Case Report

Orbit ossifying fibroma – Case report and literature review

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INTRODUCTION

Ossifying fibroma (OF) is benign bone lesions, most frequent in young children (10 years). They are more found in the maxillary sinus and mandible (75–89%). It has also been reported in the paranasal sinuses, frontal, ethmoid, and sphenoid bones, and orbital roof.¹⁵

Primary orbital bone tumors represent 0.6–2% of all orbital tumors and 23% of all lesions involving bone, the most common of which are fibrous dysplasia (FD) and osteoma.¹⁵ The incidence of orbital OF is difficult to estimate; however, a review of 3000 fibro-osseous lesions

ABSTRACT

Background: Ossifying fibroma (OF) is benign bone lesions, most frequent in young children, more common in the maxillary sinus and mandible (75–89%), the pathogenesis of the tumor is not clear, there are many subtypes of OF. This paper aims to report an OF a case and literature review.

Case Description: Male, 19 years old, with a progressive history proptosis since 2012, diagnosed as a right supraorbital lesion at an external service and assigned to conservative management. Then, he evolved with double vision, which worsened in February of 2018, associated with a moderate headache. On admission: proptosis and downward deviation of the right orbit was noticed on the physical exam and with exception of limited right upgaze, external ocular movements were maintained. Head computed tomography showed a multiloculate expansive osteolytic lesion at the right orbital roof. On magnetic resonance imaging, the lesion had an inner content with septations, T1-weighted imaging heterogeneous signal, T2-weighted imaging high signal intensity, and peripheral contrast enhancement. The patient underwent a right frontal craniotomy with a gross total resection and the postoperative follow-up was uneventful. Menzel reported the first case in 1782. The clinical findings depend on localization. There are five subtypes. In general, the lesions have a radiological appearance with hyperdense boundary and cause deformity and destruction in bones with high recurrence risk. Radical resection is curative.

Conclusion: As a result, the correlation of clinical, radiologic, and pathologic data is significant while going for a specific diagnosis in cases of craniofacial fibrous lesions. Total excision is the best treatment, but it can recur.

Keywords: Cranium tumors, Neurosurgery, Orbit lesions, Ossifying fibroma, Proptosis
collected by the Armed Forces Institute of Pathology reported 30 cases.[7,15]

The pathogenesis of the tumor is not clear and a few theories have been proposed. Trauma has been suggested as a biological factor. A developmental abnormality has also been recommended since this tumor usually arises from membranous bone and occurs in patients in a young age group.[15]

They are generally slow-developing benign tumors. However, these lesions are destructive and have a high risk of recurrence unless surgeons eradicate them. The term juvenile is used primarily in children and a young adult population occurrence and can be more aggressive than adults. OF which arise on the middle face and the paranasal sinuses are also more aggressive.[6,12]

**CASE REPORT**

D.L.F., male, 19-year-old, presented a progressive proptosis in 2012, associated with occasional mild headache. On the same year, he sought medical consultation at an external service and after imaging investigation, a right supraorbital lesion was found. It was decided conservative management with imaging follow-up. Three years later, the patient joined a phosphoethanolamine clinical trial, which he left after approximately 1 year. At that time, he also presented with double vision. On February of 2018, the headache worsened, as well as the diplopia. Due to the worsening, he was referred to the Hospital das Clinicas at the University of Sao Paulo. In the neurological exam, he was alert and oriented; there was proptosis (Grade I) and downward deviation of the right eye. The external ocular movements, visual acuity, and campimetry were preserved, and there were no motor or sensorial impairments.

Head computed tomography (CT) showed a multiloculated expansive lesion on the right orbital roof, sized 3.3 × 4.0 × 2.8 cm (LL × AP × CC), with osteolytic characteristics. There was a bulging of the anterior cranial fossa and right frontal sinus superiorly. The superior orbital fissure and the optic nerve canal were preserved.

Magnetic resonance imaging (MRI) revealed a well-circumscribed multicystic lesion with peripheral hypointense capsule and mild internal septations with variable signal intensity at T1-weighted imaging (T1WI). On T2, the cystic cavities showed high signal intensity. There was the obliteration of the extra and intraconal fat tissue and inferior dislocation of the lachrymal gland and extraocular muscles. Cystic walls were enhanced after injection of gadolinium [Figure 1].

On March 7, he underwent a right frontal craniotomy for resection of the supraorbital lesion associated with high-speed drilling. Was made extended frontal pretemporal craniotomy, with wide orbital skull exposure. The main objective due to decompress optic nerve. All bone block was removed by drilling. Afterword’s the skull was reconstructed by methacrylate. The drilling was very tough. A solid bone makes it more difficult to remove. The postoperative follow-up was uneventful, with reduction of the proptosis [Figure 2]. He was discharged on the 4th postoperative day. Figure 3 shows the pathology findings.

**Figure 1:** Expansive cystic bone formation in the right orbital roof, with peripheral enhancement through contrast with thin septations, measuring 3.3 × 4.0 × 2.8 cm (LL × AP × CC). It presents clear limits, with a “frosted glass” aspect in the bone portion in its periphery, especially in its inferomedial aspect. (a-c) Magnetic resonance imaging images, axial, sagittal, and coronal view, respectively. (d and e) Computed tomography images, coronal and axial view (f) there is consequent degree I proptosis of this side and displacement of the optic nerve.
DISCUSSION

There are many differential diagnoses for bone lesions in the skull base. The spectrum of possibilities includes benign lesions, as osteomas and osteoid osteomas, reactive expansile nonneoplastic lesions, primary malignant tumors, and metastatic lesions. Although rare, fibro-osseous lesions are one of these possibilities.\[^{3,4,10,11}\]

Benign fibro-osseous lesions (BFOLS) are characterized by a fibrocellular stroma with varying degree of mineralized material leading to the replacement of physiological bone architecture with fibro-osseous tissue. FD, OF, and cemento-osseous dysplasia are the noted BFOLS.\[^{14}\]

The recent World Health Organization classification of head and neck tumors in 2017 described four fibro-osseous lesions: OF, FD, familial gigantiform cementoma (FGC), and cemento-osseous dysplasia.\[^{2,5,8,9,13}\]

OFs are benign fibro-osseous neoplasms affecting the craniofacial skeleton.\[^{4}\] There are three variants: cemento-

![Surgical images](image1)

**Figure 2:** Surgical images: (a) frontopterional approach, (b) bone exposure, (c and d) intraoperative aspect of the ossifying bone, (e and f) postoperative results: improvement of exophthalmos and paralysis of the lateral rectus muscle.

![Histological images](image2)

**Figure 3:** Juvenile psammomatoid: (a and b) hematoxylin and eosin analysis: disconnected spicules of irregularly shaped bone often with hemorrhage and occasional giant cells; prominent osteoblast lining may be seen, as well as occasional partially lamellar bone with numerous round psammoma-like calcified bodies are identified. (c-e) Immunohistochemistry analysis: the markers performed are only for a differential with meningothelial/neural sheath tumors. The three are negative: progesterone receptor, epithelial membrane antigen, and S-100.
OF (COF), juvenile trabecular OF (JTOF), and juvenile psammomatoid OF (JPOF).\textsuperscript{[5]} COF is a slow-growing benign neoplasm with an odontogenic origin. It is rare with a peak of incidence in the third and fourth decades of life and female predilection (5:1). Affects exclusively the mandible and maxilla.\textsuperscript{[6]} The surgical resection is the main treatment and recurrences are rare.\textsuperscript{[1]} JTOF is also rare with no gender predomination and occurs mainly in children between 8 and 12 years. Affects predominantly the maxilla and mandible. Extragnathic occurrence is rare. JPOF is an uncommon tumor which the mean age ranges from 16 to 33 years. It occurs mainly in the extragnathic craniofacial bones, as the orbit and ethmoid in 72% of the cases. Radiographically, the JPOF is usually well delineated at the periphery with varying degrees of radiolucency and radiopacity. Lesions that are more aggressive may show cortical thinning and perforation. Being well-demarcated distinguishes it from other fibro-osseous lesions such as FD.\textsuperscript{[1,9]} Both JTOF and JPOF are benign and the treatment should be the complete resection. However, recurrence is higher than in COF and malignant transformation has not been reported.\textsuperscript{[10,11]}

FD is a skeletal developmental anomaly in which normal medullary bone is replaced by fibrous tissue. The lesions of the ethmoidal and sphenoid sinuses are seen less commonly. The monostotic form is observed in approximately 70–80%, whereas the involvement of the craniofacial bones in these lesions is approximately 25%. The polyostotic form is observed in approximately 30%, and in these lesions, which are observed in younger ages, the involvement of the craniofacial bones is approximately 50%. The association between polyostotic disease, abnormal skin pigmentation, and endocrinopathies may configure the McCune-Albright syndrome.\textsuperscript{[2]} Clinical findings may vary according to the tumoral bone involvement (headache, facial pain, proptosis, vertigo, facial asymmetry, cranial nerve dysfunction, sinus infections/inflammation, visual or auditory impairment, and visual disturbances). The most common appearance of FD on CT scan is an expanded bone with ground-glass appearance. In monostotic FD, MRI often mimics a tumor: the fibrous tissue tends to enhance and signal intensity may be intermediate both in T1- and T2-weighted images and it enhances after gadolinium injection as a well-vascularized lesion. Sarcomatous degeneration can occur in 4–11% of the cases. Surgical resection is the main option when it is feasible and should be considered in cases of progressive growing, cranial nerve compressions, and sarcomatous transformation.\textsuperscript{[13]}

| Table 1: Summary of different types of fibrous-osseous lesions. |
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| Disease | Clinical features | Radiologic findings | Histopathology |
| Ossifying fibroma | COF | Painless expansion | Radiolucent lesions and progressive radiopaque | Encapsulated. Hypercellular fibroblastic stroma containing variable amounts of calcified structures. The stromal cells have hyperchromatic nuclei but no marked atypia. Mitosis is not easily found |
| JTOF | This rapid expansion affects maxilla with obstruction of the nasal passages and epistaxis can occur | Radiolucent with various degrees of opacification. Cortical thinning and perforation can occur | Aneurysmal bone-cyst formation |
| JPOF | Bony expansions that involve the orbit or nasal bones and sinus can result in proptosis, visual symptoms, and nasal obstruction | | |
| Fibrous dysplasia | | Diffuse radiolucent/ground glass | |
| Familial gigantiform cementoma | Early-onset of multifocal/multiquadrant progressively expansive lesions that may be massive and cause remarkable facial deformity. No other bones are affected | Expansive masses with well-circumscribed borders presenting as radiolucent areas containing radiopaque calcifications | Analogous to COF: hypercellular fibroblastic stroma with monomorphic appearing spindle-shaped fibroblasts and collagen fibers |

COF: Cemento-ossifying fibroma, JTOF: Juvenile trabecular ossifying fibroma, JPOF: Juvenile psammomatoid ossifying fibroma
FGC is a rare fibro-osseous disease. It affects the jaws characterized by a multifocal lesion with progressive growth. It can be autosomal dominant but sporadic cases are also described. The treatment is challenging and complete surgical resection usually is impossible and the recurrence rates are high.\cite{5}

Cemento-osseous dysplasia is the most common benign fibro-osseous lesion of the jaw. It occurs exclusively in the tooth-bearing regions of the jaw. In general, require no treatment and can be monitored during routine dental appointments.\cite{5}

Clinical, radiographic, and microscopic parameters that distinguish among OFs and FD are summarized in Table 1.

CONCLUSION

There are many differential diagnoses in skull base lesions. Most common are benign bone tumors; however, fibro-osseous lesions should be remembered mainly FD and OFs as possible causes. In general, these lesions must be treated with a complete surgical resection, minimizing the recurrence rates.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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HIGHLIGHTS

• OF are benign bone lesions, most frequent in young children, and maxillary sinus and mandible (75–89%)
• Primary orbital bone tumors represent 0.6–2% of all orbital tumors and 23% of all lesions involving bone
• Head CT showed a multiloculate expansive osteolytic lesion at the right orbital roof. On MRI, the inner content had septations, T1WI variable signal intensity and T2-weighted imaging high signal intensity with fluid-fluid level, and contrast enhancement of its walls
• Pathology: disconnected spicules of irregularly shaped bone often with hemorrhage and occasional giant cells; prominent osteoblast lining may be seen, as well as occasional partially lamellar bone. The immunohistochemical markers performed are only for a differential with meningotheial/neural sheath tumors. The three are negative: progesterone receptor, epithelial membrane antigen, and S-100
• As a result, the correlation of clinical, radiologic, and pathologic data is significant while going for a specific diagnosis in cases of craniofacial fibrous lesions. Total excision is the best treatment, but recurrence can be seen.