Kissing Molars

Paolo Boffano, MD, Cesare Gallesio, MD, DDS

Abstract: Kissing molars are an extremely rare condition. They are impacted permanent molars that have occlusal surfaces contacting each other in a single follicular space with roots pointing in opposite directions. Unfortunately, because of the rarity of this clinical finding, it is difficult to propose clinical procedure protocols. In this article, we report the management of a patient with kissing molars.

Key Words: Kissing molars, impacted molar, surgical extraction

Kissing molars are unusual findings. As described by Van Hoof[1] in 1973, they are impacted permanent molars that have occlusal surfaces contacting each other in a single follicular space with roots pointing in opposite directions.[2] We were able to find 5 patients with this condition in the literature.[1,3–6] This report documents the management of a patient with kissing molars.

CLINICAL REPORT

A 42-year-old-man was referred to the Oral and Maxillofacial Surgery Division of the University of Turin with pain in the right retromolar region.

During a clinical intraoral examination, a 5-mm pocket was found distal to the lower right second molar. Therefore, a panorex was suggested. The panoramic radiograph revealed the presence of impacted mandibular right third and fourth molars, which had the characteristics of the so-called kissing molars (Fig. 1).

Surgical intervention was performed under local anesthesia. Local nerve block anesthesia of the inferior dental, lingual, and buccal nerves was induced with 2 capsules of 1.8 mL of 2% mepivacaine containing 1:100,000 adrenalin (Carboplyina; Dentsply Italia, Roma, Italy). The right mandibular third and fourth molars were extracted using a triangular vestibular flap that was limited to the second molar, lifting a full-thickness flap. Osteotomy necessary to visualize the impacted third molar was performed using a no. 8 tungsten carbide round bur mounted on a high-speed handpiece. To minimize the quantity of bone removed, the third and fourth molars were sectioned into 2 parts with a carbide fissure bur mounted on a high-speed handpiece. After sectioning, the 4 fragments were removed. Follicular tissue underwent histopathologic analysis. This examination did not show any evidence of disease. The remaining socket was rinsed with physiological saline. We decided to fill it with granular calcium sulfate (NewPlaster NP170; ClassImplant, Rome, Italy) to help bone regeneration in the obtained socket. Primary closure of the flap was performed using 3- silk suture. The patient was prescribed an antibiotic (1 g of amoxicillin every 12 h for 5 d [Zimox; Pfizer Italia, Milan, Italy]), a nonsteroidal anti-inflammatory drug (80 mg of ketoprofene every 12 h for 3 d [OKI granulare; Dompé, L'Aquila, Italy]), corticosteroids (1 mg of betamethasone every 12 h for 3 d [Bentelan; Defiante Farmaceutica LDA, Rome, Italy]). Postoperative instructions and use of the prescribed drugs were explained orally and on a printed sheet of paper that was given to the patient. Beginning the day after surgery, patients rinsed twice daily with 0.20% chlorhexidine solution for 10 days. The sutures were removed 10 days later. On the 10th day after intervention, the patient complained no pain, and he showed neither paresthesia nor anesthesia in the region of the chin. No sign of infection was observed. Root canals of the second right molar were treated then because the vitality test result was negative at follow-up visit on the 60th day after surgery. The patient was clinically monitored during the year after the surgery.

DISCUSSION

Extractions of kissing molars are challenging, and in asymptomatic patients, a close observation without surgery is advisable. The surgical removal of the third and fourth right mandibular molars of our patient was decided.

Preoperative assessment of surgical difficulty is fundamental to correctly plan the extraction of the impacted third molars.[7] It was extremely important to assess the various elements that could influence the extraction, such as relative depth, angulation and form of the root, number of roots, relationship of the tooth to the ramus, proximity to mandibular canal, and lack of periodontal membrane space.[8]

We decided to section both third and fourth molars to limit the bone removal that was, however, necessary because of the position of the 2 impacted teeth.

Great attention was paid not to damage the inferior alveolar nerve and not to cause iatrogenic mandibular fracture. On the 10th day after intervention, the patient showed neither paresthesia nor anesthesia in the region of the chin. No intraoral dehiscence was present. No signs of infection were observed.

When surgery is the chosen option, a bone graft to augment the weakened mandible could be considered.

Multiple rosetting of the molars has been associated with mucopolysaccharidoses.[3,9] This association should be considered in uncertain cases to perform further investigation.

Kissing molars are an extremely rare condition. Unfortunately, because of the rarity of this clinical finding, it is difficult to propose clinical procedure protocols.

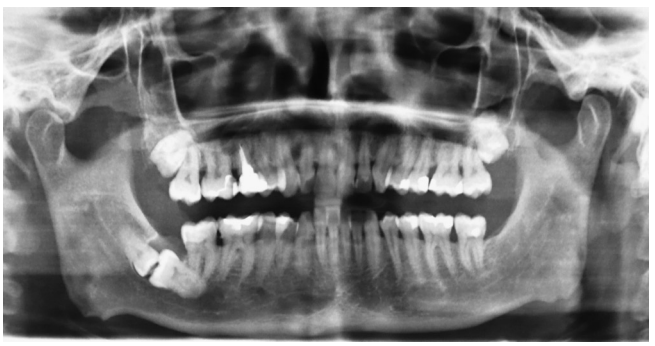

FIGURE 1. Panoramic radiograph showing impacted right third and fourth mandibular molars with occlusal surfaces contacting each other in a single follicular space.

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Carcinoma Cuniculatum: A Rare Entity in the Oral Cavity

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Background: Carcinoma cuniculatum, a well-differentiated squamous cell carcinoma, is a rare neoplasm with a low risk of metastasis. **Methods and Results:** A 74-year-old female patient is presented with a carcinoma cuniculatum of the right maxilla, a very rare variant of squamous cell carcinoma that usually occurs in the skin of the lower extremities, in particular, in the skin of the plantar surface of foot. It is described by an invasive growth pattern, but metastases to regional lymph nodes are rare. In the oral cavity, only very few cases have been published.
Conclusion: The correct diagnosis of this entity with the knowledge that it is a variant of a low-grade carcinoma with low risk of metastasis is important, so that elective neck dissection must not be obligate.

Key Words: Carcinoma cuniculatum, oral cavity, squamous cell carcinoma

Carcinoma cuniculatum (CC) was first described by Aird et al[1] in 1954.The name derives from *cuniculus* (Latin): rabbit, because of a "rabbit burrow" appearance of the complex branching keratin-filled crypts. It is considered to be a variant of verrucous carcinoma (VC). Both tumors are slow-growing lesions with endophytic and exophytic growth pattern, and both present a discrepancy between their clinical malignant appearance and their benign-appearing pathohistology with minimal propensity to metastasize to lymph nodes.[2] Carcinoma cuniculatum most commonly occurs in the skin of the plantar surface of feet. It is locally invasive, but metastases to local lymph nodes have been only rarely described.[2,3]

We present a case of CC in the anterior maxilla.

PATIENT

A 74-year-old female patient was referred to the Department of Craniomaxillofacial and Oral Surgery of the University Hospital Zurich with a 3-month history of intraoral ulceration of the anterior upper jaw (Figs. 1 and 2). Her medical and family histories did not contribute to this problem, and she had no history of smoking or alcohol abuse. The intraoral examination revealed a mass measuring 2×1.5 cm in region 15–15. No palpable lymph nodes were detected in the neck. The initial diagnosis was squamous cell carcinoma, and this opinion was supported by the histopathologic biopsy result.

The biopsy specimen presented a multilayered squamous epithelium with thin papillary projections covered by thick keratin layers and without significant cytologic atypia. No mitoses were observed. A clear invasive growth pattern could not be assessed on this superficial biopsy sample, but in correlation with imagery findings, the biopsy report was indicative of a well-differentiated squamous cell carcinoma.

Under low magnification, the superficial areas of the lesion showed a well-differentiated squamous epithelium with thin papillary projections (Fig. 3). In deeper areas, the tumor showed an endophytic growth pattern with multiple, large, branching, and

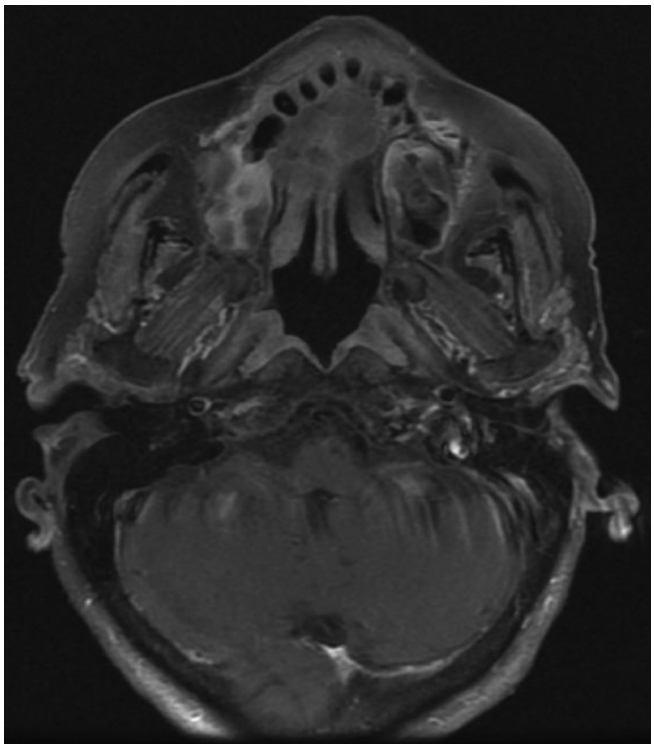

FIGURE 1. Magnetic resonance imaging scan of tumor mass of the upper jaw.

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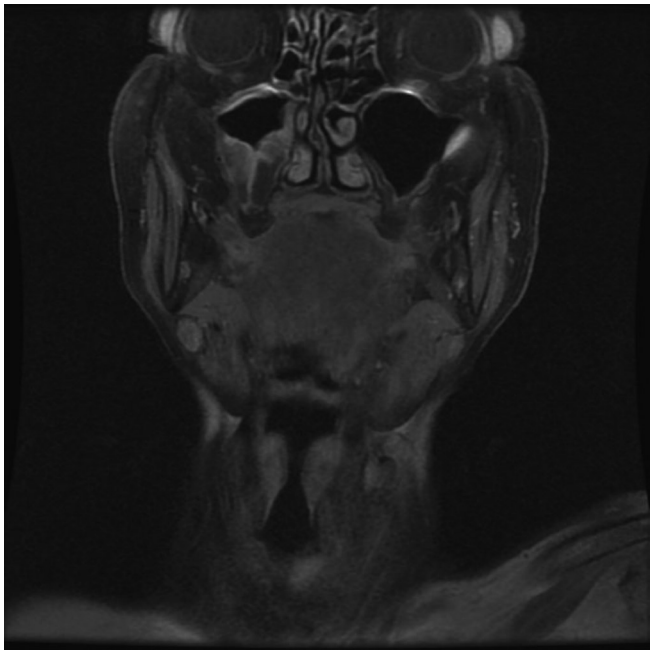

FIGURE 2. Tumor infiltration into the right maxillary sinuses.

interconnected cysts. Focal bone destruction was observed. The keratin-filled cysts were surrounded by a well-differentiated squamous epithelium. Tumor cells exhibited only mild cytologic atypia and few mitoses. Immunohistohemistry results for Ki-67 showed proliferative activity in basal and suprabasal layers of the epithelium (Fig. 4). The tumor cells were negative for p53. The peculiar endophytic invasive growth pattern and the mild cytologic atypia were typical of CC. No evidence of human papillomavirus could be demonstrated.

An anterior maxillectomy including bilateral selective neck dissection level I-III with immediate prosthodontic rehabilitation (obturator) was performed. No recurrence or lymphadenopathy was observed over a 2-year follow-up period.

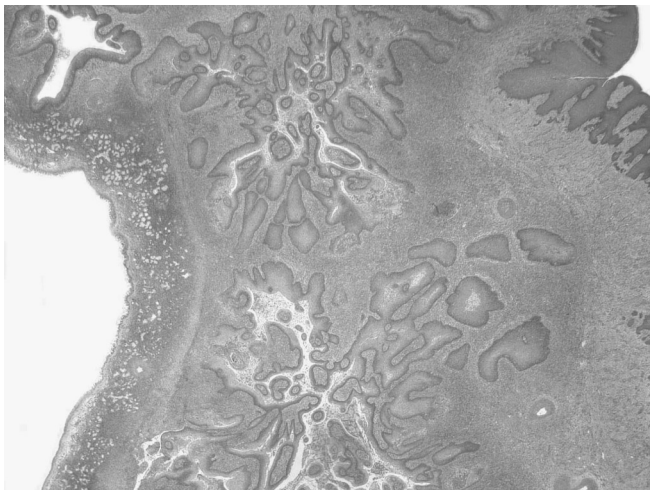

FIGURE 3. Keratin-filled crypt surrounded by a well-differentiated squamous epithelium.

DISCUSSION

Carcinoma cuniculatum, the rare variant of squamous cell carcinoma, appears typically as a slowly growing lesion with a long history of ulceration. It has been described most frequently in the cutis, particularly in the plantar surface of the feet. Concerning oral mucosa, only 19 cases have been reported in the literature (Table 1); female-male ratio was 5:14; mean age, 55.4 years; and primary location was 36.8% in the upper jaw.

Histologic characteristics of CC is the endophytic growth with multiple expansive and branching keratin-filled cysts, hence the appellation of cuniculatum (*cuniculus*, rabbit burrow). The cysts are covered by a well-differentiated epithelium with no or only mild cytologic atypia and few mitoses. Differential diagnosis includes other variants of squamous cell carcinoma such as papillary cell carcinoma. On biopsy samples, assessing the malignant character of CC can be quite challenging because the infiltrative growth pattern is not evident on superficial probes. In addition, categorical criteria

Table 1. Review of 19 Published Cases of Carcinoma Cuniculatum of the Upper Aerodigestive Tract (1977–2008)

Author	Year	Patient	Age, y	Sex	Location
Flieger and Owiński[4]	1977	1	50	M	Upper jaw alveolus
		2	60	M	Upper jaw alveolus
		3	9	M	Upper jaw alveolus
		4	69	F	Hard palate
Kahn et al[5]	1991	5	52	M	Anterior floor of mouth
		6	62	M	Upper jaw alveolus
		7	54	F	Lower jaw alveolus/ floor of mouth
		8	49	F	Lateral floor of mouth
Delahaye et al[6]	1994	9	51	M	Retromolar
		10	55	M	Peritonsillar/ floor of mouth
		11	63	M	Larynx
		12	31	M	Hard palate
		13	52	M	Buccal mucosa
Huault et al[7]	1998	14	55	F	Lower jaw alveolus
Allon et al[8]	2002	15	56	M	Upper jaw alveolus
De Petris et al[3]	2005	16	73	M	Esophagus
		17	58	M	Esophagus
Raguse et al[9]	2006	18	81	F	Lower jaw alveolus
Puxeddu et al[10]	2008	19	72	M	Larynx

M indicates male; F, female.

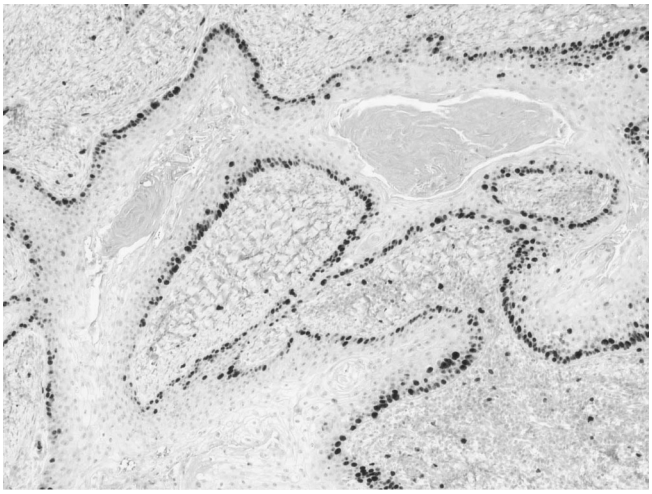

FIGURE 4. Ki-67 staining showing proliferative activity in basal and suprabasal cell layers of a well-differentiated squamous epithelium.

of malignancy are missing. Immunohistochemistry (E-cadherin, β-catenin, Ki-67, p53) is of no help in differential diagnosis because normal and tumoral epithelium show the same staining pattern. Knowledge of the clinics and correlation with radiologic findings are necessary for histologic interpretations and diagnosis on biopsy samples.

Concerning differential diagnosis in the oral mucosa, CC keratin-filled crypts are typical, and in VC, the keratinization pattern is vertical or "church-spire–like." The pattern of hyperplasia in CC is a formation of canaliculi, and in VC, a bulbous expansion. In comparison to papilloma with no inflammation and papillary squamous cell carcinoma with a moderate chronic, nongranulomatous inflammation, VC and CC show heavy inflammation.[3]

Etiology and pathogenesis of CC are still unknown. A human papillomavirus association has been discussed[11] but cannot be verified in all cases.[8] Alcohol and tobacco were discussed as possible etiologic factors in some reported cases.[3]

Epidemiologic data are less to be found in literature. Bouquot et al[11] described in US population an incidence rate of 0.1 per 100,000 person-years.

CONCLUSION

The correct diagnosis of this entity with the knowledge that it is a variant of a low-grade carcinoma with a lower risk of metastasis is important, so that neck dissection must not be obligate.

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Chordoma in Paranasal Sinuses

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Abstract: Chordoma is a rare malignant intercranial tumor. Cranial base chordomas usually occur extradurally. In the current study, a 44-year-old man with chordoma in cranial base and paranasal sinuses was reported. The patient was managed with combination of surgery and radiotherapy. Surgical phase comprised anterior cranial base osteotomy, removal of lesions, and replacement of osteotomy segment. One-year follow-up showed no evidence of recurrence of lesions. Aggressive surgery with postoperative radiotherapy for treatment of chordoma in the cranial base can be suggested as a proper treatment modality.

Key Words: Chordoma, tumor, paranasal sinus

Chordoma was first described by Luschka[1] in 1856. Chordoma is a rare intercranial tumor (approximately 0.2% of 6000 brain

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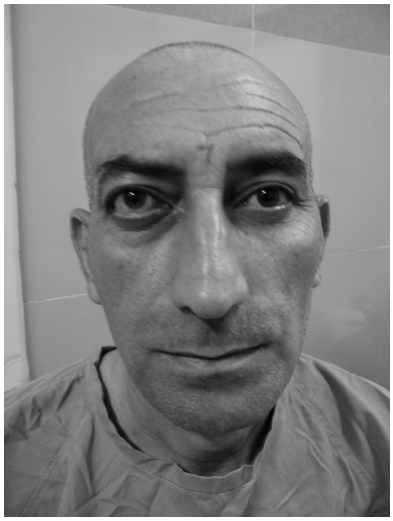

FIGURE 1. Preoperative view of patient showed severe exophthalmos in the right eye.

tumors).[2] Chordomas develop from remnants of notochord. The remnants of notochord are usually found only in the nucleus pulposus of intervertebral disks but may be seen in the clival bone marrow and even in the intradural clival area.[3,4] Cranial base chordomas usually occur extradurally, although chordomas may also occur intradurally in a few patients.[5] Extradural chordomas invade the dura mater laterally, especially in aggressive tumor. Chordoma is a midline tumor, but a few cases of lateral origins have been reported.[6] Macroscopically, chordoma is soft and gelatinous, with a smooth or lobulated grayish white outer surface. Its surface is homogeneous, but calcification and infrequently hemorrhage are not unusual.[2] Chordoma is a malignant lesion with population of cells that have small, oval, or round eccentric nuclei with a dense chromatin pattern. Multiple vacuoles of different sizes are a specific feature of these cells, leading to the term *physaliphorous cells*. Capsulation is not seen with chordoma.[4] Chordoma is a locally aggressive, slow-growing tumor with a high local recurrence rate.[7–9]

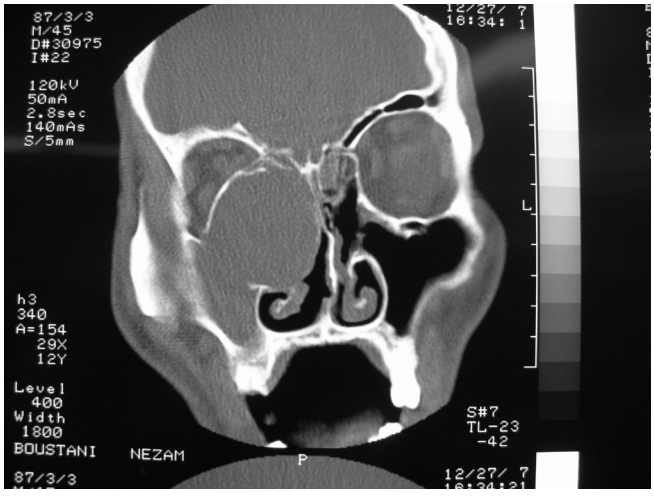

FIGURE 2. Computed tomography scan shows huge tumor in paranasal sinuses with invasion to the right orbit.

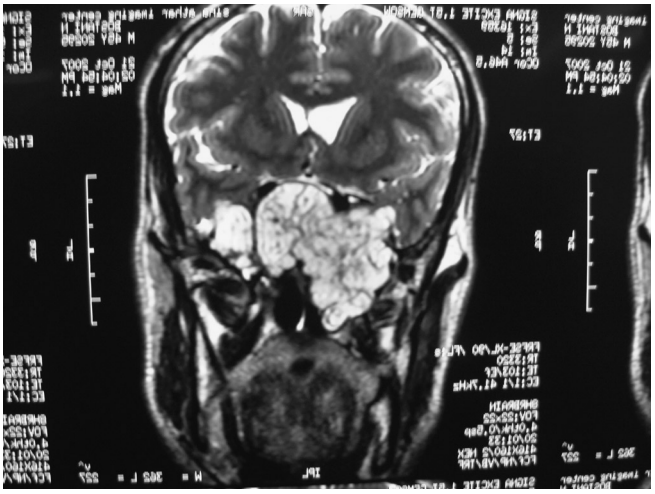

FIGURE 3. Magnetic resonance imaging view reveals hyperdense, lobulated, huge tumor in the anterior cranial base.

This report describes the treatment of a skull base and paranasal chordoma in a 44-year-old man.

CLINICAL REPORT

A 44-year-old man with a history of intercranial surgery for removal of tumor in 2003 presented to the Department of Oral and Maxillofacial Surgery, Craniomaxillofacial Surgery Research Center, University of Shiraz Medical Science, Shiraz, Iran, in 2006. In his initial surgery, the patient with loss of right eye vision managed with surgical removal of the tumor without radiotherapy. Chordoma has been reported by the pathologist right after the biopsy was taken in the first surgery.

In his first visit to the clinic, the patient expressed his complaint of severe, gradually progressive headache and diplopia in his left eye for several months. Clinical examination revealed the presence of severe pain and diplopia in the left eye. In the radiologic examination, computed tomography scan revealed a huge tumor mass in intercranial frontal lobe with invasion to anterior cranial base and paranasal sinuses. Extensive destruction in medial wall of the right eye was seen (Figs. 1 and 2). Magnetic resonance imaging views revealed the presence of hyperdense, lobulated, huge tumor in the anterior skull base (Fig. 3).

The patient was operated on in the Department of Neurosurgery for removal of tumor intercranially in the first surgery. After 2 months, the patient was referred to our department for treatment of the skull base and paranasal extension of tumor for the second surgery. Further evaluation showed no metastases in other sites of the body. Both eyes lost vision. He had headache and nasal stuffiness. Throughout the surgery, subcranial extended anterior approach was performed for skull base tumors. Osteotomy of the frontonaso-orbital external skeletal frame provided optimum anterior access to the orbital and sphenoethmoid planes and to the nasal and paranasal cavities (Fig. 4). After enucleation of tumors, polyethylene tubes were inserted bilaterally into the subcranial compartment and externalized through the nasal lumen. Then, osteotomy segment was replaced and fixed with miniplate and screws (Fig. 5). He was referred to radiotherapy 4 weeks after the second surgery. One-year follow-up revealed no evidence of infection or cerebrospinal fluid leakage. After biopsy was performed, the histologic examination showed lobulated mass composed of cuboidal to

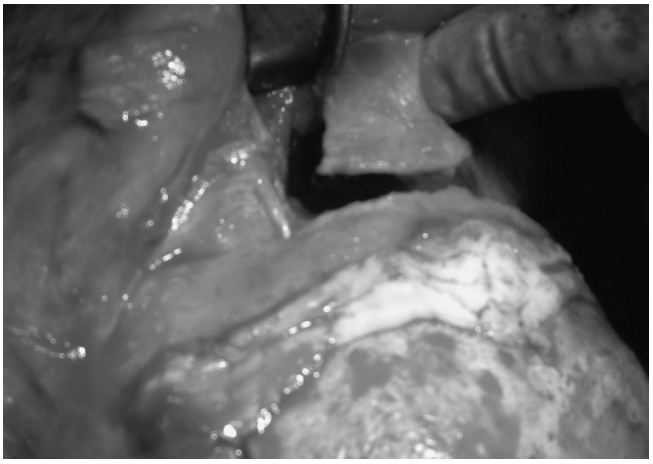

FIGURE 4. Intraoperative view of the anterior cranial base osteotomy.

polygonal clear cells with eccentric nuclei and pools of extracellular mucin. Few tumor cells typically had the appearance of physaliferous cells. The cells were positive for antikeratin monoclonal antibodies AE1/3 epithelial membrane antigen, S100, and vimentin. According to these histologic and immunohistochemical findings, the diagnosis of chordoma was justified (Fig. 6).

DISCUSSION

Chordomas are slow-growing, uncommon malignant tumors that occur from the embryonic remnants of the notochord along the craniococcygeal axis.[10] They are classified in relation to the region of origin into craniocervical, vertebral, and sacrococcygeal groups. The most common location is in the sacrococcygeal region, and chordomas rarely arise in other regions of the vertebral column.[11] The greatest incidence is found in the fifth and seventh decades.[12]

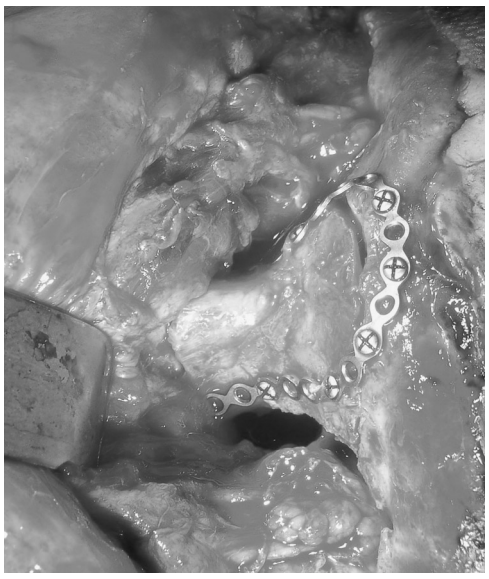

FIGURE 5. Intraoperative view showed replacement osteotomy segment after removal of lesions.

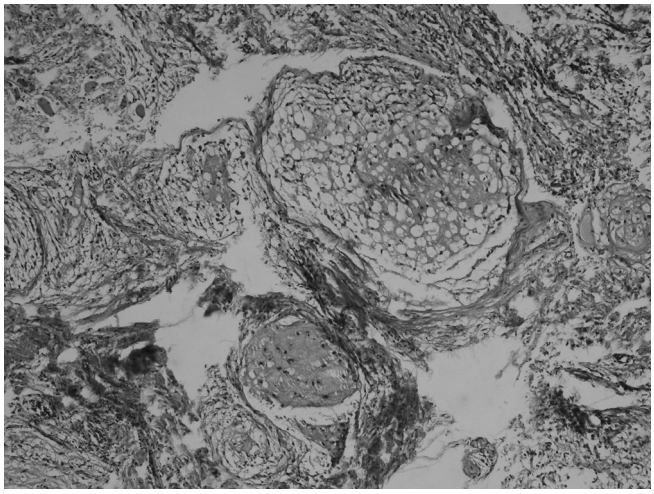

FIGURE 6. Histologic view of lesion demonstrates a chordoma.

Chordomas are tumors with low metastatic potential because only 10% spread to distant sites.[13] Most patients with chordomas present with headaches and diplopia, which are observed in about 55% to 60% of patients.[14]

Symptoms of diplopia tend to be intermittent in approximately half of these patients. According to our experience, the patients had severe headache and diplopia, which progress to blindness. Local extension of these tumors into the retropharyngeal area or nasal cavity can account for dysphagia, nasal obstruction, and epistaxis. Most primary chordomas of the paranasal sinuses are found in women in their third decade of the life.[15] All primary chordomas of the paranasal sinuses are slow growing and locally invasive.[16] They usually destroy the adjacent structures. Computed tomography scan without contrast enhancement usually shows hyperdense lesion, but some tumors appear isodense. On contrast-enhanced computed tomography scan, almost all tumors show enhancement with varying degrees. Magnetic resonance imaging is very helpful in revealing the extent of the tumor and its margin. Ideal treatment for chordomas is controversial and depends on the experience of the surgeon. The aim of this treatment is to increase survival rate along with decrease the possible risk of recurrence. Surgery, surgery followed by radiation, and biopsy followed by radiation are recommended by authors in the treatment of chordomas. Many patients need multimodality therapy. Because of high local recurrence rate and area of tumors, extensive surgery is preferable. Most authors agree that surgical resection is important for a preferable outcome.[17,18]

It is difficult to be sure of achieving absolute resection and obtaining a frozen section of bony margin. Radiotherapy is still controversial but often applies to incomplete excision case or palliation of recurrence. Because complete resection of chordomas in the skull base and paranasal region is extremely difficult or impossible, postsurgical radiotherapy may be also effective in controlling local invasion.

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Surgical Treatment of an Oral Cyst With Respiratory Epithelium

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Abstract: Cysts with respiratory epithelium are rare entities in the oral cavity. In the literature, there are only few cases of oral cystic masses lined by respiratory epithelium but lacking gastrointestinal epithelium. In this paper, we present the surgical treatment of a cyst with respiratory epithelium in the floor of the mouth affecting a 35-year-old woman. The asymptomatic lesion was noticed 6 years previously. On histopathologic examination, the cyst showed walls that were composed of connective tissue covered by pseudostratified ciliated columnar epithelium, interspersed by a few goblet cells. Oral cysts lined by respiratory epithelium are benign lesions. Cure is effected by surgical excision. This lesion should be considered in the differential diagnosis of masses involving the anterior tongue or the floor of the mouth.

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Key Words: Oral cyst, respiratory epithelium, lingual choristoma, foregut duplication cyst

Cysts with respiratory epithelium are rare entities in the oral cavity. They are often called lingual choristomas or foregut duplication cysts.[1] When they do occur, they usually involve the tongue or, more rarely, the soft tissue of the floor of the mouth. In the literature,[1–4] there are only few cases of oral cystic masses lined by respiratory epithelium but lacking gastrointestinal epithelium (Table 1).

In this paper, we present the surgical treatment of a cyst with respiratory epithelium in the floor of the mouth.

CLINICAL REPORT

A 35-year-old woman presented to the Maxillofacial Surgery Division at San Giovanni Battista Hospital in Turin with a swelling in the floor of the mouth, which was noticed 6 years previously. Initially, the lesion was asymptomatic, but later, it slightly grew. No difficulty in eating or respiratory compromise was reported.

TABLE 1. Oral Cysts Lined by Respiratory and Squamous Epithelium, Without Gastrointestinal Epithelium

Author of the Article	Age of the Patient	Sex of the Patient	Histopathology
Manor et al[2]	11 years old	Male	Respiratory epithelium and cuboidal cells
Kim et al[3]	27 years old	Male	Respiratory epithelium, squamous metaplasia, and calcium deposition in the cyst cavity
Ho and Crean[4]	6 weeks old	Male	Keratinized squamous epithelium, supported by fibrous tissue, adipose tissue, sebaceous glands, and hair follicles
Burkart et al[1]	Infant	Male	Squamous epithelium, interspersed focally by respiratory type epithelium, mucous glands, and dermal appendages
Burkart et al[1]	Infant	Male	Squamous epithelium and focal areas of ciliated pseudostratified respiratory epithelium, mucous glands, and lymphoid tissue

On examination, the surface appearance of the floor of the mouth was normal. Palpation showed a 1 × 2-cm soft, tender mass in the oral floor.

Under local anesthesia, an incision in the anterior floor of the mouth was made, exposing a uniloculated mass. Then, the cyst was enucleated by blunt dissection.

Grossly, the specimen consisted of a cystic round mass of elastic consistency, 2 cm in diameter. It showed a smooth outer surface, and it contained a yellowish cloudy material. The cyst was submitted for pathologic examination.

Histopathologic examination result demonstrated that the cyst was composed of connective tissue covered by pseudostratified ciliated columnar epithelium, interspersed by a few goblet cells (Figs. 1 and 2).

The immediate postoperative course was uneventful, without significant edema of the floor of the mouth. Healing was normal, and there has been no recurrence a year after operation.

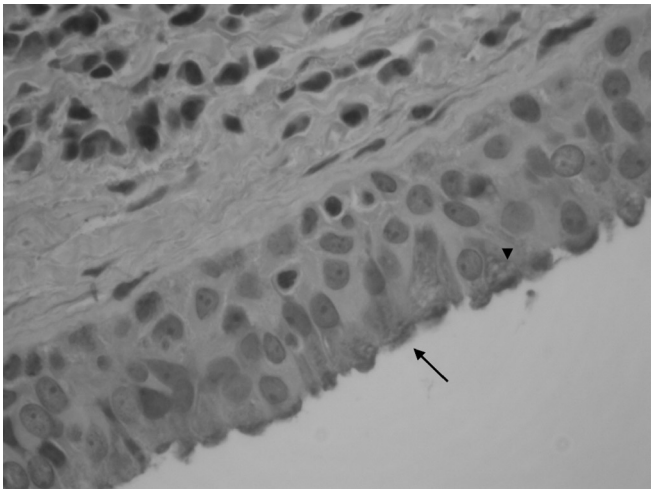

FIGURE 2. Representative histologic section of the cyst, demonstrating respiratory epithelium with ciliated cells (arrow) and mucinous cells (arrowhead; hematoxylin-eosin, original magnification ×160).

DISCUSSION

When a mass is encountered involving the anterior tongue or the floor of the mouth, a wide spectrum of entities must be considered in the differential diagnosis; these include teratoma, dermoid cyst, thyroglossal duct cyst, hemangioma, lymphangioma, mucocele or ranula, epidermoid cyst, lymphoepithelial cyst, salivary gland neoplasms, and lingual choristomas or foregut duplication cysts.[1,5]

The cyst lined by respiratory epithelium usually manifests itself as an asymptomatic swelling in the floor of the mouth. In approximately 30% of those affected, difficulty in feeding, swallowing, respiration, and speech have been reported.[5]

The presence of respiratory-type mucosa in oral cysts is attributable to the fact that the tracheobronchopulmonary tree, such as part of the mouth, also develops from the endoderm of the primitive foregut.

In the literature,[6–9] several theories regarding the embryogenesis of cyst lined by respiratory epithelium have been described. This particular cyst could arise from a developmental derangement of the foregut. During differentiation in the third week of embryonic development, the foregut undergoes a dichotomy, a ventral part that forms the respiratory tract and a dorsal part that creates the esophagus, stomach, and duodenum. Because of the proximity of the primitive foregut to the pharyngeal arches (which contain the developing tongue), embryonal rests may be misplaced and entrapped between the parts of the tongue, the lateral lingual swellings and the tuberculum impar, during the fourth week. These entrapped rests, which are pluripotential, are exposed to different environments and inductive influences and can differentiate to respiratory or well-organized, highly differentiated gastrointestinal epithelium.[2,10]

In our case, given that the pathophysiology of these lingual cysts is poorly understood, we did the diagnosis of cyst with respiratory epithelium according to the indications of Manor et al,[2] who suggested descriptive terminology rather than current terminology of lingual cyst of foregut origin.

Oral cysts lined by respiratory epithelium are benign lesions. They can be treated by surgical excision that seems to be efficacious with relatively minimal morbidity.

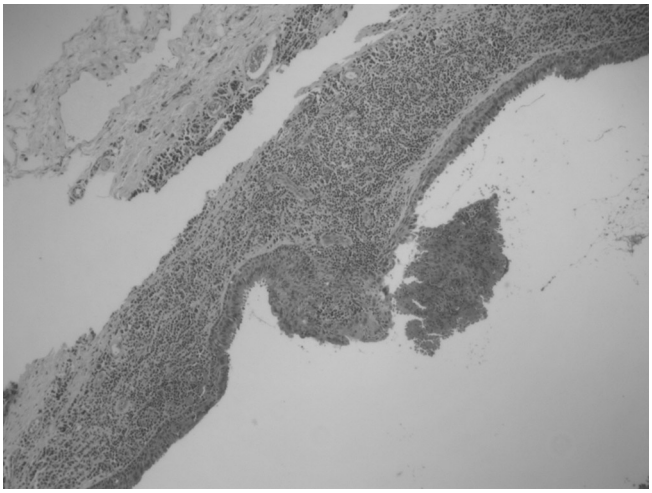

FIGURE 1. Representative histologic section of the cyst, demonstrating epithelial cyst with subepithelial chronic inflammation (hematoxylin-eosin, original magnification ×8).

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Successful Treatment of a Huge Congenital Cervical Teratoma

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Abstract: Congenital cervical teratomas are extremely rare tumors with high perinatal mortality and morbidity rates particularly due to compression and distortion of the infant's airway. Hence, these mostly benign malformations require immediate excision, whereas surgery of these tumors is challenging for a multidisciplinary team. We report on a recent case of congenital cervical mature teratoma with total excision and cure. The aim of this case study is to report the authors' experience in managing a case of congenital cervical teratoma to provide a structured approach and help in decision making, once prenatal diagnosis is made.

Key Words: Congenital, cervical teratoma, surgical therapy

Congenital cervical teratomas are extremely rare because they account for approximately 1.5% to 5.5% of all pediatric teratomas.[1] The incidence is reported to be between 1 in 20,000 and 1 in 80,000 live births.[2,3] In some studies, associations to other deformities are described.[4–6] Teratomas are composed of tissue originating from all 3 embryonic layers as they derive from pluripotential primitive germ cells. Histologically, they are classified as mature, benign in 95% of cases, immature, and malignant transformation. Immature elements in congenital cases are not considered a sign of malignancy but conditional on the infant's immaturity. The term "malignant" is kept for cases with histologic signs of malignancy or with distant metastases.[7,8] Such a malignant transformation occurs in less than 5% of all congenital cervicofacial teratomas.[1] Differential diagnoses are connatal malformations such as cervical cysts, struma, vessel malformations, lymphangioma, thymus cysts, and dermoid cysts.[9] Clinical features that indicate a teratoma are a well-circumscribable tumor with both solid and cystic configuration. Calcifications could be a further hint for a teratoma because they are reported in 50% of the cases.[10] Depending on the localization and size, the tumor can be diagnosed prenatally by ultrasonography.[11] This allows for a safer planning of the parturition and the following surgical therapy.

CLINICAL REPORT

A 28-year-old female gravida 1, para 0, at 32 weeks gestation, was referred with polyhydramnios for antenatal ultrasonography. In comparison to the routine antenatal ultrasound at 27 weeks gestation, it revealed the presence of a large cervical mass (72 × 55 × 66 mm) and confirmed the presence of polyhydramnios (Fig. 1). Amniotic fluid reduction by amniocentesis was undertaken at this time. A lymphangioma was suspected. For observation and preparation of the delivery and the postnatal multidisciplinary therapy, the woman was hospitalized. At 32 weeks of pregnancy, a caesarean section was performed due to ruptured membranes. An alive boy with a body weight of 2700 g and a body height of 47 cm was delivered, exhibiting a large particularly right-sided neck mass (Fig. 2). Orotracheal intubation was successfully performed after delivery, and the child was transferred to the neonatal intensive care unit.

Respiratory distress syndrome and hemodynamic instability required mechanical ventilation, the replacement of surfactant and treatment of hypotension. Initially, there was no chance to probe the esophagogastric way. Total parenteral nutrition was necessary until surgical intervention. A magnetic resonance imaging was carried out on the sixth postnatal day, which showed a 90 × 50 × 60-mm complex lesion containing solid and cystic regions, consistent with a teratoma. It was seen to pass across the midline in the anterior neck across the trachea. The trachea and the carotid sheath were displaced to the posterior space. No tumor infiltration of the adjacent structures was observed (Fig. 3).

Further growth of the tumor required early surgery, and extirpation of the tumor was processed on the 11th day of life. A transversal

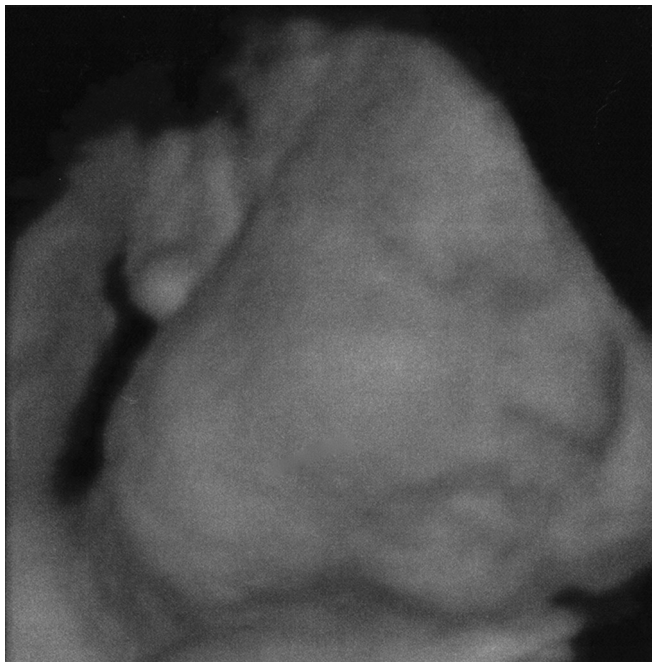

FIGURE 1. Ultrasonography at 32 weeks gestation.

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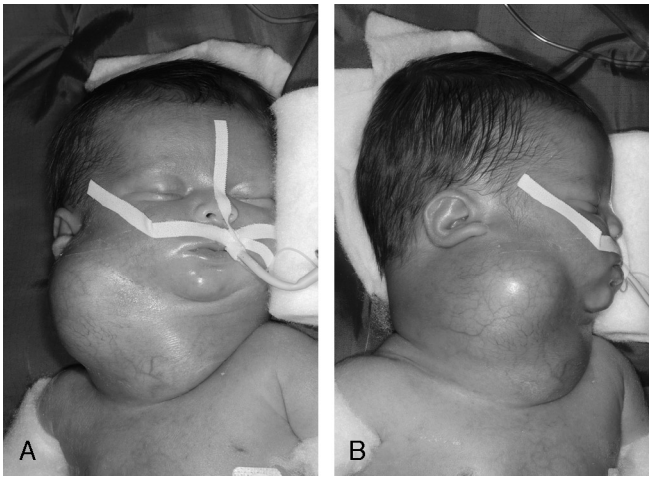

FIGURE 2. Preoperative situation: frontal view (A) and lateral view (B).

incision over the lesion was performed and brought out an encapsulated tumor. Dissection proceeded along the capsule of the lesion. The 100 × 50 × 60-mm tumor adherent to the upper part of the trachea was resected en bloc, and all significant locoregional structures (carotid artery, internal jugular vein, larynx, trachea, esophagus, left lobe of the thyroid gland) were identified and preserved (Fig. 4). After tumor extirpation, intubation was changed from orotracheal to nasotracheal before wound closure ensuring a correct tube position. Histologic investigation revealed a 200-g mature teratoma containing epithelial and mesenchymal structures with existence of chondral and thyroid tissue. Therefore, we concluded that the teratoma had arisen in the right lobe of the thyroid gland because it meets the criteria defined in the literature.[12–14]

The postoperative period was complicated by a capillary leak syndrome with massive general edema. However, the infant fully recovered after forced diuresis and was extubated 2 weeks postoperative offering an adequate respiration. Except for a mild larynx stenosis, a bronchoscopy carried out before extubation revealed no pathologic findings. The boy was discharged at the age of 8 weeks without any respiratory or neurologic deficit (Fig. 5). At this time, thyroid hormones were arranged in the lower normal range. Parathormone was located in the upper reference range accompanied by a regular calcium level. The value of α-fetoprotein, which was

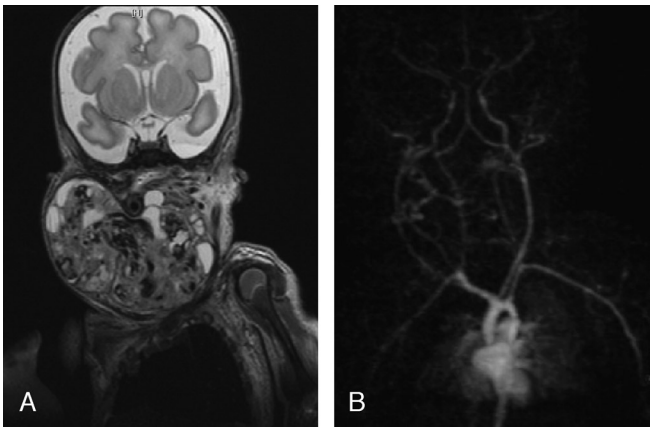

FIGURE 3. A, Magnetic resonance imaging of the tumor. B, Angiography of the head and neck vessels.

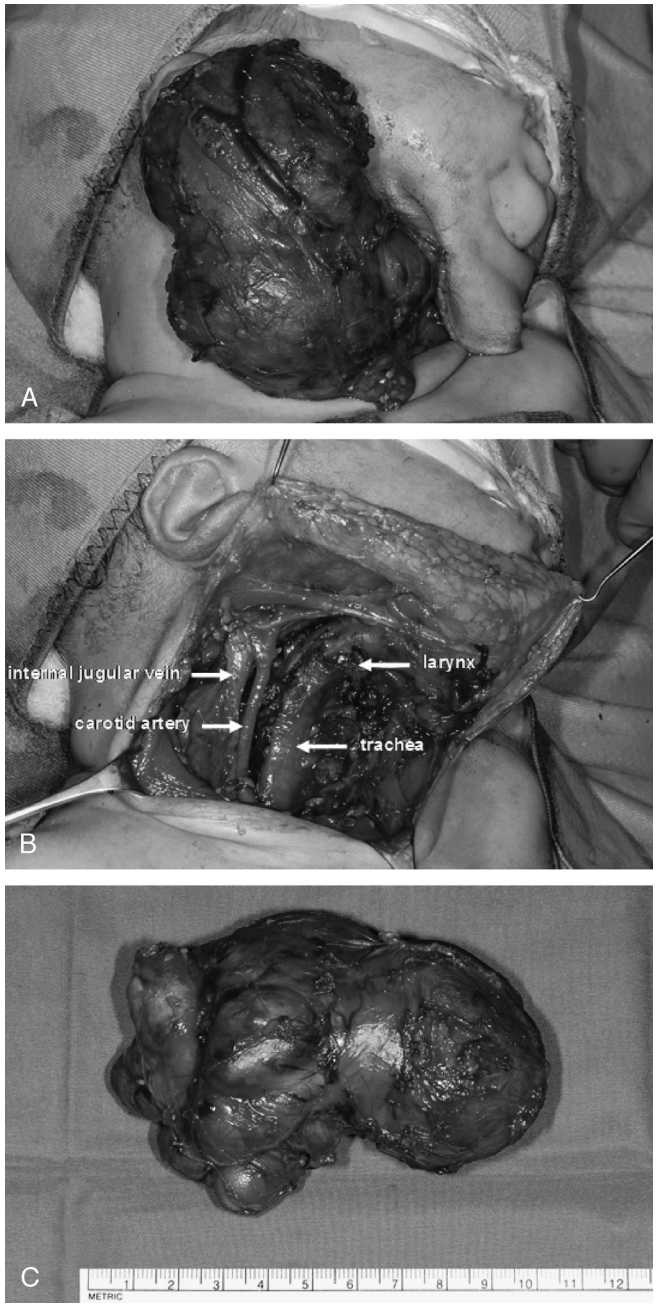

FIGURE 4. A, Intraoperative situs before tumor removal. B, Intraoperative situs after tumor extirpation. C, Tumor en bloc.

149,323.6 ng/mL after delivery, decreased continuously up to an extent of 1,324.8 ng/mL at the time of discharge.

DISCUSSION

Although congenital cervical teratomas are usually histologically benign, the untreated mortality ascribed to these lesions is 80% to 100% because of compression and distortion of the infant's airway caused by the significant size they may attain.[10] Prenatal diagnosis is usually reached with the aid of ultrasonography after the 15th week

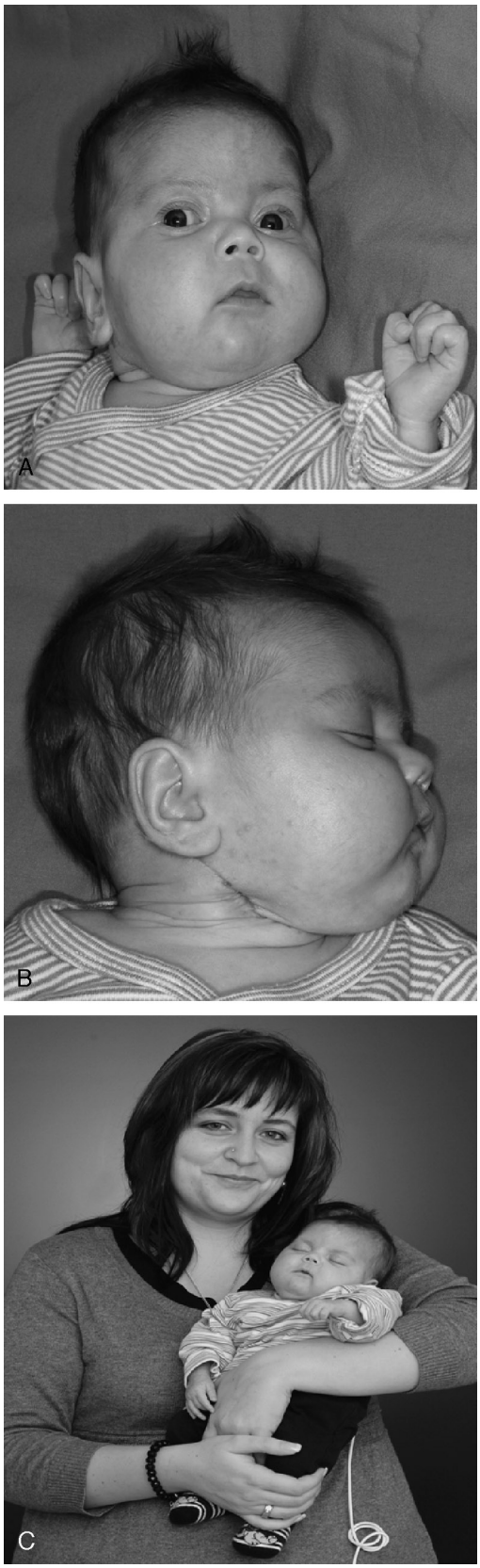

FIGURE 5. Patient 6 weeks postoperative. frontal view (A), lateral view (B), and patient with his mother (C).

of gestation, detecting the tumor mass and its vascularization plus a polyhydramnios.[15] Fetal magnetic resonance imaging represents a complementary technology because this is also a safe and noninvasive modality.[16] A quantitative analysis of α-fetoprotein[17] and a cytogenetic study[18] could be continuative. These screenings help in preparing a multidisciplinary team to plan parturition, postnatal airway support, and surgical intervention once decision for continuing the pregnancy is made.

Up to 50% of the newborns with huge cervical teratomas experience severe dyspnea immediately after delivery.[19] The prognosis is determined by the severity of the associated airway obstruction. Ex-utero intrapartum treatment is a technique to establish a patent airway before completion of birth. It provides the luxury of time to obtain airway control by intubation, or tracheostomy, or, if necessary, tumor resection on placental support.[20] Rapid growth of teratomas is reported for many cases. Therefore, postnatal intubation is recommended even if the newborn does not show any dyspnea, initially.[21]

Complete surgical removal is curative. In a review containing approximately 200 cases, the perioperative and postoperative mortality is stated to be 13%.[1] In contrast, untreated cases have an unfavorable prognosis.[22]

To the authors' mind, if possible, a tracheotomy should be avoided to decrease the postoperative morbidity. But then, preoperatively and postoperatively, a tracheotomy is often inescapable because in many cases, it represents the only method to save the airway.

Thyroid and parathyroid insufficiency may be present at birth in cervical teratomas and can be aggravated by surgery. Such circumstances necessitate a substitution with thyroxine, vitamin D, and calcium.[23] In the case presented here, postoperatively, thyroid and parathyroid hormones were arranged in the reference range, indicating that no serious injury of the respective glands has occurred. Nevertheless, a periodical checkup is recommended also to control problems associated with prematurity in our patient, especially the neurologic outcome. Against the background of the fact that teratomas are typically correlated with a high level of α-fetoprotein, this marker should be controlled regularly as well. After tumor removal, the level of α-fetoprotein decreases continuously up to the reference value. A new increase of α-fetoprotein could be a sign of recrudescence.[17] Another part of the aftercare is a 6-monthly computed tomography or magnetic resonance imaging to discover a recurrence.

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Medial Orbital Wall Reconstruction Through Subciliary Approach: Revisited

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Abstract: The aim of this study is to determine the safety and complication of subciliary approach through the retrospective review of our experiences.

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From 2005 through 2008, the subciliary skin-muscle flap methods were used in 30 patients undergoing medial orbital wall reconstruction. Preoperative and postoperative ophthalmic findings including diplopia, Hertel exophthalmometry, and occurrence of complications were checked. Resorbable polylactic acid sheet or porous polyethylene sheet was trimmed and molded in L shape, vertical portion to cover the medial wall defect and horizontal portion for stability in orbital floor.

In the follow-up of diplopia, half of the patients (3 cases) presenting with diplopia improved during the first month of follow-up, and all of them improved by 6 month. For hypesthesia, all patients improved by 3 months. Enophthalmos of 1 patient improved after operation and did not recur. No patients complained of visible scar 6 months postoperatively, and no ectropion was observed.

We think that medial orbital wall could be reconstructed safely through skin-muscle flap subciliary approach without resulting in ectropion or lacrimal canaliculus injury.

Key Words: Orbital fractures, reconstructive surgical procedures

Surgical approaches to the medial orbit have been driven by access, safety, cosmesis, and versatility.

Since the first of introduction of medial cutaneous approach by Lynch, several methods have been presented.[1] The subciliary incision is rendered a higher risk of scarring and lid retraction than transconjunctival approach.[2] Transconjunctival approach provides only limited exposure.[3] Although transcaruncular approach is growing in popularity,[4,5] this approach carries a risk of early ophthalmic complication including underaction of inferior oblique and conjunctival scarring.[6]

Since 2005, in an effort to achieve better exposure of the medial orbital walls, we have used mainly a subciliary approach to the medial orbit.

The aim of this study is to determine the safety and complication of subciliary approach through the retrospective review of our experiences.

MATERIALS AND METHODS

From 2005 through 2008, the subciliary skin-muscle flap methods were used in 30 patients undergoing medial orbital wall reconstruction. The patients' ages ranged from 16 to 56 years (mean age, 32.1 years). There were 24 men and 6 women. Eight patients were reconstructed with porous polyethylene sheet (Medpor; Porex Surgical Inc, Newnan, GA), whereas 22 patients were reconstructed with resorbable polylactic acid sheet (Inion Co, Tampere, Finland).

Preoperative and postoperative ophthalmic findings including diplopia, Hertel exophthalmometry, and occurrence of complications were checked.

OPERATIVE TECHNIQUE

Surgery was performed on the patient under general anesthesia. Subciliary incision was designed and infiltrated with 2 mL of 1% lidocaine solution containing epinephrine (1:100,000). After the incision through skin and orbicularis oculi muscle, skin-muscle flap was elevated just superficial to the orbital septum to the arcus marginalis. Dissection was extended just below the anterior lacrimal

crest to increase the exposure. Using the malleable retractor, fracture site in medial orbital wall was visualized.

Resorbable polylactic acid sheet or porous polyethylene sheet was trimmed and molded in L shape, vertical portion to cover the medial wall defect and horizontal portion for stability in orbital floor (Fig. 1). After the molded plate was put in position, periosteum and skin were closed.

RESULTS

On physical examination before surgery, diplopia was the most common (58.5%) associated complication, followed by hypesthesia (19.5%), enophthalmos (19.5%), limitation of ocular movement (19.5%), and hematoma (2.4%). The cases were analyzed according to the reconstruction materials (Table 1).

The postoperative complication rate was 10.0% (3 cases among 30), and these were diplopia (2 cases) and enophthalmos (1 case) (Table 2).

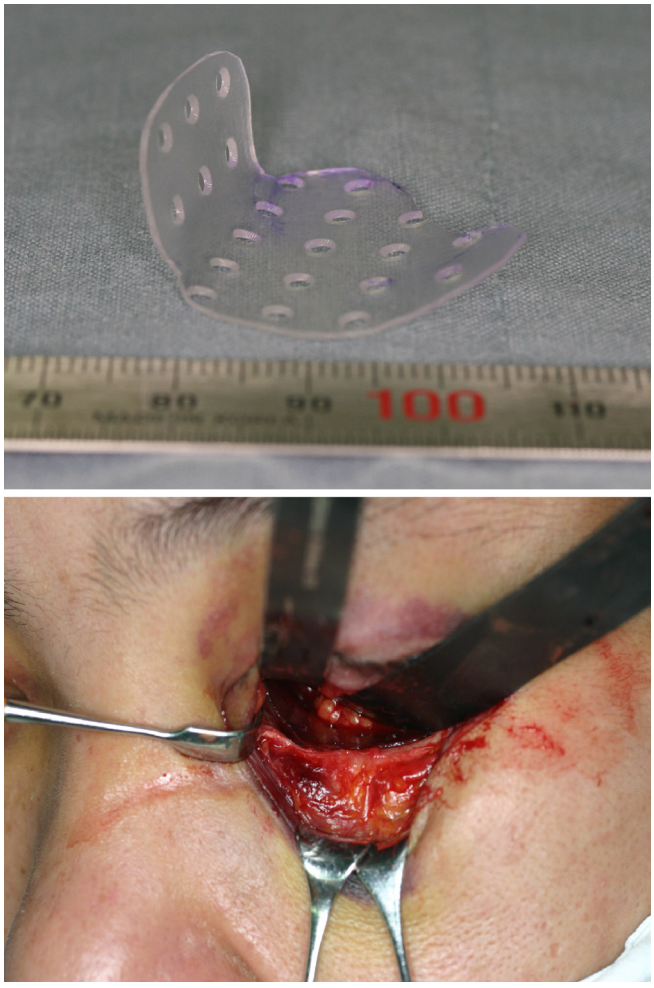

FIGURE 1. Resorbable polylactic acid sheet was trimmed and molded in L shape, vertical portion to cover the medial wall defect and horizontal portion for stability in orbital floor (Top). After the molded plate was put in position, periosteum and skin was closed (Bottom).

Table 1. Signs and Symptoms of Presentation in Preoperation Associated With Medial Orbital Wall Reconstruction Through Subciliary Approach

Physical Examination	Medpor	Resorbable Sheet	Total, n (%)
Diplopia	26	19	24 (48.4)
Hypoesthesia	9	7	16 (17.2)
Enophthalmos	4	8	12 (12.9)
Limitation of ocular movement	7	0	1 (7.5)
Others	0	0	0 (0)

In the follow-up of diplopia, half of the patients (3 cases) presenting with diplopia improved during the first month of follow-up, and all of them, both the Medpor and resorbable sheet groups, improved by 6 months (Table 3; Fig. 2).

In the follow-up of hypesthesia, all patients improved by 3 months (Table 3; Fig. 3). Enophthalmos of 1 patient improved after the operation and did not recur by 1-year follow-up (Table 3).

DISCUSSION

Holtmann et al[3] reported on ectropion in 42% patients after subciliary incision. In Figure 4 of their study, skin incision was 2 mm below lid margin, inferior skin flap was elevated, and incision was made through orbicularis oculi muscle and periosteum over infraorbital rim.

The outcome of our study revealed that among 30 patients operated on with subciliary incision, no patients complained of visible scar 6 months postoperatively. Furthermore, no ectropion was observed. The reason for low incidence of ectropion, we think, is the method of flap elevation. We used skin-muscle flap instead of skin flap. We think that skin-muscle flap might prevent ectropion.

The transcaruncular approach carries a risk of ophthalmic complications, including permanent underaction of inferior oblique, or

Table 2. Comparison of the Postoperative Complications Associated With Medial Orbital Wall Reconstruction Through Subciliary Approach

Complications	Medpor, n (%)	Resorbable Sheet, n (%)	Total, n (%)
Diplopia*	0 (0)	1 (25)	1 (10)
Hypoesthesia*	2 (33.3)	1 (25)	3 (30)
Hematoma	0 (0)	1 (25)	1 (10)
Limitation of ocular movement*	1 (16.7)	0 (0)	1 (10)
Enophthalmos*	2 (33.3)	1 (25)	3 (30)
Infection	1 (16.7)	0 (0)	1 (10)
Others	0 (0)	0 (0)	0 (0)
Total	6 (100)	4 (100)	10 (100)

*They are number of aggravated signs and symptoms compared with preoperation.

Table 3. Follow-Up of the Postoperative Signs and Symptoms With Medial Orbital Wall Reconstruction Through Subciliary Approach

Physical Examination	Time	Medpor, n (%)	Resorbable Sheet, n (%)	Total, n (%)
Diplopia	Preoperation	26 (100)	19 (100)	45 (100)
	Immediate operation	12 (46.2)	13 (68.4)	25 (55.6)
	1 mo	7 (26.9)	8 (42.1)	15 (33.3)
	3 mo	3 (11.5)	1 (5.3)	4 (8.9)
	6 mo	3 (11.5)	0 (0)	3 (6.7)
	12 mo	0 (0)	0 (0)	0 (0)
Hypoesthesia	Preoperation	9 (100)	7 (100)	16 (100)
	Immediate operation	5 (55.6)	3 (42.9)	8 (50)
	1 mo	3 (33.3)	2 (28.6)	5 (31.25)
	3 mo	2 (22.2)	3 (42.9)	5 (31.25)
	6 mo	3 (33.3)	2 (28.6)	5 (31.25)
	12 mo	0 (0)	0 (0)	0 (0)
Enophthalmos	Preoperation	4 (100)	8 (100)	12 (100)
	Immediate operation	3 (75)	1 (12.5)	4 (33.3)
	1 mo	3 (75)	0 (0)	3 (25)
	3 mo	0 (0)	1 (12.5)	1 (8.3)
	6 mo	0 (0)	0 (0)	0 (0)
	12 mo	0 (0)	0 (0)	0 (0)

conjunctival scarring.[6] When the inferior and medial walls were addressed, the transcaruncular approach was combined with an inferior conjunctival fornix approach. In this case, inferior oblique muscle has to be cut at its origin and, later, sutured back to the periosteum at its origin or simply repositioned.[7]

We used subciliary incision, and dissection was extended just below the anterior lacrimal crest to increase the exposure. According to our previous article,[8] lacrimal canaliculus is relatively safe in this approach.

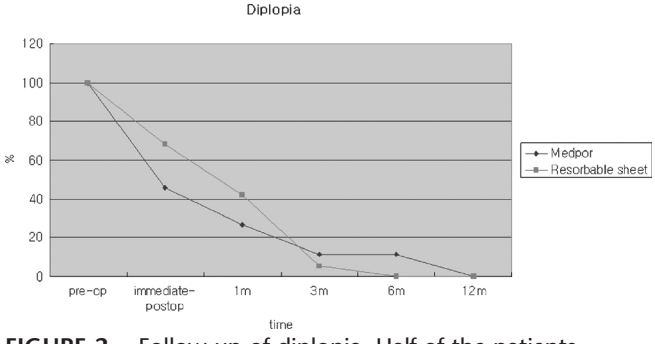

FIGURE 2. Follow-up of diplopia. Half of the patients (3 cases) presenting with diplopia improved during the first month of follow-up, and all of them, both the Medpor and resorbable sheet groups, improved by 6 months.

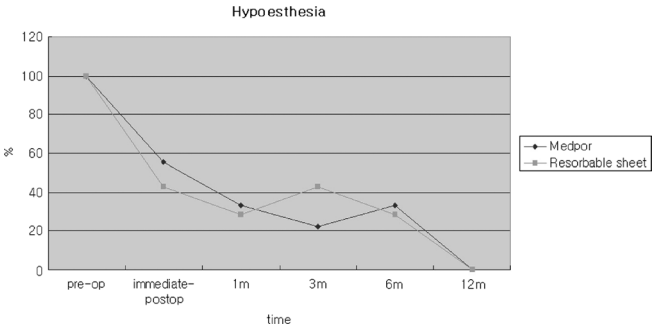

FIGURE 3. Follow-up of hypesthesia. All patients improved by 3 months.

We think that medial orbital wall could be reconstructed safely through skin-muscle flap subciliary approach without resulting in ectropion or lacrimal canaliculus injury.

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Malignant Triton Tumor of the Infratemporal Fossa

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Abstract: Malignant triton tumor is a very aggressive type of sarcoma that comprises rhabdomyoblasts and malignant Schwann cells. It is a different entity from malignant schwannoma, characterized by

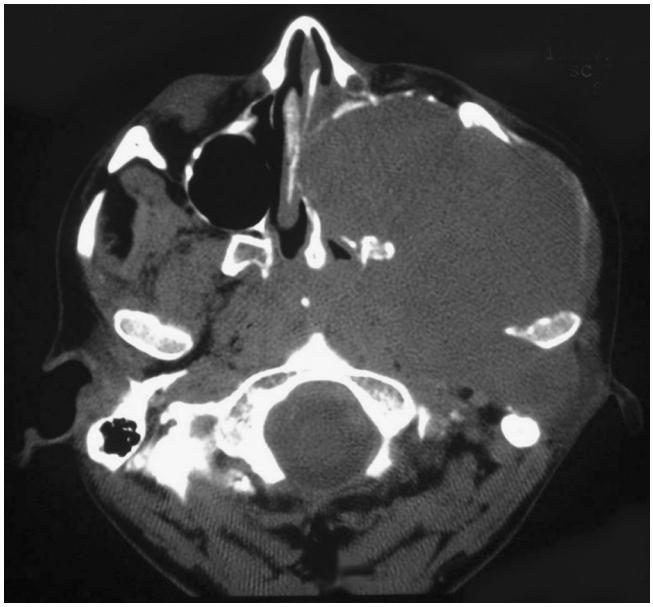

FIGURE 1. Preoperative axial CT scan revealing a large mass that entirely filled the left temporal and infratemporal fossae; left maxillary, ethmoidal, and sphenoidal sinuses; left nasal cavity; left parotid region; and periorbit. The tumor extended to the middle cerebral fossa through the foramen ovale and affected the left petrous apex and the left parasellar region. There was involvement of the mandibular condyle and the left temporomandibular joint dislocation.

their aggressiveness and poor prognosis. Head and neck location is frequent, and early diagnosis and complete resection followed by radiation therapy is important for long-term survival. However, the therapeutic plan should be individualized, taking into account the location and size of the primary tumor. The use of adjuvant chemotherapy and molecular therapies should be considered in the treatment of these tumors. We report an unusual presentation of a malignant triton tumor located in the infratemporal fossa, describing its clinical and pathologic features, and we try to update the knowledge in the management of these tumors, including the use of molecular therapies.

Key Words: Triton tumor, schwannoma, rhabdomyosarcoma, infratemporal fossa

Malignant triton tumor (MTT) is an aggressive soft tissue sarcoma consisting of a malignant schwannoma with rhabdomyoblastic differentiation.[1] Malignant triton tumor was first described by Masson[2] in 1932, and the name *triton tumor* derives from Locatelli's[3] experiments in which a sciatic nerve implanted subcutaneously in the back of a triton salamander caused the growth of supernumerary limbs consisting of neural and muscular tissues.

There are also benign triton tumors, also called neuromuscular hamartomas, composed of benign peripheral nerve fibers mixed with striated muscle fibers.

Malignant triton tumor is a rare entity with fewer than 100 cases reported in the literature, in which slightly more than 25% are developed in the head and neck.[4] It is diagnosed at a relatively young age[5] and shows very aggressive behavior and poor prognosis.[6] An MTT can be sporadic or associated with neurofibromatosis type 1 (NF1).

We present a sporadic MTT located in the infratemporal fossa and arising from the trigeminal nerve, where 3 other cases have already been described.[7–9] After reviewing the literature, this case is the second arising from the trigeminal nerve and not associated with neurofibromatosis. The aim of this study was to describe an unusual case to try to upgrade the management of MTT, emphasizing the use of recent treatments like retinoic acid derivatives, which have been used in few cases so far.

CLINICAL REPORT

A 23-year-old woman presented to our institution in October 2006 with a history of quickly progressive swelling on the left side of her face.

She referred that she had noticed the appearance of the swelling 10 months ago, as well as pain, muscle tension, and difficulty to open the mouth. The history of the patient was otherwise negative.

The patient had a complete left-side nasal obstruction, a slow progressive left hearing loss, a lancinating left periorbital pain, and episodes of dysarthria related to difficulty of mouth opening. She also complained of headache and general syndrome.

Physical examination showed a large painless mass of a hard consistency on the left temporal region that stretched to the zygomatic region and contacted with the jaw's angle. The patient presented anesthesia in the areas innervated by the second and third branches of the trigeminal nerve. The otoscopic exploration showed a protrusion of the lower wall of the left external auditory canal. On anterior rhinoscopy, there was a swelling of the outer wall of the left nostril that caused its complete obliteration. Exploring the oral cavity, there was a swelling of the left palate and a medial protrusion on the left side of the oropharynx. Vision and ocular motility were normal.

An enhanced computed tomographic (CT) scan confirmed the presence of a lytic and expansive mass that destroyed nearly all bones of the left hemiface (Fig. 1). A magnetic resonance image (MRI) was also obtained (Fig. 2).

Abdominal, pelvic, and thoracic CTs and MRIs indicated no evidence of a metastatic disease.

A biopsy specimen under nasal endoscope was obtained and disclosed histologic components of both a rhabdomyosarcoma and a malignant schwannoma. The tumor cell density was high, and the predominant component was constituted by the proliferation of rhabdomyoblasts scattered between fascicles of spindle-shaped cells (Fig. 3). The pleomorphism was moderate, and few mitoses were present. There were no areas of hemorrhage or necrosis. Inmunohistochemistry was uniformly positive for vimentin and *bcl-2* and erratically positive for desmin and common muscle actin. CD99, CD34, CD117, and smooth muscle actin staining were negative. Myo-D1 staining was positive in most cells, and S100 showed a reversed pattern of desmin (Fig. 4) and Myo-D1 staining. KI67 showed an irregular rate of proliferation. These findings confirmed the diagnosis of MTT, according to the criteria of Woodruff et al.[1]

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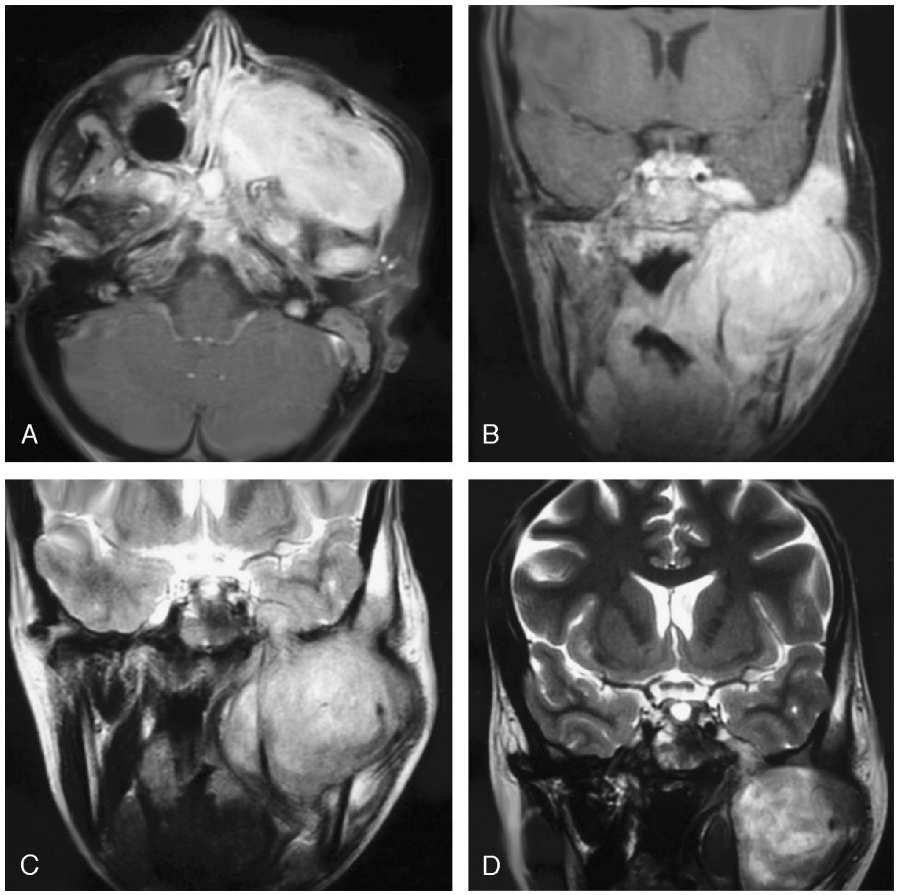

FIGURE 2. Magnetic resonance image. A, T1 (Gd)-weighted MRI demonstrating the large and heterogeneous left infratemporal fossa mass. Lesion occupied the left ethmoid and maxillar sinus, projected into nasal fossa and eroded the bones around. B, T1–weighted MRI showing the involvement of the left parapharyngeal space and the temporal fossa, extending to the skull base. C and D, T2-weighted MRI showing the tumor extension to the middle cerebral fossa through the foramen ovale and the affection of the left parasellar region. The mass did not involve the dura.

She had not received prior radiation therapy, and the family history and physical examination were negative for NF1 and other possible congenital lesions.

The tumor was initially deemed unresectable because of its large size and its extensive facial, orbit, and skull base involvement, so the patient received ifosfamide (2 g/m^2 per day) and adriamicine (60 mg/m^2 per day) for 6 days. The tumor failed to regress except in the parasellar region, where its size was reduced, so it was decided to attempt an excision of the tumor, understanding the potential for incomplete excision and the risk of esthetics sequelae.

The patient underwent resection of the lesion by a subtemporal-preauricular infratemporal fossa approach combined with a left transfacial approach on March 2007, finding a large tumor that occupied the whole of the infratemporal and pterygopalatine fossae, and introduced intracranially by the foramen ovale, infiltrating the dura mater. The tumor destroyed the maxillary and ethmoidal sinuses, the zygomatic process, and the ascending branch of the jaw; infiltrated soft tissues of this area; and also invaded the left orbit through the superior orbital fissure.

The tumor was resected in monobloc with negative margins for residual tumor except in the parasellar region, where tumor persistence could not be excluded. Excision included removal of the zygomatic arch; the vertical branch of the mandible; the maxillar, ethmoid, and lateral nasal walls; the parotid gland, the V2, and the

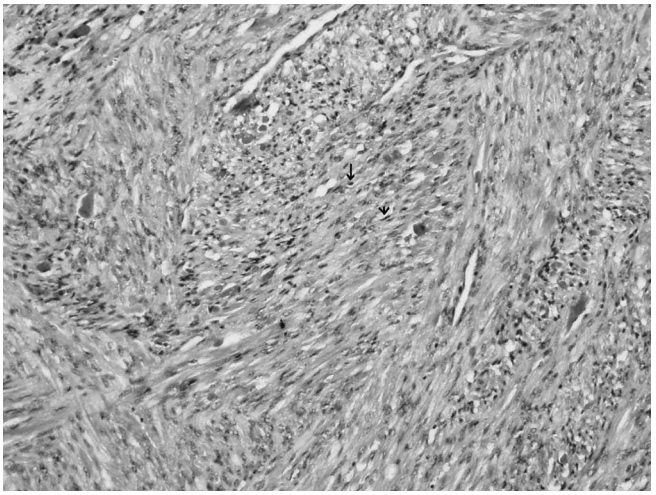

FIGURE 3. Rhabdomyoblasts (round cells having abundant eosinophilic cytoplasm and eccentric nuclei; arrow) are scattered throughout intersecting fascicles of elongated spindle cells (arrow head; hematoxylin-eosin, original magnification ×10).

V3. The resection site was reconstructed with an anterolateral thigh free flap and a titanium mesh for orbital floor reconstruction. The result of the pathologic examination of the piece was believed to represent an MTT.

After surgery, the patient received 66 Gy (220 cGy per fraction) of radiotherapy by the helicoidal tomotherapy system, beginning from the 12th postoperative week. However, during radiotherapy, the patient presented a tumoral growth located in the left parasellar region, cavernous sinus, posterior cranial fossa, around the pons, and along the path of the trigeminal nerve (Fig. 5).

After treatment, the tumor seemed to be locally controlled, but on January 2008, 6 months after radiotherapy, the patient developed a local progression in the posterior cranial fossa, skull base, and left parotid region and several metastatic lesions in the lungs, ribs, sacrum, and pelvis. Because of this progression, the patient presented with frequent headaches, dyspnea, walking difficulty, behavioral changes, and eye problems.

Additional chemotherapy was considered on February 2008, and the patient received a cycle of Dacarbazin (1.2 g/m^2) that was not well tolerated. The clinical situation worsened, and it was decided to apply 30 Gy of radiotherapy (10 fractions), with palliative intent to the pelvic and thoracic regions, but it had to be suspended before the end of the treatment on April 2008 by a general deterioration.

At the same time, it was decided to analyze the possible presence of retinoic acid receptors (RARs) in the tumor specimen because of their existence in the previously reported cases[10] of tumors that responded favorably to treatment with interferon alfa and isotretinoin. We determined the receptors and retinoid X receptor (RXR) and the amplification was positive, so the treatment with interferon alfa (10 million units subcutaneus, 3 times per week) and isotretinoin (1 mg/kg per day) was applied, showing good tolerance but with low efficiency.

On May 2008, the patient died of her disease because of intracranial progression.

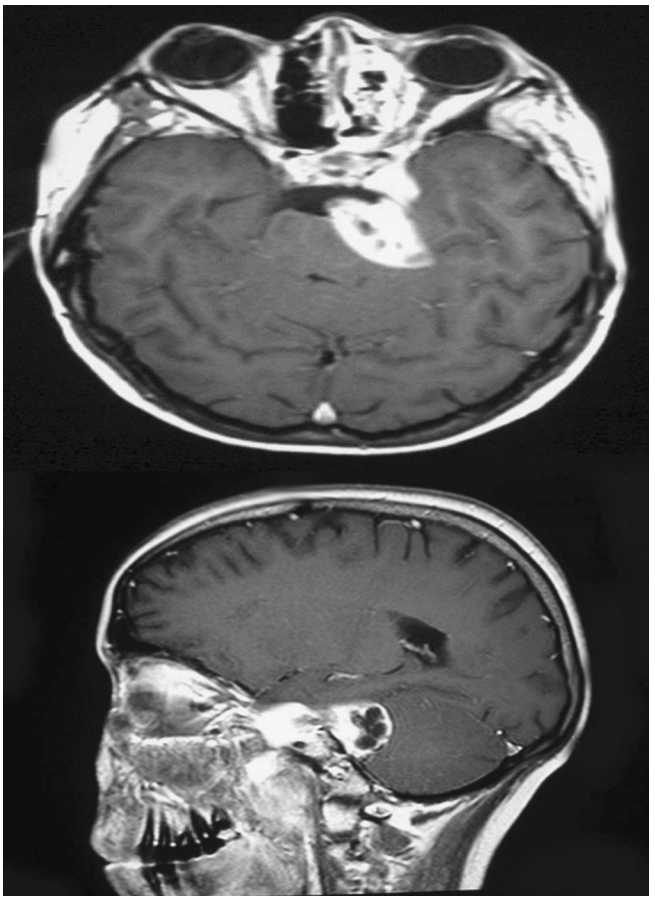

FIGURE 5. Follow-up MRI demonstrating tumor progression to the middle and posterior fossae.

DISCUSSION

Malignant triton tumor is a rare subtype of malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation. Because it is an uncommon entity, the knowledge of the optimal management of these patients is limited and no general suggestions can be made regarding the best treatment and follow-up of these tumors.

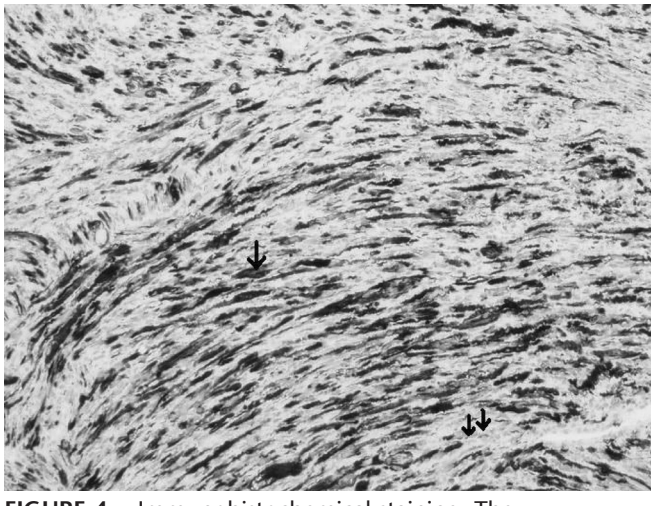

FIGURE 4. Immunohistochemical staining. The rhabdomyoblastlike cells (arrow) showing positive cytoplasmic reaction for desmin and the spindle cells (double arrow) demonstrating the immunoreactivity for S100.

Malignant triton tumors are highly malignant sarcomas that are locally infiltrating and often result in multiple recurrences. Finally, there is a metastatic spread.

This tumor usually arises sporadically, but it sometimes occurs associated with NF1 (33% of cases[8]). Radiation had been implicated as an etiologic factor in the development of MTT, with a latency period of 10 to 20 years.

Fewer than 100 cases have been reported, and head and neck region is one of the most frequent sites of involvement. The most common site of involvement is the neck (33%), arising from the branches of the brachial and the cervical plexuses and the vagus nerve branches. However, other places have been described.

The main presenting symptom is an enlarging mass that invades and destroys slowly surrounding structures. In most cases, these tumors are diagnosed in advanced stages in which structures such as the orbit or the skull base are affected and, sometimes, intracranial invasion exists. In our patient, the mass was very large, and there was extensive destruction of the facial skeleton and intracranial invasion. Advanced stages will affect the treatment and prognosis of these patients. At the time of diagnosis, these patients usually do not have regional or distant spread of the disease.

Performing an early diagnosis is important because the tumor size is the most important parameter associated with survival. A biopsy of the lesion will establish the diagnosis. Woodruff et al[1] established 3 histopathologic criteria for the diagnosis of MTT.

The typical pathologic finding is a proliferation of spindle cells with a fibrillar, elongated, eosinophilic cytoplasm and prominent bipolar extensions, similar to Schwann cells. These cells are mixed with others that have striations and are similar to immature striated muscle fibers. Immunohistochemical staining helps to prove the origin of the cells. In our case, we used antibodies to vimentin, desmin, myosin, and common muscle actin to confirm the skeletal muscle origin, and antibodies to S100 were used to confirm the nerve sheath differentiation. The different rates of cell proliferation, estimated by Ki-67 staining, indicate the heterogeneity of the tumor.

Before treatment, it is necessary to perform a CT scan or an MRI to determine the extent of the disease. Magnetic resonance imaging is the imaging modality of choice to evaluate this type of soft tissue tumor.[11] However, the CT is more useful to detect bone involvement.

As mentioned, most MTT are diagnosed in advanced stages and the choice of treatment modality should be individualized. As with any other sarcoma, resection of the tumor with a wide margin followed by radiotherapy is generally recommended.[12] However, sometimes, the tumor invades unresectable regions, such as the cavernous sinus, and it is necessary to use other therapeutic modalities, such as radiosurgery.

Radiotherapy should always be used after surgical resection to remove any remaining tumor. In our case, radiotherapy was applied in the form of tomotherapy, which is a system of image CT-guided intensity-modulated radiation therapy. Through this modality, radiation can be applied with high precision near vital structures.

The role of chemotherapy has not been clearly defined.[8] In cases similar to ours, which had a large tumor size, adjuvant chemotherapy can be used to try to reduce the tumor size and therefore facilitates surgery.[13]

Recently, Köstler et al[10] published a report on a patient with an MTT, which showed a high expression of RARs. Our patient's tumor had amplification of RAR, and the treatment was administered. However, because this was done in a very advanced stage of the disease, the response was low and not measurable. We believe that this treatment could be used in early stages of the disease because it is well tolerated and has little adverse effects.

These patients have poor prognoses. Some authors[14,15] reported that patients with MMT have the same survival as those with usual malignant schwannoma, but others[5] describe that these tumors are more aggressive. Forty percent of the cases have several local recurrences, which become unresectable, and 48% of patients develop distant metastases.[5] The mean 2-year survival rate for MTT is between 25% to 33% in all regions,[4,5,8,9] but in some locations, such as paranasal sinuses, this rate is higher.[16,17] Survival of patients with head and neck MTT ranges between 4 months and 22 years.[5] Our patient survived 19 months.

In conclusion, we present an MTT of the infratemporal fossa. In this special location, the prognosis is worse because of the difficulty of early diagnosis and complete resection of the tumor. These tumors should be treated at centers with expertise in skull base surgery to achieve complete surgical resections and with experience in reconstructive surgery to repair the esthetic and functional defects. Early detection and aggressive treatment are recommended to try to control disease progress. New treatments, such as retinoic acid derivatives, should be used to verify its usefulness.

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Orbital Fracture Deterioration After Scuba Diving

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Abstract: Sinus barotrauma is a common disease in divers. However, it is not familiar to maxillofacial surgeon. We presented orbital fracture deterioration by sinus barotrauma in scuba diving and a review of literatures. We also discussed the clinical features, the prevention, and the possible mechanism of orbital fracture deterioration after scuba diving.

Key Words: Orbital fracture, scuba diving, sinus barotrauma

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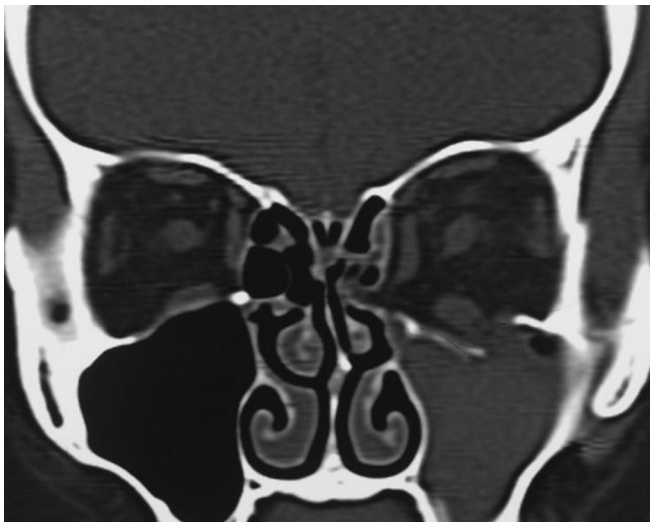

FIGURE 1. Coronal CT scan after injury shows fractures of the orbital floor and medial wall with minimum bone displacement.

A 28-year-old woman was hit on her left eye by another person's elbow. She was diagnosed with fractures of the orbital floor and medial wall with minimum bone displacement on computed tomographic (CT) scan (Fig. 1). She did not show any disturbance of her left eye movement, and her eye examination was normal. Because she did not show enophthalmos 2 weeks after injury, she was treated conservatively. She scuba dived on the 16th day after injury. She had epistaxis when she came up to the water surface from approximately a 15-m depth. Then she noticed gradually deteriorating diplopia and left enophthalmos a few weeks after scuba diving. She reconsulted our hospital 3 months after scuba diving. She showed prominent left enophthalmos and restriction of upward movement of her left eye. Computed tomographic scan revealed a hammock-shaped bone displacement of orbital floor and medial wall and the ipsilateral maxillary sinusitis (Figs. 2A, B). Reconstruction of the orbital floor was performed under general anesthesia at 4 months after scuba diving. We tried to reduce the displaced orbital floor by subciliary and gingivobuccal transmaxillary approach. We identified purulent fluid collection and edematous mucosa in the maxillary sinus. We performed split thickness calvarial bone graft (3 × 3.5 cm) from her parietal region to the orbit floor. Her enophthalmos and disturbance of her left eye movement improved a few weeks after surgery. In 3 months after surgery, she felt dull pain on her left cheek. Computed tomographic scan showed recurrence of left maxillary sinusitis. This condition responded well to medication.

DISCUSSION

In recent years, scuba diving had become a popular sport. However, sinus barotrauma in scuba diving is not familiar to maxillofacial surgeons. Orbital fracture without symptom is generally treated conservatively. Surgeons should know the effect of scuba diving on orbital fractures.

Orbital fracture deteriorated after scuba diving in our case. Because of the hammock shape of the bone fragment, it is reasonable to think that negative pressure in the paranasal sinus pulled the bone fragment into the sinus. If the sinus orifice remains patent, pressure change, which occurs in nasal cavities during scuba diving, is directly transmitted to the sinus cavity. Therefore, in our case, we presumed that edema of the mucous membrane after the original fracture narrowed the orifice of the maxillary and ethmoid sinus and made pressure control between the nasal cavity and paranasal sinus difficult. Intraoperative findings showed purulent fluid collection and edematous mucosa in the maxillary sinus. The patient did not have any history of disorder. We concluded that the cause of deterioration of orbital fracture in this patient is sinus barotrauma squeezing.

Sinus barotrauma is a common disease in divers, following middle ear barotrauma.

Sinus barotraumas occurs because the rigid walls of the cavity cannot expand and contract under conditions of changing pressure. Boyle gas law relates pressure to volume change. There are 3 mechanisms of sinus barotraumas: squeezing, reverse squeezing, and mixed. They have been described in detail by Paul et al.[1] In our case, during descent, an increase of external pressure is transmitted through the sinus wall, leading to vascular congestion and edema in the sinus mucosa. When the elastic limit of the mucosa is exceeded, hemorrhage occurs, either into the mucosa and submucosal space or into the sinus. At the same time, the air in the sinus, in accordance with Boyle law, decreases in volume. If the sinus fills with blood, mucosa, and transudate, during ascent, the air present in the sinus will expand and be at relatively higher pressure than the ambient pressure. Therefore, epistaxis may be expelled during ascent.[1] In our case, when the patient ascended to the surface of water, she noticed only epistaxis without any other symptoms.

In a prior study, it was shown that when the pressure in the paranasal sinus cavities and outer body reached 100 to 150 mm Hg, the paranasal sinus mucosa became edematous or a serous nasal discharge occurred. When the pressure reached 260 to 300 mm Hg, the mucosa bled.[2] She dived to a 15-m depth. The pressure

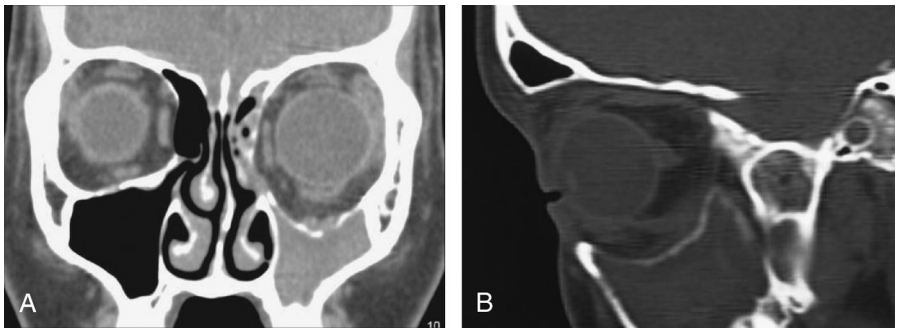

FIGURE 2. A, Preoperative coronal CT scan shows hammock-shaped bone displacement of the orbital floor and medial wall and ipsilateral maxillary sinusitis. B, Preoperative sagittal CT scan shows hammock-shaped orbital wall deformity.

difference between the water surface and a depth of 15 m is approximately 1140 mm Hg. If pressure is not adjusted in the sinus, much negative pressure occurs, which causes bleeding.

Becker et al suggest that depending on the extent of barotrauma, the diver may usually return to diving within 6 weeks if x-ray films show that the sinuses have cleared and if any underlying predisposing conditions have been remedied (i.e., by treatment of coexistent infection, allergy, septal deviation, or polyps). They advise divers that nasal congestion from any cause (e.g., upper respiratory infection, sinus infection, allergy, or smoking) will predispose them to barotraumas.[3] They suggest the following guideline. Diving should not be done if nasal congestion is present, during, for example, an upper respiratory infection or allergic or nonallergic rhinitis. Intranasal pathology, for example, nasal polyps or septal deviation compromising the ostiomeatal complex, may require correction.[4] Lawrence[5] reported that the pathology and symptoms produced by dysbarism are dependent on the degree of pressure differential and the time over which it occurs. Divers should descend and ascend slowly, equalizing continuously. According to these reports, we should permit patients who had orbital fracture to scuba dive after confirmation of normal sinus drainage and no mucous edema on CT scan. That is the prevention of orbital fracture deterioration after scuba diving, and that usually requires 4 to 8 weeks after orbital fracture.

Three months after operation, CT scan revealed the recurrence of left maxillary sinusitis in our case. There were no other nasal disorders. We assumed that the orifice of maxillary sinus was constricted by fracture at scuba diving, and the constricted orifice was not reduced completely by operation.

SUMMARY

In this report, we presented orbital fracture deterioration and its mechanism after scuba diving. To our knowledge, this is the first report about fracture by squeezing. Scuba diving is a popular sport, and therefore, it is important to note that scuba diving can worsen orbital fractures. After an orbital fracture, evaluation of the paranasal sinus on CT scan should be performed before scuba diving is undertaken. Divers should descend and ascend slowly, equalizing continuously for prevention of orbital fracture deterioration.

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Aplasia Cutis Congenita: Management of a Large Skull Defect With Acrania

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Abstract: Aplasia cutis congenita is a rare disorder characterized by absence of skin. Lesions typically occur on the vertex and are sometimes small, but they can affect deep tissues such as the skull bone and dura. Mortality is related to the depth and size of the lesion and can amount to a rate of more than 50% when full thickness is involved. The treatment remains controversial—both surgical and conservative managements are described. Minor lesions can be controlled with nonsurgical treatment, but large defects require early surgery. We report the case of a female newborn with acrania and scalp aplasia cutis congenita, which was treated with a bipedicle scalp flap based on the temporal vessels. Full- and partial-thickness skin grafts were used to cover the donor site on the temporo-occipital region. Postoperatively, the patient developed a liquorice cyst, which was treated with a shunt, and she has been followed up for evaluation of the bony defect closure and skull morphology. Her neuropsychomotor development is normal.

Key Words: Aplasia cutis congenita, acrania, large skull and scalp defect, congenital deformity

Aplasia cutis congenita (ACC) is a rare condition characterized by noninflammatory, well-demarcated defects of all skin layers and subcutaneous tissue, also affecting, although less often, deep tissues such as the periosteum, the skull bone, and the dura. It is an ubiquitous disease, which was first described in the extremities by Cordon, in 1767,[1] and in the scalp, by Campbell, in 1826.[2]

In the head, the lesions typically occur on the vertex, where they may be as small as pinpoints, but they may also affect significant areas of the skull.[3–13] Rarely does ACC involve the deep tissues, but it if does, scalp, skull, and dura may be absent at birth.[3,9,13] These structures are being usually covered by a thin glistening membrane that eventually ulcerates.[8]

The etiology of ACC is unknown, but genetics certainly does play a role therein, as do vascular accidents, amniotic adhesions, thrombotic events, teratogens, and intrauterine pressure and infection.[4,6,9,12,14] Aplasia cutis congenita may be present in a isolated condition or be associated with Adams-Oliver syndrome, a rare congenital condition that is usually sporadic but can be transmitted in a autosomal dominant mode of inheritance.[4,7,12,15]

The involvement of the dura, bone, and scalp entails the risk of infection, meningitis, venous thrombosis, and sagittal sinus hemorrhage.[3,5,9,10,13,15,16] Depending on the size and depth of the defect, the mortality rate, in such cases, can increase to more than 50%.

The management of large skull defects resulting from cutis aplasia remains controversial. Both surgical intervention and conservative treatment have been described in the literature.[3,4,7,14,17] Minor scalp lesions can be treated conservatively. However, large

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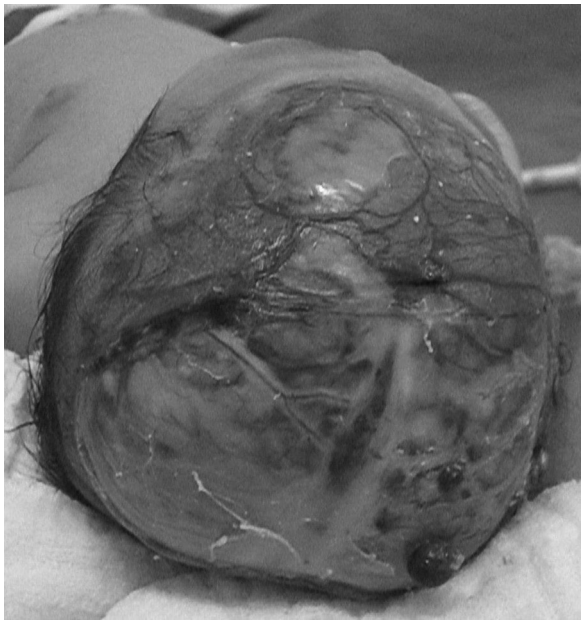

FIGURE 1. Superior view of the scalp defect (2 d of life).

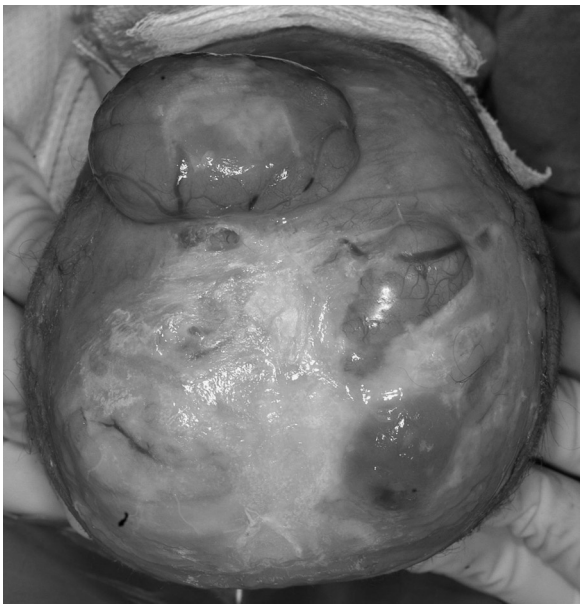

FIGURE 3. Frontal encephalocele (14 d of life).

defects require surgery and preferably the use of rotation scalp flaps.[4,17]

CLINICAL REPORT

The female patient had been born by cesarean delivery and transferred from another hospital. The mother was 31 years old (gravida 4, para 3). There was no abnormality in the family history. The newborn infant weighed 3255 g and received Apgar scores of 8 and 9. The skin defect was noted at birth: a large lesion measuring 14 × 12 cm, involving the full thickness of the cranium. The dura mater was also missing, with only a thin glistening membrane overlying the encephalon (Fig. 1). The large, grossly dilated superficial veins were a possible site for a potentially fatal hemorrhage. There was no immediate leak of cerebrospinal fluid (CSF). A computed tomographic scan revealed normal intracranial structures, but the overlying bone was missing (Fig. 2). There was no other malformation, and nothing remarkable was detected on neurologic examination.

Initially, the lesion was dressed once a day with topical collagenase and chloramphenicol gauze to keep the wound moist and aseptic. Intravenous antibiotics (oxacillin and cefotaxime) and phenobarbital were administered to prevent infection and seizures.

On day 14, probably in consequence of a hypertensive crisis, an encephalocele developed through a tear in the fragile membrane, and the patient started fever episodes (Fig. 3). Although a CSF examination result did not reveal meningitis, the antibiotics were

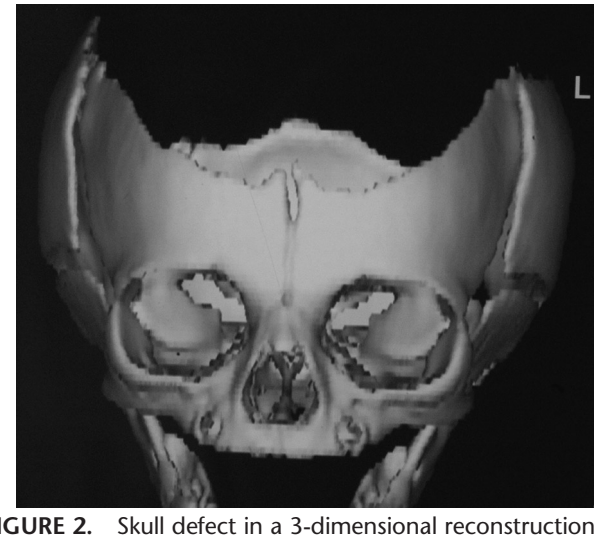

FIGURE 2. Skull defect in a 3-dimensional reconstruction computed tomographic scan.

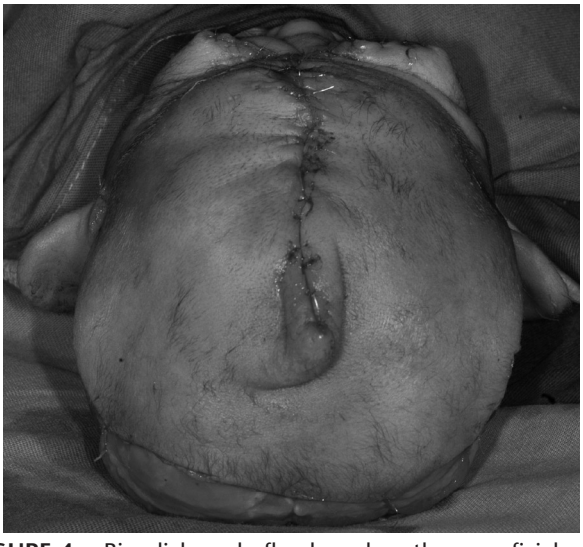

FIGURE 4. Bipedicle scalp flap based on the superficial temporal vessels (superior view).

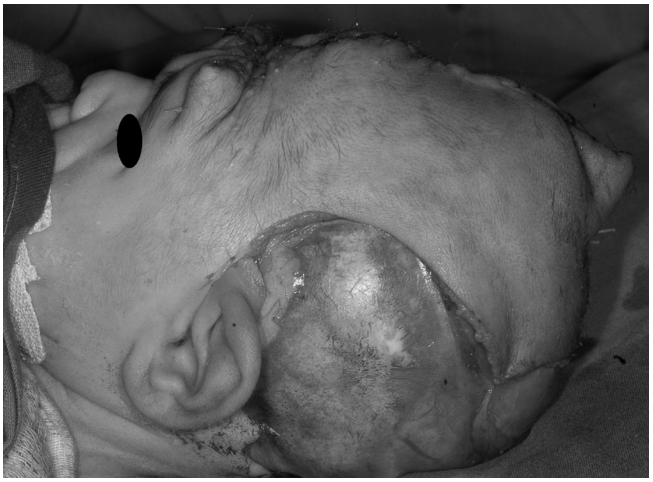

FIGURE 5. Bipedicle scalp flap based on the superficial temporal vessels (lateral view).

changed to cefepime and vancomycin, and early surgery was planned.

On day 20, in the operating theater, the neurosurgical team placed a patch of lyophilized dura mater (Duraform Johnson & Johnson, Raynham, MA) on the left half of the defect to repair the encephalocele. A huge bipedicle flap of skin and galea, based on the right and left superficial temporal vessels, was made to cover the defect (Figs. 4 and 5). The donor site on the temporo-occipital region was left to granulate and be grafted in a posterior surgery. On the second postoperative day, the infant presented a CSF leak through the vertex of the flap, which healed after a lumbo-peritoneal deviation procedure. Thirty days after the first operation, the donor area was covered with full- and partial-thickness skin grafts, and the healing was uneventful.

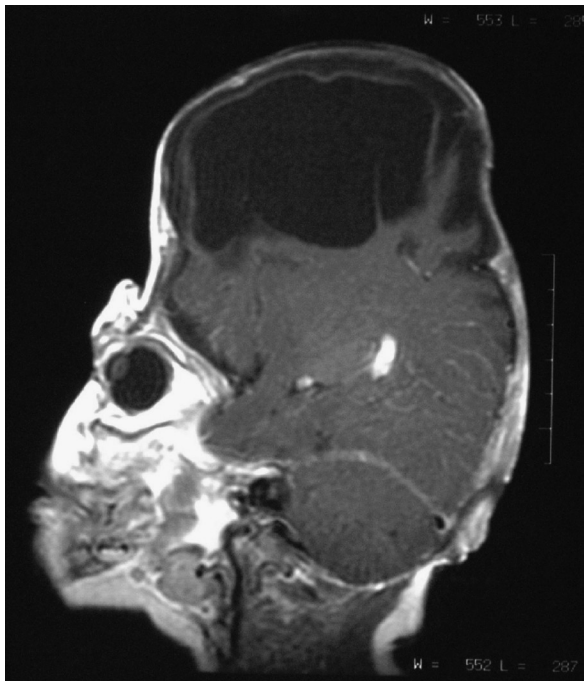

FIGURE 6. Liquorice cyst on magnetic resonance imaging.

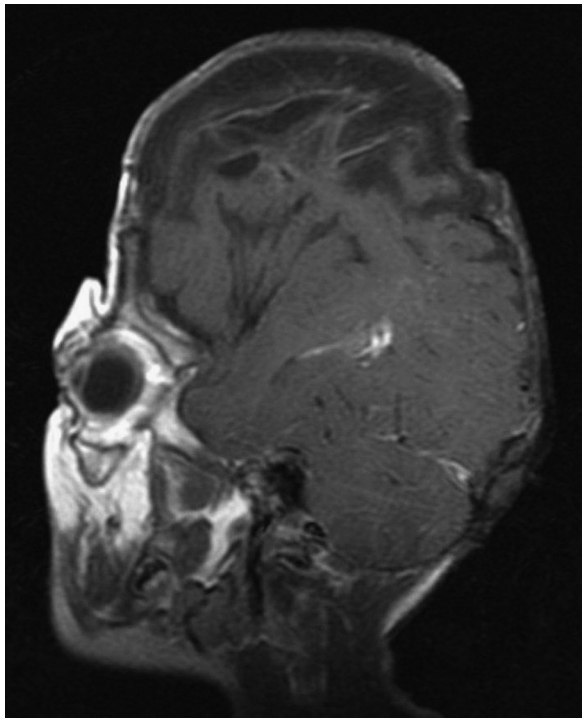

FIGURE 7. Magnetic resonance image at 8 months after shunt.

During the first year of follow-up, the patient developed an oxycephalic aspect owing to a liquorice cyst that was treated by the neurosurgery team 1 year after the first procedure (Figs. 6 and 7).

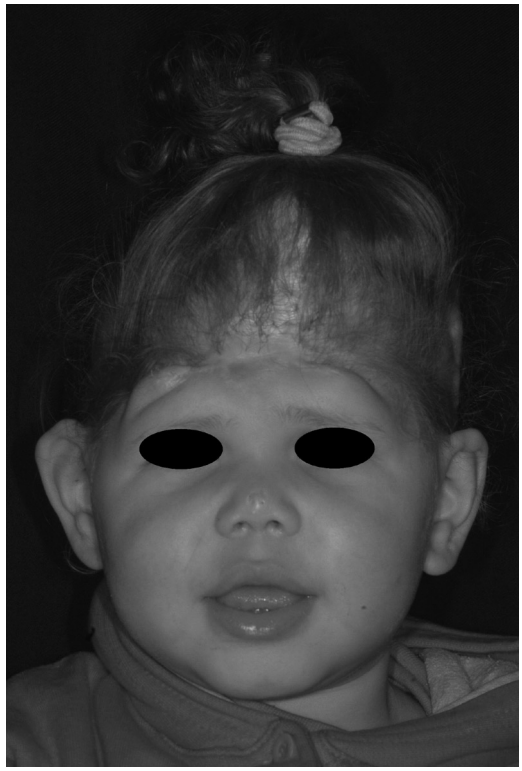

FIGURE 8. Frontal view at 21-month follow-up.

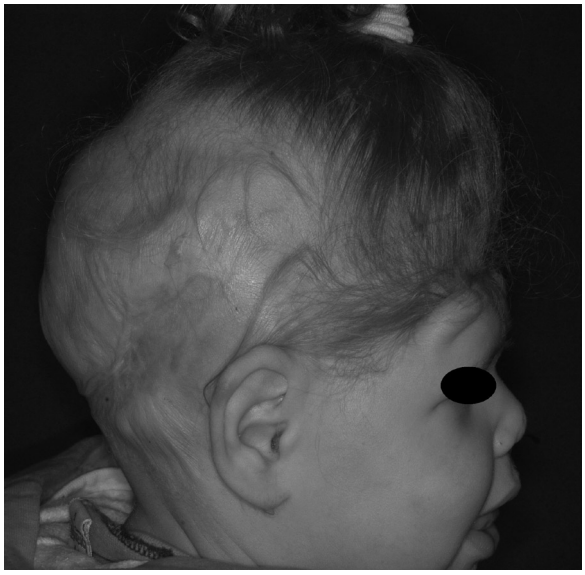

FIGURE 9. Lateral view at 21-month follow-up.

Currently, 21 months postoperatively, she is being followed up for closure of the bony defect and evaluation of skull morphology. Her neuropsychomotor development is completely normal (Figs. 8 and 9).

DISCUSSION

The involvement of the skin, skull, and dura by ACC is a rare condition, and the female-male incidence ratio is approximately 7:5.[5] The extensive defects represent a challenge to the treatment approach, as the etiology remains uncertain and management is still controversial. The goal is to achieve complete closure of the defect, avoiding important risks such as meningitis, hemorrhages, and brain trauma.

Conservative management has been advocated by several authors.[7,12,14,15] Treatments consisting of a variety of saline, silver sulfadiazine or bacitracin cream dressings, special wound-dressing materials (Acticoat, Smith & Nephew, St. Petersburg, FL; Elasto-Gel Southwest Technologies, Kansas City, MO), and engineered skin procedures. The reason for such an approach is to avoid operations and risks associated therewith for a newborn. There seems to be a consensus that conservative treatment with local wet and antibiotic dressings is the therapy of choice for patients with small defects.[8,18]

However, some reports of deaths after massive hemorrhage in patients with ACC who were treated conservatively have highlighted the problem of operative management.[18,19] In the case being reported, our patient developed an encephalocele during the initial conservative management, and 2 encephaloceles that were developed with that same treatment had been described by Ribuffo et al.

Patients with large skull defects should, therefore, be considered for surgical treatment because of the risk of infection or early eschar formation leading to massive bleeding.[3–6,8,10,11,16–20] Surgical treatment options include partial- or full-thickness skin graft, scalp rotation flaps, pericranial flaps, split rib grafts with free flaps, and tissue expansion.[5] The skin grafts entail a risk of partial or total failure, and hemorrhage can occur during future replacement with skin flaps.[9] Periosteal flaps covered with skin grafts have been reported by Moscona et al,[17] with the goal of creating a viable site for bony regeneration. The free flaps such as the latissimus dorsi are indicated for future cranioplasty in older infants or when a local flap has failed.[3] The extreme rarity of these cases and small series reported make it difficult to evaluate what is the best surgical repair. We have not found any reports describing an ACC skull defect as large as our patient's, and we believe that the best approach is early surgery and placement of rotation skin flaps, if necessary combined with split skin grafts to cover the donor area.

Patients with ACC of the scalp have an abnormal arterial architecture, which allows the blood to flow between the left and right sides, the anterior and posterior regions of the scalp. However, the site of superficial arteries can be displaced, and a previous Doppler study should be done to determine a safe pedicle. In our case, a bipedicle flap was planned, based on superficial temporal vessels, to avoid possible partial necrosis reported by other authors[4,12]; as bipedicle flaps seem to have better viability than single flaps.[5,9]

When dealing with a tear of the fragile tissue, or a laceration of the dura mater, the best option, in our opinion, is complete repair of the dura and not of only a part of it. In our case, had we done that, we should probably have avoided the CSF leakage. The weakness at the bony defect allows the growing brain to exert an expanding force on the local flaps.[3] The oxycephalic aspect of the face generally improves with time and does not require operation. When this appearance is because of liquorice cyst, however, treatment is necessary. The best examination for diagnosis and follow-up is magnetic resonance imaging. Reconstruction cranioplasties with rib grafts or with cranial vault splitting are advised when the patient is 3 or 4 years old.[9,21]

What we have presented here is our experience in treating a rare and extensive lesion of ACC. For most patients, the conservative local treatment will be sufficient. For large defects, when complications are to be expected, we believe an early and well-planned surgery should be performed. Bipedicle flaps are safe and represent a good option without excessive morbidity. Consultation with neurosurgery and plastic surgery teams is of critical importance in the treatment of patients with ACC.

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Huge Arteriovenous Malformation in Masseter Muscle

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Abstract: Arteriovenous malformation is a tumor characterized by direct connection between an artery and vein without capillaries in-between, and it is commonly located intracranially. Intramuscular arteriovenous malformations are rare in the head and neck region. Less than 1% of the vascular tumors are localized in a muscle, 15% of them are in the head and neck muscles. Among the head and neck muscles, masseter muscle is the most common location, with the rate of 4.9%. The condition of a 36-year-old patient who applied to our clinic with the complaints of progressively increasing pain and progressively growing mass in the right cheek that appeared 1.5 years ago was diagnosed as arteriovenous malformation located in the masseter muscle. After preoperative embolization, the mass was successfully treated with total excision. In this case report, diagnostic and therapeutic tools addressing arteriovenous malformation located in the masseter muscle are discussed in the light of current literature.

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Key Words: Arteriovenous malformation, vascular tumors, intramuscular arteriovenous malformations, masseter muscle, treatment of arteriovenous malformation

CLINICAL REPORT

A 36-year-old patient was admitted to our clinic with the complaints of progressively increasing pain and progressive swelling in the right cheek that first appeared 1.5 years ago. On physical examination, a painless, mobile, pulsatile mass that is rubberlike in consistency, measuring 6 × 6 cm, was palpated (Fig. 1).

Cranial magnetic resonance imaging (MRI) and angiography were performed. In the cranial MRI, an expansile lesion with distinct borders measuring 6 × 5 × 4 cm that was located in the subcutaneous region showing serpiginous flow void vascular structures consistent with arteriovenous malformation was detected (Fig. 2). In cerebral angiography, the mass was reported to be supplied by right facial artery and branches of internal maxillary artery and to drain to the facial and external jugular veins in a mixed fashion (Fig. 3).

Preoperative embolization of the mass was performed. Under general anesthesia, after modified Blair incision, the trunk and all of its branches were isolated and dissected from the tumor. Facial artery and vein that was supplying the mass inferiorly were both ligated. In the proximity of the mass, multiple connections between arteries and veins were noted (Fig. 4). The mass infiltrated the masseter muscle. The mass was excised together with the masseter muscle after dissecting from the parotid tissue. Histological

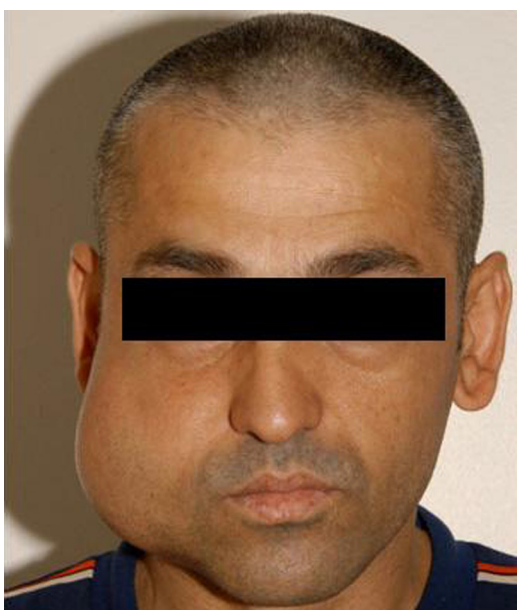

FIGURE 1. Mobile, pulsatile mass that is rubberlike in consistency, measuring 6 × 6 cm in the right buccal region, is seen.

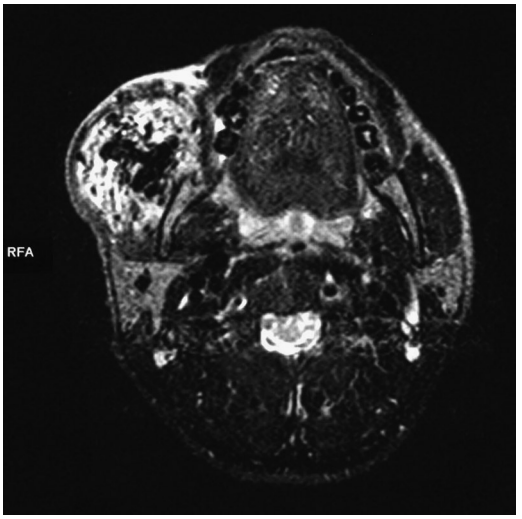

FIGURE 2. Cranial MRI showing an expansile lesion, with distinct borders measuring 6 × 5 × 4 cm, that is located in the subcutaneous region with serpiginous flow void vascular structures consistent with arteriovenous malformation.

examination findings of the removed tissue showed arteriovenous type angiomatous formation and fresh thrombus in lumen of some vessels (Fig. 5).

No residual lesion was detected during follow-ups neither in the physical examination at 3 monthly intervals nor in the MRI that was performed 1 year after the operation (Fig. 6).

DISCUSSION

Arteriovenous malformation is a direct connection between an artery and vein without capillaries in-between. Vascular malformations are commonly detected at birth or in early childhood. However, some may manifest in the adolescent and early adult period.[1,2] Arteriovenous malformations are commonly located intracranially. Although skeletal muscle vascular tumors are very

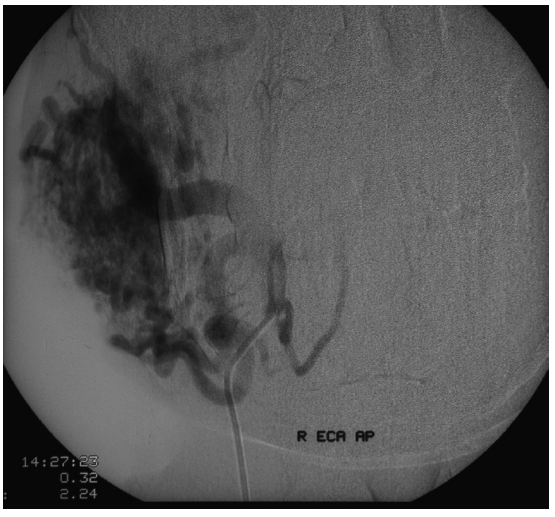

FIGURE 3. Cerebral angiogram shows that the tumor was supplied by the right facial artery and branches of the internal maxillary artery.

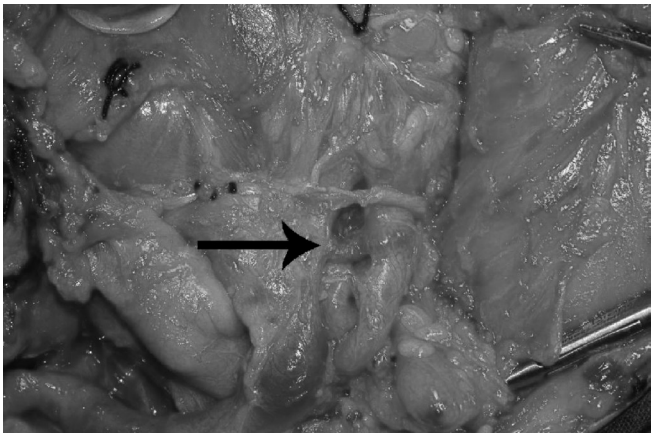

FIGURE 4. Intraoperative photo showing connection between arteries and veins around the tumor.

rare (1% of all vascular tumors[3]), they are commonly found in the head and neck region (15%). Among the muscles affected, the most common location is the masseter muscle (4.9% of all intramuscular malformations) followed by the sternocleidomastoid and trapezius muscles.[4]

The causes of vascular tumors are unknown. Placental cells, such as the trophoblast, may be the cell of origin for vascular malformations.[5]

Intramuscular vascular malformations present with swelling, and pain may accompany. They become more evident with muscle contraction.[1] These lesions may be described as rubberlike in consistency, tense, pulsatile, and mobile in the vertical axis of the muscle. They are commonly deeply seated and therefore are difficult to be recognized with physical examination alone.[3] Skin discoloration due to vascular abnormality is rare. In contrast to the vascular lesions of the skin, they do not grow with time.[6,7]

Vascular malformations of the masseter muscle present with preauricular or buccal swelling with or without pain. Facial paralysis may be seen if expansion of the mass results in pressure on the nerve.[8,9] Lesion becomes more prominent with Valsalva maneuver or contraction of the masseter muscle.[3]

Ultrasound can diagnose these lesions but may not be able to delineate their extent; computed tomography/MRI is more useful.

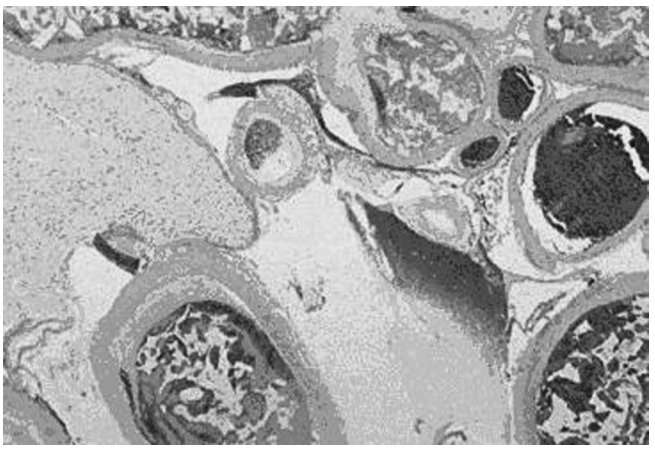

FIGURE 5. Arteriovenous type angiomatous formation and fresh thrombus in lumen of some vessels. Original magnification ×100.

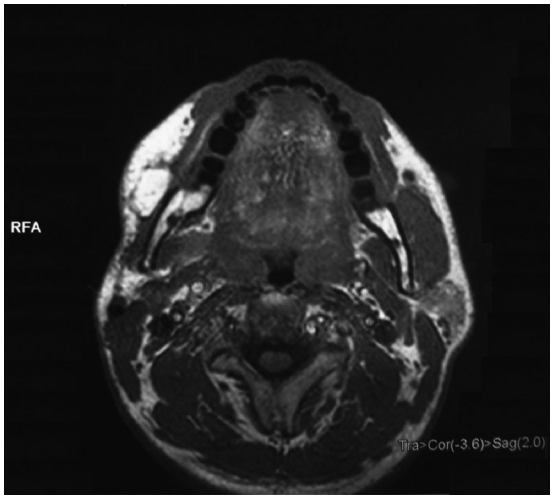

FIGURE 6. Follow-up MRI at the first year showing no recurrence.

Management of intramuscular vascular malformation should be individualized based on tumor location, accessibility, depth of invasion, patient age, and cosmetic considerations. Low-flow vascular malformations can be managed well with medical therapy (high-dose steroid), sclerotherapy, laser therapy, or cryosurgery. High-flow lesions require embolization followed by surgical therapy.[10]

Preauricular incision extended to upper cervical skin line provides sufficient exposition in most cases. Superficial parotidectomy and retraction of the facial nerve enable the surgeon to reach the masseter muscle and work in a more bloodless field. In masseter masses close to the oral mucosa, intraoral approach may be used. However, this approach is not advised because it may cause facial nerve damage and limited bleeding control.[11] Recurrences are commonly seen after intraoral excision. The drawback of extended parotidectomy is facial asymmetry. This asymmetry can be decreased with superior reflection of sternocleidomastoid muscle as a flap. Despite the characteristic appearance, the diagnosis is made through histopathologic examination.

Complications from the disease process include hemorrhage, infection, function problems, and ulceration. Treatment complications include recurrence and surgical complications (hemorrhage, airway compromise, hematoma, and skin necrosis).

Rare vascular malformation cases located in the masseter muscle are present. However, in the case we presented, lesion manifested in the adult period and increased in size. Therefore, in intramuscular tumors showing progressive growth in adults, vascular malformations should be considered.

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Management of Paragangliomas in Otolaryngology Practice: Review of a 7-Year Experience

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Background and Aims: Paragangliomas of the head and neck are highly vascular lesions originating from paraganglionic tissue located at the carotid bifurcation (carotid body tumors), along the vagus nerve (vagal paragangliomas), and in the jugular fossa and tympanic cavity (jugulotympanic paragangliomas) and should be considered in the evaluation of all lateral neck masses. The aim of this study is to review an institutional experience in the management of these tumors.

Materials and Methods: Twenty-six patients with 27 paragangliomas were treated in our institution during a period of 7 years (2000–2007). There were 15 women (57.6%) and 11 men (42.4%) with a mean age of 33.5 years. A painless lateral neck mass was the main finding in 16 patients (61.5%). There was no evidence of a functional tumor. Carotid angiography was performed on all of our patients (100%) to define the vascular anatomy of the lesion. Twenty-two paragangliomas (of the 25 operated paragangliomas; 88%) underwent selective embolization of the major feeding arteries. We performed surgery on 24 (92.3%) patients. Two patients were treated with radiotherapy.

Results: Most lesions were paragangliomas of the carotid bifurcation (n = 14 [51.8%]), whereas 6 patients were diagnosed with jugular (22.2%), 1 with a vagal (3.7%), 1 with a tympanic paraganglioma (3.7%), 2 with jugulotympanic paraganglioma (7.4%), and 1 with laryngeal paraganglioma (3.7%). In 1 patient (3.8%), bilateral paragangliomas in the carotid bifurcation were detected.

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There was an evidence of malignancy in all cases (3.8%). Preoperative embolization has proven successful in reducing tumor vascularity in approximately 22 (of 25 who accepted surgery; 88%) paraganglioma patients. The common preoperative complication was vascular injury, which occurred in 6 (23%) of 26 patients; the main postoperative complication was transient cranial nerve deficit in 4 (15.3%) of 26 patients; and a permanent Horner syndrome was documented in 2 patients (7.6%). Cerebrospinal fluid leak occurred in 1 patient (3.7%). Postoperatively, stroke was occurred in 1 patient (3.7%). Two patients with jugular paraganglioma were treated with irradiation because of skull base extension with significant symptomatic relief.

Conclusions: The primary therapeutic option for paragangliomas is complete excision of tumor with preservation of vital neurovascular structures. Combined therapeutic approach with preoperative selective embolization followed by surgical resection is the safe and the effective method for complete excision of the tumors with a reduced morbidity rate.

Key Words: Paragangliomas, therapy, glomus caroticum, glomus jugulare

Paragangliomas of the head and neck are hypervascular, benign neoplasm embryologically derived from neural crest cells of the autonomic nervous system.[1] They were previously nominated as chemodectomas or glomus tumors, but the term paraganglioma is more accurate. They most frequently (60%) occur at the carotid bifurcation (carotid body paraganglioma, CBP). Additional sites of origin include the jugular bulb (jugular paraganglioma, JP), the vagal nerve (vagal paraganglioma, VP), the middle ear (tympanic paraganglioma, TP), the orbit, the nasopharynx, and the paranasal sinuses.[2] Cervical paragangliomas are most frequently unilateral and occur in a sporadic pattern. Multiple paragangliomas account for 11% to 22% of all cases. The 10% to 50% of paragangliomas are familial.[3] Bilateral CBPs are seen in approximately 31.8% of familial cases instead of 4.4% in sporadic ones.[4] Inherited forms are likely to be multicentric and bilateral and with an earlier onset of symptoms.[4–6] Malignant forms are uncommon, with an incidence ranging from 6.4% for CBP to 17% for VP.[7,8] For most cases, the ascending pharyngeal artery provides the primary blood supply to these paragangliomas.[9]

MATERIALS AND METHODS

A retrospective review of the medical records of the patients diagnosed with cervical paragangliomas was performed during the period from 2000 to 2007 years. All patients were treated at a single center of otolaryngology clinic (Istanbul University, Cerrahpasa Medical Faculty, Otolaryngology and Head and Neck Surgery Department). This study included 26 patients with 27 paragangliomas. There were 15 women (57.6%) and 11 men (42.4%) with a mean age of 33.5 years. In a female patient with CBP, bilateral involvement was noticed.

The most common presenting symptom was a painless lateral neck mass (16 of 27 patients; 61.5%). Other symptoms included dysphagia, voice hoarseness, tinnitus, and vertigo. No patient complained of symptoms related to a functional tumor with catecholamine hypersecretion such as palpitations, hypertension, tachycardia, or tremor.

Color Doppler ultrasonography (US) was performed as a first-line diagnostic procedure on 20 (76.9%) of our patients. Computed tomography (CT) with contrast enhancement was performed as a first diagnostic examination in 6 patients and as an adjunctive method in the remaining 20 (Fig. 1). Magnetic resonance imaging was performed on 10 patients in cases of suspected invasion of the skull base.

Digital subtraction angiography (DSA) through femoral artery catheterization followed the initial evaluation and proved a diagnostic condition in every case. Microcatheter embolization after visualization of vessels feeding the tumor was performed on 22 patients using straight and curled microcoils (Microcoils; William Cook Europe, Denmark) as the main embolic material together with Gelfoam and polyvinyl alcohol. Control angiography was performed to determine the level of vascular occlusion at the end of the procedure.

We graded CBP using the Shamblin classification (8). Type I tumors are defined as localized in between the internal and external carotid artery and can be easily resected. Type II tumors are adherent to or are partially surrounding the vessels, and type III tumors are tumors completely encasing the carotids.

Surgery was performed within 48 hours of embolization to avoid development of collateral vessels for tumor blood supply and the initiation of postembolization inflammatory effect. Surgical excision was performed under general anesthesia with an adequate approach through a semi-apron incision (a standard apron incision for the case of bilateral cerebellopontine angle). Proximal and distal control of the internal carotid artery and the external carotid artery was obtained with care to avoid injury to the adjacent cranial nerves. The dissection was performed in the relatively avascular subzadventitial plane between the artery and the tumor. In patients with class III tumors, a division and ligation of the external carotid artery were performed in advance on 4 cases. Opening of the common carotid artery was foreseen at 2 patients with glomus caroticum. No carotid shunts were used. Infratemporal fossa tip A incision was performed for JP and jugulotympanic paraganglioma. Follow-up

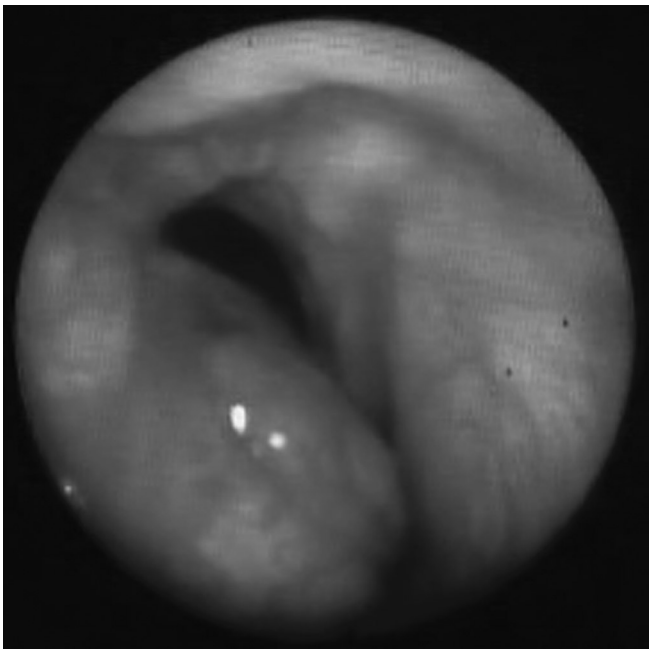

FIGURE 1. The axial CT revealed right-sided supraglottic laryngeal mass.

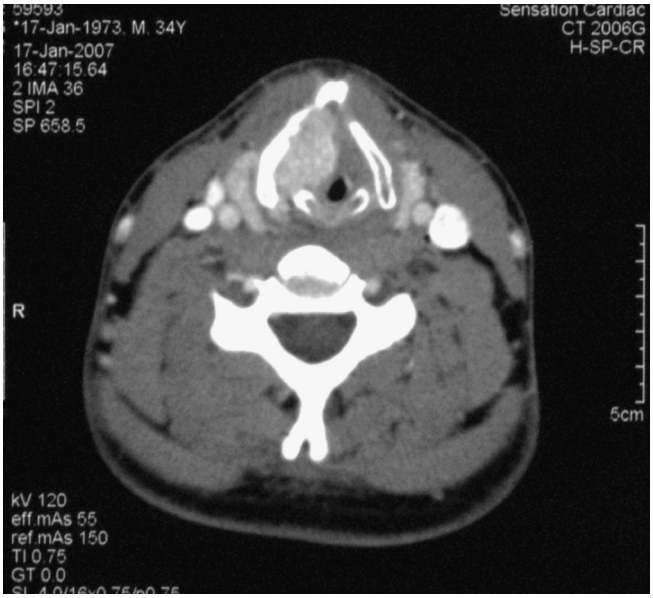

FIGURE 2. Laryngeal endoscopy demonstrating laryngeal paraganglioma in a patient with stridor.

examination consisted of carotid Doppler US and clinical examination in all patients annually.

All resected paragangliomas were examined under light microscopy for the characteristic arrangement of the tumor cells in distinctive cell balls, which is called Zellballen.

Two patients who refused the surgical treatment were treated with radiotherapy.

RESULTS

Carotid body paraganglioma was diagnosed in 13 (12 unilateral and 1 bilateral; 50%) patients. There were 6 patients with JP, 1 patient with a TP, 1 patient with a VP, 2 patients with jugulotympanic paraganglioma, and 1 patient with laryngeal paraganglioma (Fig. 2). Bilateral involvement was found in 1 patient having CBP. All patients were subjected to DSA to define the vascular supply of the tumor. We realized that ascending pharyngeal and superior thyroid arteries were the more common feeding vessels. Microcoil embolization to 22 paragangliomas (88%) was performed during the initial DSA. Three tumors had an unclear arterial supply through multiple tiny collateral vessels that could not be selectively embolized. Surgical excision of the both tumors was accomplished with a 3-month interval for patients with bilateral CBP.

According to Shamblin classification, concerning tumor–vessel relationship, 6 tumors (42.8%) were grouped into class I, 5 patients (35.7%) into class II, and 3 patients (21.5%) into class III. The mean size of the tumors was 3.6 cm (range, 2–8 cm). Radical resection of the paraganglioma was possible in all patients who accepted surgery. No perioperative mortality was documented. No transient ischemic attack or stroke was noticed in any patient as a complication of the embolization procedure. In 4 (16.6%) of our patients, a transient neurologic deficit was documented. In the 2 of all cases, the hypoglossal nerve was temporarily affected. Other nerve deficits included the vagal (n = 1) and the glossopharyngeal nerve (n = 1). All patients had resolution of their symptoms in a period of 2 to 3 months. Two patients with the VP and JP developed permanent Horner syndrome because of sympathetic chain damage (8.3%). Cerebrospinal fluid leak occurred in 1 patient with jugulotympanic paraganglioma during the operation; dura reparation was performed preoperatively for this patient. In 4 patients (16.6%), internal carotid artery injury developed during the surgery. These injuries were repaired with suturing the opening area. Common carotid artery dissection occurred in 2 patients (8.3%) during the operation; common carotid artery was repaired with suturing and ligation. In 1 patient, stroke was noted postoperatively. The other was normal postoperatively.

Pathological examination was diagnostic in every operated patient under light microscopy by recognition of the characteristic architecture of the tumor cells in cell balls (Zellballen). Indirectly, all tumors may be classified benign except one because no local recurrence and no regional and/or distant metastases were documented during a median follow-up period of 72 months. The patient with malign paraganglioma was treated with adjuvant chemotherapy and radiotherapy.

DISCUSSION

Paragangliomas are usually benign, slow growing tumors arising from widely distributed paraganglionic tissue thought to originate from the neural crest. Paraganglia are distributed throughout the head and neck and superior mediastinum along the course of the major vasculature. Paraganglia are also found in the orbit, the larynx, and along the course of the vagus nerve. Many different terminologies have been used in the past to describe these tumors based on their histopathologic and anatomic presentations. The tumors are thus divided into adrenal paragangliomas or pheochromocytomas and extra-adrenal paragangliomas. Cervical paragangliomas are described by their site of origin and are often given special names: carotid paraganglioma, glomus tympanicum, glomus jugulare, VPs, and other rare paragangliomas (laryngeal, optic, etc).

Carotid body paragangliomas characteristically present as slowly growing lateral neck masses. Horner syndrome may result from cervical sympathetic chain invasion. In our series, all of 15 patients with CBP referred us with a palpable neck mass. A patient with the CBP referred us with Horner syndrome in additional to cervical mass. Jugular paraganglioma originates in the jugular bulb near the skull base and extends both inferiorly in the parapharyngeal space and intracranially. It is not often accompanied by a neck mass. Symptoms are related to adjacent cranial nerve involvement (IX, X, XI, and XII).[10] In our series, 2 of 6 patients with a JP presented with symptoms indicating involvement of the XII cranial nerve. Vagal paraganglioma develops just below the skull base at the level of the nodose ganglion and is most frequently associated with progressive dysphagia and hoarseness with increasing tumor size. Also, 1 female patient with a VP presented with X and IX cranial nerve deficit.

Laryngeal paragangliomas are uncommon neuroendocrine neoplasms of neural origin that are thought to arise from the superior or inferior laryngeal paraganglia. More than 90% of laryngeal paragangliomas occur in the supraglottic larynx. Dysphonia is the most commonly reported symptom, followed by stridor, dysphagia, and dyspnea.[11–14] In our case, also there was supraglottic paraganglioma, and the patient applied us with stridor (Fig. 2). Carotid body paraganglioma represents the most common form of cervical paraganglioma that accounts for 60% of all neoplasms and is located at the carotid bifurcation. In our study, also a major group was CBP.

Most cervical paragangliomas are benign lesions. Malignant forms are uncommon, with an incidence ranging from 6.4% for carotid body tumors to 17% for glomus vagale.[7,8] Familial forms

have a decreased incidence of malignancy compared with sporadic forms.[4] The main criterion for malignancy is the presence of metastasis to the cervical lymph nodes or distant sites such as lung, bone, breast, and the liver.[15] In our 7 years' experience, we just saw a patient with malign paraganglioma with the presence of metastasis to the cervical lymph nodes.

A high index of clinical suspicion for the presence of a CBP is required to avoid an open or percutaneous biopsy that can lead to significant hemorrhage. Differential diagnosis is very important because a variety of lesions can have the same clinical appearance such as branchial cleft cysts, lymphomas, parotid gland tumors, thyroid masses, metastatic cervical lymphadenopathy, and tuberculosis. Color Doppler US represents the first step for the assessment of a cervical lesion.

In our study, 76.9% of patients were screened initially with Doppler US. During the period of this retrospective study, there has been no uniformly accepted diagnostic algorithm in the evaluation of a lateral neck mass. Eight patients were admitted to our department with a CT scan as a first imaging study. In the case where a cervical paraganglioma is suspected by US, a CT scan or magnetic resonance imaging represents the most important imaging technique for defining the relationship of the lesion to the adjacent anatomic structures. Computed tomography will show an enhancing mass. Bone windows can reveal skull base erosion and intracranial extension. Magnetic resonance imaging is more sensitive than CT for lesions involving the skull base or extending within the cranial vault.[2] The criterion standard with respect to detection of small paragangliomas is still DSA.[16–21] The ascending pharyngeal artery can be considered as the artery of the paraganglioma because its branches can supply tympanic, jugular, vagal, carotid, and even laryngeal paragangliomas.[22]

Surgical resection of paragangliomas can be complicated by profuse bleeding because of their high vascularity. Preoperative embolization can reduce intraoperative blood loss significantly. This is especially true for vagal and jugulotympanic paragangliomas.[10,23] The benefit of performing a preoperative embolization for carotid body tumors and TPs is poor. Hemostasis can easily be achieved during resection of carotid body tumors when the course of the ascending pharyngeal artery is taken into consideration. This artery supplies the tumor from rostral to caudal, and therefore, the surgical approach should focus on more distal vascular control to block rostral–caudal blood flow.[5,24] In our series of patients, microcoil embolization was the main technique performed after superselective catheterization of the major feeding arteries in percentage of lesions. Surgical approach depends on the localization and extension of the tumor.

Radiation therapy (RT) and radiosurgery may be indicated. Both classic fractionated RT and stereotactic radiosurgery (eg, gamma knife surgery) are successful in long-term control of tumor growth and in decreasing catecholamine excretion in functional tumors; however, the short duration of observation after stereotactic radiosurgery does not allow for definite conclusions. Radiation treatment is advised as the unique treatment modality for older or infirm, symptomatic patients, especially those with extensive or growing tumors. We used classic RT for 2 patients who refused the surgical treatment. One of them had huge glomus jugulare tumors that have intracranial extension. The other was an older patient with glomus jugulare.

In conclusion, our series shows that CBP is the most common tumor in glomus tumors as the literatures show. Laryngeal glomus is rare and may manifest as stridor. Doppler US is the first step to diagnose glomus tumor. Preoperative embolization can reduce intraoperative blood loss significantly. The primary treatment for glomus tumors is surgery, if necessary, followed by radiotherapy.[25,26]

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Simultaneous Pleomorphic Adenomas of the Hard Palate and Parapharyngeal Space

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Abstact: We report a very unusual presentation of simultaneous pleomorphic adenomas of 2 different locations, hard palate and parapharyngeal space. Patient age of presentation is rare because these tumors are seen in younger patients. We treated a 70-year-old woman with these 2 tumors, resecting both lesions with intraoral and cervical approaches. Pleomorphic adenomas are frequent lesions, but in the literature reviewed, we have not found articles reporting 2 simultaneous pleomorphic adenomas of these locations. Complete surgical resection is very important to avoid recurrences.

Key Words: Pleomorphic adenoma, parapharyngeal space, palate, salivary glands

The most common parotid gland tumor is pleomorphic adenoma, and these represent 70% to 80% of all benign parotid tumors.

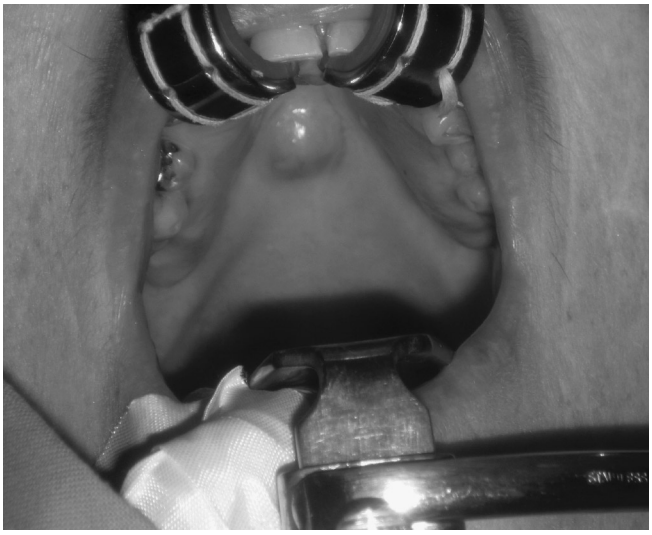

FIGURE 1. Surgical view of the pleomorphic adenoma of the hard palate, showing the integrity of the mucosa.

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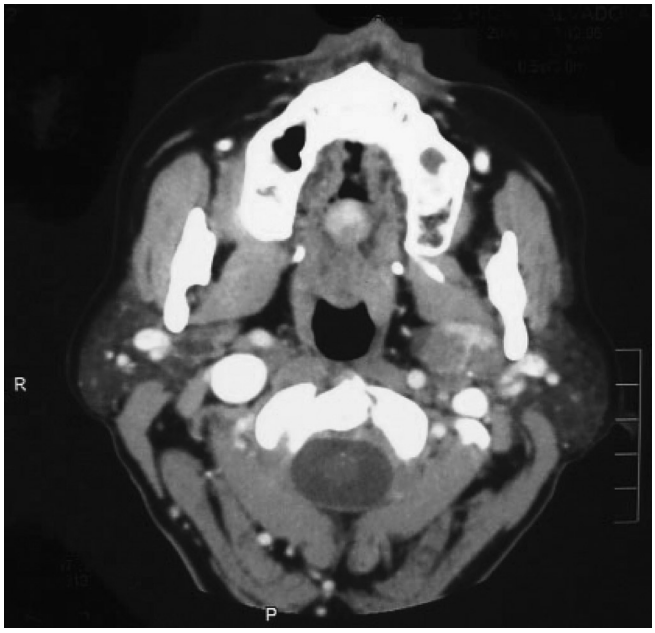

FIGURE 2. Axial CT scan with contrast showing the known lesion of the hard palate and the unknown lesion of the left parapharyngeal space.

These are typically seen in middle aged women and present as a painless slowly growing mass. These masses are typically solitary and well demarcated. They may appear heterogeneous secondary to hemorrhage, calcification, and necrosis.[1]

Computed tomography is an important diagnostic tool in tumors of parapharyngeal space because it helps in determining the extent of disease and local spread and also helps, to some extent, in determining the type of tumor. Presence of intact fat plane helps in distinguishing benign tumors from malignant. The treatment of pleomorphic adenoma is essentially surgical.[2]

Complete surgical removal is curative, but if the initial surgical procedure does not completely remove the tumor, there is a low recurrence rate of less than 2%. Sarcomatous transformation is seen in only 2% to 5% of cases and is usually associated with tumors that have been present for 10 to 15 years. The most common site of a pleomorphic adenoma of the minor salivary gland is the palate.

In the present study, we want to report the very unusual presentation of a patient with a simultaneous pleomorphic adenoma of the hard palate and of the parapharyngeal space.

CLINICAL REPORT

A 70-year-old woman presented with a 1-month history of a mass in the hard palate, without pain, bleeding, or any other symptoms (Fig. 1). The patient underwent computed tomography (CT) with intravenous contrast that showed the known lesion on the hard palate of 2 cm, without bone erosion, but there was another rounded lesion of 4 × 2 cm on the left parapharyngeal space, without infiltration of the surrounding structures, displacing back the vascular structures (Figs. 2 and 3). Both lesions were vascularized and took contrast.

Because of the findings of the CT scan, the patient underwent intraoral resection of the hard palate mass, which did not erode the bone and preserved the integrity of the palate mucosa, and a left cervicotomy to approach the parapharyngeal space, which found an encapsulated mass without continuity with the deep parotid lobe.

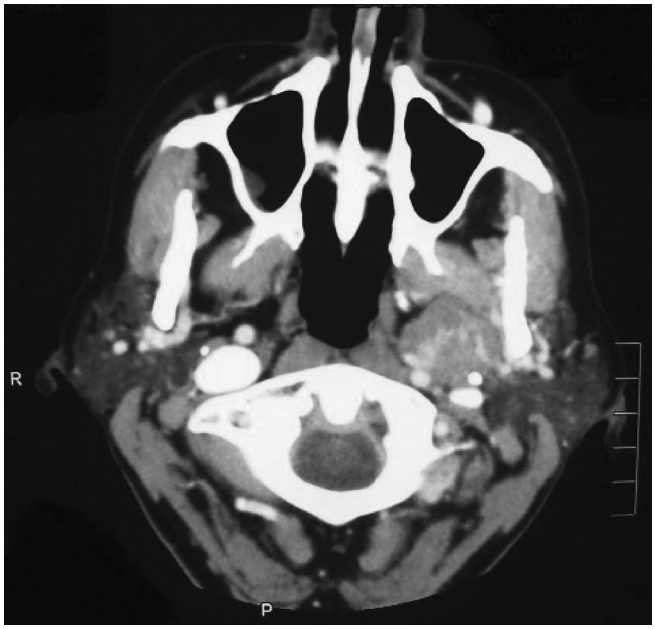

FIGURE 3. Axial CT scan showing how the tumor compresses the left internal jugular vein reducing its diameter.

Both lesions were completely resected and were sent for pathologic analysis that will confirm the diagnosis of pleomorphic adenomas.

The patient was discharged 8 days after the surgery.

DISCUSSION

Salivary gland tumors account for 3% of all head and neck neoplasm. Most of these occur in the parotid gland, and 80% are benign;[3] 50% to 80% of them are pleomorphic adenomas, and between 5% and 20% are Warthin tumors. Various primary neoplasms of the salivary glands are uncommon, Warthin tumor being the most common. The occurrence of multiple primary pleomorphic adenomas of the salivary gland is rare. Tumors of the major and minor salivary glands are extremely rare. The incidence of this kind in patients was only 1.8% in the largest reports.[4] The parotid and submandibular glands are the most prevalent locations.[5] Multiple primary pleomorphic adenomas in minor salivary glands are extremely rare, and we have not found articles describing this kind of presentation.

The most common site of a pleomorphic adenoma of the minor salivary gland is the palate followed by lip, buccal mucosa, floor of mouth, tongue, tonsil, pharynx, retromolar area, and nasal cavity.[6,7]

Although benign tumors of the minor salivary gland in the oral cavity present as a painless submucosal swelling, those from the parapharyngeal space may show symptoms, such as otalgia, neuralgia, dysphagia, palsies of 9th, 10th, or 11th cranial nerves, or trismus.[8]

In this case, tumors were simultaneously found in minor salivary gland of the hard palate and minor salivary gland of the parapharyngeal space, independent from the deep lobe of the parotid gland, without symptoms.

Computed tomography is the preferred diagnostic tool in tumors of parapharyngeal space because it allows determining the extent of disease and local spread and also in determining the type of tumor. If a fat plane is seen, it helps in distinguishing benign from malignant tumors; if a fine lucent line, representing the compressed layer of fibroadipose tissue between the tumor and deep lobe of parotid, is seen, we can assert that the pleomorphic adenoma depends on the deep lobe of a parotid gland; and if it is not seen, the tumor probably is arising de novo in the parapharyngeal space.[6]

The treatment of pleomorphic adenoma is surgery. These tumors are well encapsulated, but resection has to include an adequate margin of grossly normal surrounding tissue, to prevent local recurrence because these tumors have microscopic pseudopodlike extension through the capsule.[9]

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