Cranial Ossifying Fibroma Causing Visual Disorder

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Abstract: Fibrous lesions of the head and the maxillofacial region are generally slow-developing benign tumors that may aggressively take course in clinical terms. These locally destructive lesions show a high recurrence rates unless they are removed gross totally. Sarcomatoid degeneration is also reported to develop in recurring lesions. Difficulties in the diagnosis, classification, and treatment make this entity complex for several years. These tumors are frequently seen among children and young adults. The maxilla and the mandibula are the 2 most common sites that the tumor originates.

In our article, we discuss a 59-year-old man whose condition was diagnosed as an ossified fibroma revealing craniofacial extension. Accompanied by literature findings, it has been underlined that, as well as with the pathologic criteria, clinical features and radiologic images should be evaluated together for a specific diagnosis in fibrous lesions.

Key Words: Fibrous lesions, ossifying fibroma, cranial involvement

(*J Craniofac Surg* 2010;21: 768–770)

Fibrous lesions of the head and maxillofacial bones are generally slow-developing benign tumors that may aggressively take course in clinical terms. These destructive lesions have a high risk of recurring unless they are removed totally by surgeons.[1] The maxilla and the mandibula are the most common locations where the fibromas arise; however, they can also take place in the cranium, the nasal bone, the ethmoid, and the orbita, too. Classifying, identifying, and treating these lesions have resulted in many difficulties during several years. Benign tumors of the cranium are divided in 4 groups. Among these, except osteoma, ossifying fibroma, osteoid osteoma, and osteoblastoma rarely take place in the cranium.[2] Localized ossified fibroma (OF) cases around the head and neck regions are generally observed in the mandibula and the maxilla. Ossified fibroma cases on the nasal bone, the ethmoidal sinus, and the temporal and occipital bones have rarely been reported in the literature.[3,4]

In our article, we report a localized OF extending from the temporal region to the lateral orbital wall and the superior maxillary region. However, this lesion is suspected for the developing of exophthalmos in the right eye.

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Received November 15, 2009.
Accepted for publication November 27, 2009.
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The authors report no conflicts of interest.

ISSN: 1049-2275
DOI: 10.1097/SCS.0b013e3181d80a02

CLINICAL REPORT

A 59-year-old man was admitted to our clinic with the complaints of right-sided visual disorder and palpable lump on his right temporal region. She experienced this visual disorder nearly for 2 years since the lump appeared on his temporal region. He was not admitted to the hospital until he experienced severe headache and the lump started to grow up. Cranial computed tomography (CT) revealed an irregular multilobulated ossified mass that originated from the right lateral orbital wall and extending into the greater wing of the sphenoid bone and the frontolateral regions and outside the cranial cavity, extending in and out (Figs. 1A, B). The ossified mass was seen to contract the inferior orbital fissure opening that resulted in the ridging of the right superolateral orbital wall to the inside of the orbita. Physical examination showed a palpable mass lesion extending from the temporal region to the right orbital wall (Fig. 2). The lower part of the mass was invasive to the upper maxillary region. Amaurosis was diagnosed in the ophthalmologic examination of the right eye. The patient underwent right frontotemporal craniotomy and orbital decompression surgery under general anesthesia. The mass lesion was excised totally (Fig. 3). Asymmetric appearance on the face disappeared after the surgery. Postoperative CT did not reveal any residual tumor (Fig. 4). Histopathologic diagnosis was consistent with OF (Figs. 5A, B). The patient was discharged uneventfully.

DISCUSSION

Several heterogeneous fibrous lesions related to the craniofacial bones have been described. Classifying, identifying, and treating these lesions have caused various difficulties for several years. Today, OF, cementing fibroma (SF), cemento-ossified fibroma (SOF), fibrous dysplasia (FD), and periapical cemental dysplasia are the most commonly identified lesions in this group. The first 3 lesions are classified subjectively owing to the production capability of cementlike materials and osteoid within fibrous stroma.[5,6] As opposed to FD and periapical cemental dysplasia, it is accepted that these lesions are of

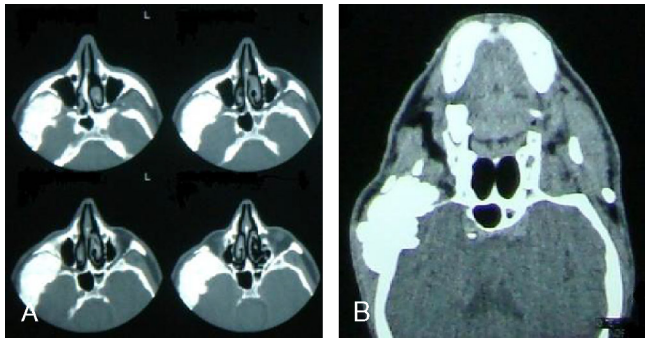

FIGURE 1. A, Preoperative cranial CT scan revealing an ossified mass extending from the right lateral orbital wall to greater wing of the sphenoid bone and frontolateral section. B, Irregular mass extending in and out of the cranium.

FIGURE 2. Lump causing asymmetry on the patient's face is seen on the right.

neoplastic structure. Today, the most common ones of these lesions are gathered under the title of *fibro-osteo-cemental lesions*. Benign, well-limited, unilocular or multilocular fibrous lesions show a slow and nodulelike development, and a gross total excision is sufficient for preventing recurrence. Rarely, a tumor may quickly develop, enlarge, and cause various cosmetic and functional problems. Tumors displaying such aggressive courses are defined as aggressive, juvenile, or active OF, SF, or SOF.[1,7,8] The term *juvenile* is used especially in the lesions that are seen around children and a young adult population. An aggressive course observed on the middle part of the faces may cause local destruction and recurring. Although the aggressive course has been assumed for tumors that have eroded or invaded the bones, the relation between histologic appearance and clinical attitude is not clear enough.[1,9] For this reason, especially tumors showing an aggressive course need to be distinguished from malignant tumors such as osteosarcoma, by means of experience in radiologic and surgical areas.[10] Many retrospective studies stated that lesions located on the maxilla might have a more aggressive course. Marvel et al[11] stated that OF cases that had arisen on the middle face and on the paranasal sinuses were more aggressive, and they suggested local excision for providing the control.

Many subtypes of OF have been named *juvenile ossified fibroma*, *juvenile aggressive ossified fibroma*, *juvenile active ossified fibroma*, *cemento-ossified fibroma*, or *psammomatoid ossified fibroma*, and all these terms are being used instead of OF today.[12–14] In 1782, for the first time in history, Menzel identified a big tumor that took place on the mandible of a 35-year-old woman as SOF, namely, a variant of OF. In 1927, Montgomery used the term *OF* for the first time. Liechtenstein and Jaffe were the first to use the term *FD* in 1938. The monostotic form of FD was identified by Liechtenstein and Jaffe, whereas Albright described its polyostotic form in 1937. Thus, McCune-Albright syndrome that consists of pigmentation changes on the skin, endocrine disorders, and polyostotic FD was identified.[11,15] Liechtenstein and Jaffe proposed that all fibrous lesions of the facial bones should be accepted as the subtypes of FD, and this opinion was valid until 1963.[3] In 1963, Reed distinguished these lesions according to their clinicopathologic qualities and accepted FD as a development defect, whereas OF as a benign neoplasm.[11] In 1968, Hamner accepted these lesions as tumors including cement.[16] First comprehensive classification of these lesions has been done by the World Health Organization in 1971, and they divided these tumors into 4 groups: (1) FD, (2) OF, (3) SF, and (4) SOF.[16,17] After some clinical experience, it has been seen that this classification cannot answer all questions, and new lesions were added after it has been reviewed. For instance, in 1983, Makek identified desmo-osteoblastoma, which develops from facial and cranial bones. Desmo-osteoblastoma originating from osteoblastic cells were very aggressive in behavior and had 2 subtypes called *psammous* and *trabecular*.[4] Later, in 1987, Makek divided fibrous lesions into 4 large subgroups: (1) development disorders, (2) reactive-reparative lesions, (3) fibromatosis, and (4) tumors. Benign tumors have been divided into 2 groups as the ones with odontogenic roots and nonodontogenic ones taking place on the face and head bones.[4] Oral pathologists traditionally classify these lesions according to whether they have odontogenic roots or not.[18] These lesions, which are observed frequently between the second and fourth decades of life, are seen more frequently in women.[18,19] In some studies, as opposed to adults, development has been reported to be faster and had a more aggressive course on children.[3,20] Regarding this fact, authors interpreted that the proliferative activity of the tumor decreased as time passed.[3] As opposed to FD, OF is

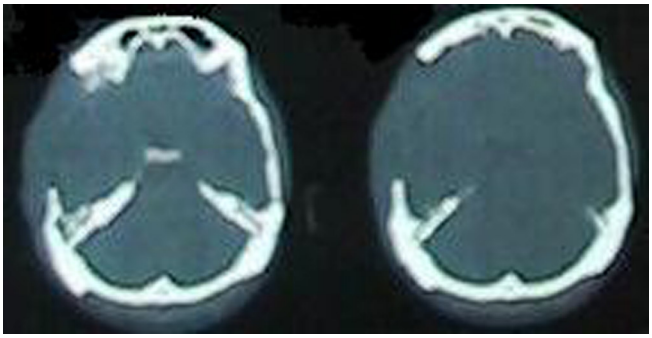

FIGURE 4. Postoperative cranial CT scan revealing no residual tumor.

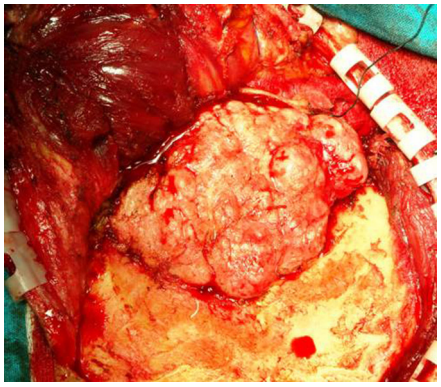

FIGURE 3. View of the mass lesion.

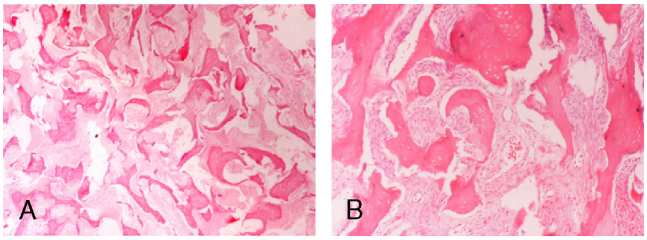

FIGURE 5. A, Pathologic view of the mass showing spindle cell proliferation and bone formation (hematoxylin and eosin, original magnification ×40). B, Narrow, curved, misshaped bone trabeculae, characteristic fishhook configuration, and fibrous tissue of variable cellularity (hematoxylin and eosin, original magnification ×100).

monostotic, and multiple bone involvement of less than 5% is seen. Mandible involvements that are observed in some series, approximately 75%, are the most frequent location for the lesions to take place. The ethmoidal, frontal, and sphenoid sinuses; the orbita; the occiput; and the temporal bone follow.[18] The most common location for FD to take place is the maxilla. The Lesions of the ethmoidal and sphenoid sinuses are seen less commonly. The monostotic form is observed approximately 70% to 75%, whereas the involvement of the craniofacial bones in these lesions is approximately 30%. The polyostotic form is observed approximately 30%, and in these lesions, which are observed in younger ages, the involvement of the craniofacial bones is approximately 50%.[18] Clinical findings may vary according to the tumoral bone involvement. Lumps, sensation disorder, and asymmetry are the most common symptoms especially in patients with a lesion of the jaw.[8,11] Unilateral proptosis, diplopia, persistent nasal obstruction, rhinorrhea, epiphora, recurring epistaxis, and hemoptysis may be observed in the lesions of the middle face and the paranasal sinus.[11]

Radiologic appearance varies according to ossification rate and location. Lesions having unilocular or multilocular characters may be radiolucent or mixed radiolucent-radiodense. Generally, lesions are surrounded by a radiodense boundary.[7,8,21] Sometimes, invasion or erosion on the surrounding bone may be observed.[7,22] Histopathologically, normal lamellar bones in these lesions are replaced by osteoid or bone that has taken place in vascularized fibrous stroma, and most of which has been hardened.[17] Osteoblastic chain and randomly spread osteoclasts are observed around the bone trabeculae in stroma in OF that has a richer character than the cell in which swollen fibroblasts have been monitored.[1,17,19] Especially on the periphery of the lesion, transformation is observed on the lamellae on bone trabeculae. These lesions, which are of local destructive characters and which may cause deformity in bones, have a high risk of recurring unless they are removed totally by surgery.[8] Radical resection, conservative local excision, and enucleation by curettage are the treatment options.[1,8] Incomplete resection especially in the cases of aggressive tumors causes recurrence.[10,20] It is reported that sarcomatoid degeneration develops in cases of long-term recurring lesions.[16]

Our case, in which there are a cosmetically asymmetric disorder on the face and visual loss, is rarely seen with a craniofacial involvement. One of the important points is that the tumor has a clear boundary on the sound bone structures that it has invaded, and it has been spread over a very wide area. As a result, correlation of clinical, radiologic, and pathologic data is very important while going for a specific diagnosis in cases of craniofacial fibrous lesions. Total excision is curative.

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