cases with clear surgical margins, the 5-year survival rate was about 80%.[5,7] In the mandible, segmental mandibulectomy is commonly performed. The free fibula osteomyocutaneous flap is the common source of tissue used for oral and mandibular reconstruction.[20] Because of the complex anatomy, the reconstruction of maxillary cannot achieve satisfactory result. Three-dimensional model simulation technique and free fibula osteomyocutaneous flap flow-through from radical forearm flap were used by He et al[21] to reconstruct a total maxillectomy defect.

Radiotherapy was formerly commonly used as an adjuvant treatment before the introduction of chemotherapy. Recently, radiotherapy is mostly introduced to the cases with positive surgical margins. The benefit of chemotherapy for JOS has not been established. Smeele et al[22] investigated the value of chemotherapy in the treatment of craniofacial OS by analyzing 201 cases and concluded that the survival rate significantly improved with chemotherapy. However, some other authors argued that chemotherapy cannot improve the survival rate.[2,5,8]

In conclusion, we report a GCRO originating from the mandible and review literature of JOS. Like other types of JOS, GCRO of the mandible usually presented as a painless mass. It usually shows an osteolytic lesion on panoramic radiograph. Histologically, direct formation of osteoid by the malignant cells is the key factor in diagnosing GCRO and difference from GCRG. Surgical extensive resection should be performed on GCRO, similar to OS, with postoperative radiotherapy and chemotherapy. Although GCRO of the jaw is extremely rare, it should be considered when making a diagnosis of giant cell lesions.

REFERENCES

1. Fletcher CD, Unni KK, Mertens F. Osteogenic tumors. In: *Pathology & Genetics of Tumors of Soft and Bone*. Lyon: IARC, 2002:264–270
2. Mardinger O, Givol N, Talmi YP, et al. Osteosarcoma of jaw. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2001;91:445–451
3. Bathurst BN, Sanerkin N, Path FR, et al. Osteoclast-rich osteosarcoma. *Br J Radiol* 1986;59:667–673
4. Huvos AG. Osteogenic sarcoma in craniofacial bones. In: *Bone Tumors: Diagnosis Treatment and Prognosis*. Philadelphia, PA: WB Saunders; 1990:170–200
5. Clark JL, Unni KK, Dahlin DC, et al. Osteosarcoma of the jaws. *Cancer* 1983;23:2311–2316
6. Fernandes R, Nikitakis NG, Pazoki A, et al. Osteogenic sarcoma of the jaw: a 10-year experience. *J Oral Maxillofac Surg* 2007;65:1286–1291
7. Canadian Society of Otolaryngology-Head and Neck Surgery Oncology Study Group. Osteogenic of the mandible and maxilla: a Canadian review (1980–2000). *J Otolaryngol* 2004;33:139–144
8. Nissanka EH, Amaratunge EA, Tilakaratne WM, et al. Clinicopathological analysis of osteosarcoma of jaw bones. *Oral Dis* 2007;13:82–87
9. Ogunlewe MO, Ajayi OF, Adeyemo WL, et al. Osteogenic sarcoma of the jaw. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2006;101:76–81
10. August M, Magennis P, Dewitt D. Osteogenic sarcoma of the jaws: factors influencing prognosis. *Int J Oral Maxillofac Surg* 1997;26:198–204
11. Junior AT, de Abreu F, Pinto CA, et al. Clinicopathological and immunohistochemical analysis of twenty-five head and neck osteosarcomas. *Oral oncology* 2003;39:521–530
12. Sato K, Yamamura S, Iwata H, et al. Giant cell–rich osteosarcoma: a case report. *Nagoya J Med Sci* 1996;59:151–157
13. Bertoni F, Bacchini P, Staals EL. Giant cell–rich osteosarcoma. *Orthopedics* 2003;32:143–146
14. Shinozaki T, Fukuda T, Watanabe H, et al. Giant cell–rich osteosarcoma simulating giant cell tumor of bone. *Kitakanto Med J* 2004;54:147–151
15. Hong SJ, Kim KA, Yong HS, et al. Giant cell–rich osteosarcoma of bone. *Eur J Radiol Extra* 2005;53:87–90
16. Nagata S, Nishimura H, Uchida M, et al. Giant cell–rich osteosarcoma of the distal femur: radiographic and magnetic imaging findings. *Radiat Med* 2006;24:228–232
17. Kinoshita G, Yasoshima H. Giant cell–rich tumor of the rib. *J Orthop Sci* 2006;11:312–317
18. Unni KK. Osteosarcoma. In: *Dahlin's bone tumor, general aspects and data on 11087 cases*. 5th ed. Philadelphia, PA: Lippincott-Raven, 1996:143–196
19. Ha PK, Eisele DW, Frassica FJ, et al. Osteosarcoma of the head and neck: a review of the John Hopkins experience. *Laryngoscope* 1999;109:964–969
20. Donsin TB, Caruso PA, Nielsen GP. A 32-year-old man with pain and swelling of the jaw. *N Engl J Med* 2004;350:267–275
21. He Y, Zhu HG, Zhang ZY, et al. Three-dimensional simulation and reconstruction of composite total maxillectomy defects with fibula osteomyocutaneous flap flow-through from radial forearm flap. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2009; 108:e6–e12
22. Smeele LE, Kostense PJ, van der Waal JE, et al. Effect of chemotherapy on survival of craniofacial osteosarcoma: a systemic review of 201 patients. *J Clin Oncol* 1997;15:363–367

Primary Orbital Liposarcoma

Carlos Sevillano Torrado, MD, Nora Muruzábal Zaldibar, MD, Veronica Velasco-Benito, MD, Roberto Fernández-Hermida, PhD

Abstract: A 45-year-old-man presented a slightly painful proptosis and diplopia for 7 months. He had been kept elsewhere on oral steroids without evidence of any clinical response over an 8-week period with suspected diagnosis of an inflammatory pseudotumor upon referral to our clinic. An intraoperative biopsy positive for primary liposarcoma was followed by debulking surgery. Exenteration and radiotherapy were performed after pathologic confirmation of this diagnosis. No recurrence has been observed after 2 years of follow-up. We underline the importance of an accurate an early diagnosis in the management of this tumor, delayed in this case because of therapy with steroids.

Key Words: Debulking, exenteration, liposarcoma, orbit, steroids

CLINICAL REPORT

A 45-year-old man was referred to our clinic because of a slightly painful proptosis in his left orbit of 7 months of duration. Medical history was unremarkable, and the patient had been on oral corticosteroid for 2 months with a suspected diagnosis of orbit pseudotumor.

From the Department of Ophthalmology, Orbit and Oculoplastic Department, Hospital de Cruces, Baracaldo, Vizcaya, Spain.
Received August 10, 2010.
Accepted for publication September 25, 2010.
Address correspondence and reprint requests to Carlos Sevillano Torrado, MD, calle Joaquín Costa 19, 2°D; 36001 Pontevedra, Galicia, Spain; E-mail: seviojos@hotmail.com
The authors report no conflicts of interest.

ISSN: 1049-2275
DOI: 10.1097/SCS.0b013e3182108fdd

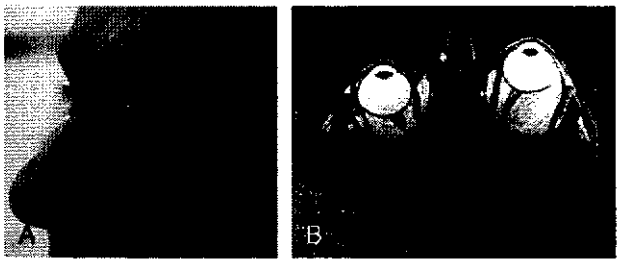

FIGURE 1. Photograph showing exophthalmos and magnetic resonance imaging showing the intraconical mass.

On examination, the patient had left-eyeball exophthalmos of 9 mm, conjunctival chemosis, and exposure keratopathy with a scleral inferior show of 5 mm (Fig. 1). A moderate reduction of both abduction and adduction was also noted. His best corrected visual acuity was 20/60. The rest of the examination (tonometry, Ishihara test, funduscopy, perimetry) was not contributory.

Chest x-ray and Mantoux test, plasma angiotensin-converting enzyme, protoplasmic-staining anti-neutrophil cytoplasmic antibodies, urine test, hemogram, and antithyroid antibodies levels were normal. The magnetic resonance imaging of the left orbit showed a large and well-limited intraconical tumor and medial displacement of the optic nerve (Fig. 1).

A lateral orbitotomy was performed to dissect and remove the mass. During the surgery, intraconal fat was found pale and ecchymotic. Intrasurgical samples of the fat were analyzed, provisional diagnosis corresponding to liposarcoma. At that stage, surgery was interrupted, and lateral wall was not replaced. Postoperative evolution was uneventful.

Two weeks later, diagnosis of primary liposarcoma of the left orbit infiltrating the muscular tissue was confirmed, and computed tomography total body revealed no other tumoral sites. The situation was discussed with the patient, and indication for extraperiosteal exenteration plus radiotherapy was settled.

Exenteration was uneventful, and socket was lined with dermoepidermal graft from lateral thigh area. External radiotherapy was started 4 weeks later, and the patient underwent a total of 33 sessions (a total of 59.4 Gy) of radiotherapy.

Histopathology showed a well-differentiated liposarcoma (adipocytic subtype) with myxoid changes; immunohistochemistry showed lipoblasts S100-positive (Fig. 2). No infiltration of the ocular globe was detected, but muscle involvement and also tumoral cells around the optic nerve were observed.

Three years after the surgery, the patient remains free of both local and systemic disease, with a good aesthetic result (Fig. 3). He is now waiting for an epithesis to be fitted.

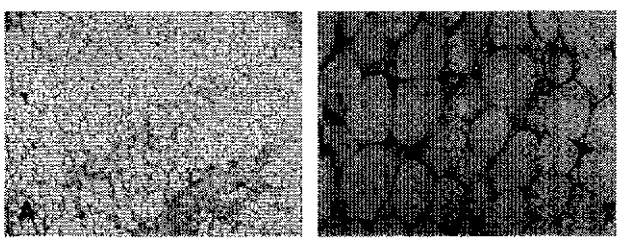

FIGURE 2. Histologic specimen showing lipoblast cells that are positive to S100.

FIGURE 3. Left socket lined with a dermoepidermal graft at 1 month after surgery.

DISCUSSION

Primary liposarcoma of the orbit occurs in the fifth to sixth decades of life,[1–7] with a predilection for sex not well established (males[2–4,7] or females[1]).

The most frequent histopathologic subtype, according to the classification of Enzinger and Winslow,[2] is the myxoid one, accounting for 45% of the cases.[1–4,6,7] The well-differentiated and pleomorphic subtypes are the least frequent (between 15% and 25%[1–3,7]). However, Cai et al[1] reported the well-differentiated subtype with myxoid changes to be the most frequent. Our patient's tumor was adipocytic.[1,2] This subtype has a subacute clinical course that can take from 3 months to as much as 7 years.[1,3–7] Painless proptosis with diplopia are common,[1] but only a few patients report loss of visual acuity or pain.[1–7] The well-differentiated subtype metastasizes only in 5%.[1,2,6,7]

Macroscopically, it usually has a gelatinous aspect with pseudocapsule[3–5,7] and color ranging from gray to orange.[3–5,7] Microscopically, it contains lipoblasts of different size with hyperchromatic nuclei,[1–7] and some myxoid material, typically with signet ring cells,[1–4,7] positive to S100 and negative to actin, myogenin, and desmin.[1–4,7] In the myxoid subtype, a typical chromosomic alteration (translocation 12,16) has been described.[1,6]

In addition, inflammatory cells can be seen, so these liposarcomas are thought to have a certain inflammatory component,[4] the reason why they can clinically worsen in a short period. Jakobiec et al[4] reported an orbit liposarcoma in a patient with Graves-Basedow disease, and Stiglmayer et al[6] reported other cases that improved partially after corticosteroid therapy. According to this, our patient developed worsening proptosis and chemosis in a 20-day period, which speaks for a high mitotic index mass (sarcoma-like), an inflammatory process, or a vascular mass with an acute bleeding inside. Diagnosis was delayed because of steroid therapy established in another center.

The myxoid subtype is the one that best responds to radiotherapy, the best therapeutic regimen being radical surgery with wide margins (or exenteration) followed by coadjuvant radiotherapy,[1,3,5–7] which reaches longer periods of survival.[1,4,5] Well-differentiated tumors respond poorly to radiotherapy,[3] but they usually have a good prognosis with wide excision.[1,7] Debulking is frequent after intrasurgical biopsy to decrease tumoral mass and to improve temporally the symptoms while a definitive diagnosis and treatment are obtained.[3–5] Because complete excision is needed to improve the prognosis[1,3–5] and muscle involvement was present,[4] we chose an orbital exenteration instead of a wide excision.

Exenteration can spare the eyelids[5,7] because the mass is confined to the intraconical space, although this is not always the case.[1] We did spare the lids. Rates of survival after surgery are close to 50% in 5 to 10 years[1,4,5,7] and significantly increase with coadjuvant

radiotherapy.[1,4,5,7] The main prognostic factors are the location and size of the tumor at the time of diagnosis and histologic type.[1,5–7] Although in our case no recurrence has been detected, we know that 3 years is too short to relax because there are cases described with recurrence after 5 years or more.[1,5,7]

In summary, a delay in diagnosis must be avoided,[4,5,7] so steroid use should be rejected or at least shortened[4,6] in cases of intraconal infiltration without a tissue diagnosis, and a biopsy should be scheduled as soon as possible.

REFERENCES

1. Cai YC, McMenamin ME, Rose G, et al. Primary liposarcoma of the orbit: a clinicopathologic study of seven cases. *Ann Diagn Pathol* 2001;5:255–266
2. Enzinger FM, Winslow DJ. Liposarcoma. A study of 103 cases. *Virchows Arch Pathol Anat Physiol Klin Med* 1962;335:367–388
3. Mridha AR, Sharma MC, Sarkar C, et al. Primary liposarcoma of the orbit: a report of two cases. *Can J Ophthalmol* 2007;42:481–483
4. Jakobiec FA, Rini F, Char D, et al. Primary liposarcoma of the orbit. Problems in the diagnosis and management of five cases. *Ophthalmology* 1989;96:180–191
5. Cockerham KP, Kennerdell JS, Celin SE, et al. Liposarcoma of the orbit: a management challenge. *Ophthal Plast Reconstr Surg* 1998;14:370–374
6. Stiglmayer N, Jandrokovic S, Miklic P, et al. Atypical lipoma: well-differentiated liposarcoma of the orbit with dedifferentiated areas. *Orbit* 2003;22:311–316
7. Madge SN, Tumuluri K, Strianese D, et al. Primary orbital liposarcoma. *Ophthalmology* 2010;117:606–614

Mucopyocele of the Concha Bullosa Presenting as a Large Nasal Mass

Mosaad Abdel-Aziz, MD

Abstract: Concha bullosa that is a pneumatization of the middle turbinate is a common anatomic variant; the obstruction of its ostium may lead to mucocele and even pyocele after infection of retained secretion. Although the condition is rare, mucopyocele of concha bullosa may be presented as a large nasal mass. However, the diagnosis could be suspected from its characteristic radiologic signs. We present an adolescent boy with mucopyocele of the concha bullosa.

Key Words: Concha bullosa, mucocele, middle turbinate, nasal endoscopy

From the Department of Otorhinolaryngology, Faculty of Medicine, Cairo University, Cairo, Egypt.
Received August 13, 2010.
Accepted for publication September 25, 2010.
Address correspondence and reprint requests to Mosaad Abdel-Aziz, MD, 2 El-Salam St, King Faisal, Giza, Cairo 202, Egypt;
E-mail: mosabeez@yahoo.com
The author reports no conflicts of interest.

ISSN: 1049-2275
DOI: 10.1097/SCS.0b013e318210b95a

Concha bullosa that is a pneumatization of the middle turbinate is one of the most common variations of the sinonasal anatomy. However, most patients are asymptomatic, but when it is large, it gives rise to nasal symptoms.[1] Mucocele is an epithelial-lined mucus-containing sac. Its common location is usually the frontoethmoid region; however, it may occur in any of the paranasal sinuses and may become infected with pyocele formation.[2] Excessive expansion of the concha bullosa with retained mucus that may become infected resulting in pyocele of the middle turbinate is a rare condition. Few cases had been reported in the English literature; in this study, we present an adolescent boy with a concha bullosa pyocele.

CLINICAL REPORT

A 17-year-old adolescent boy was presented with nasal obstruction in the right side that was progressive and of 2-year duration; the condition was associated with mucoid nasal discharge. Four days before presentation, he developed pain over the right side of his face that was referred to the ipsilateral eye. The patient has a history of facial trauma with fractured nose that was not treated. Anterior rhinoscopy revealed a large mass filling the right nasal cavity; the mass was covered with smooth intact mucosa, and it appeared to arise from the lateral nasal wall. Computed tomographic scan of the nose and the paranasal sinuses showed a soft-tissue lesion that was nearly filling the right nasal fossa and appeared to be arising from the middle turbinate. The lesion was surrounded by a thin plate of bone, and there were no findings that suggested bone destruction. The mass was expanded at the expense of the anterior ethmoid sinus with bowing of the nasal septum medially and medial wall of the maxillary sinus laterally; also, it compressed the inferior turbinate (Fig. 1). The patient was subjected to endoscopic sinus surgery for the removal of the mass, opening of the lateral aspect of the mass resulted in the appearance of a mucopurulent secretion that poured out from the sac inside the middle turbinate (Fig. 2). The wall of the sac was surrounded by a thin plate of bone that was easily crushed and removed with complete drainage. Endoscopic follow-up of the patient for 1 year showed no recurrence.

DISCUSSION

A paranasal sinus mucocele is a true cyst lined with pseudostratified ciliated columnar epithelium. It is a benign lesion and usually occurs in the ethmoid or frontal sinuses. If a mucocele becomes infected, it is referred to as a mucopyocele. Mucoceles of the paranasal sinuses develop because of the obstruction of normal mucociliary flow. This leads to a slowly expanding mass that becomes symptomatic while it impinges on nearby structures. A mucocele or mucopyocele can cause local bone erosion, diplopia, and nasal obstruction.[3]

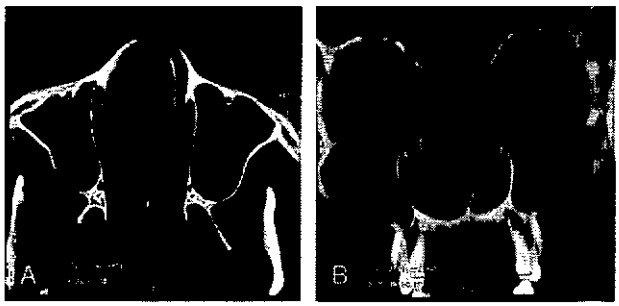

FIGURE 1. Computed tomographic scan of the nose and the paranasal sinuses axial (A) and coronal (B) views shows concha bullosa mucopyocele that fills the right nasal cavity.