

CASE REPORT

Massive Juvenile Ossifying Fibroma of the Frontal Bone

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ABSTRACT

Ossifying fibroma is a rare, benign fibro-osseous lesion composed of lamellar bone and fibrous tissue. It occurs as an osseous lesion in the craniofacial skeleton. Clinically, lesions are usually asymptomatic, slow-growing and well-circumscribed. However, in very few cases, particularly in younger patients these tumors have demonstrated an aggressive course of development. Surgical management via a wide local excision is a necessity since it is notorious for recurrence, especially paranasal sinuses lesions. Here, we describe a case of a 9-year-old male child who had left forehead swelling with painless, progressive proptosis and downward lateral displacement of the globe for approximately 8 months. Computed tomography showed a massive expansile lesion involving the left frontal bone. The mass was excised surgically via a Lynch Howarth approach and proved histopathologically to be a juvenile ossifying fibroma. The radiological interpretation will be discussed which is helpful for diagnosis and selecting appropriate surgical approach in growing child.

Keywords: Fibro-osseous lesions, Ossifying fibromas, Frontal bone.

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INTRODUCTION

Ossifying fibroma is a rather uncommon fibro-osseous tumor which is known to involve the bones of the orbit and paranasal sinuses and produce proptosis. In recent years, the term fibro-osseous lesions has grown in popularity as an overall designation for a number of rare, histologically benign lesions of the head and neck that are made up of bone, fibrous tissue and cementum.¹ Presence of cementum or bone classifies the lesion as cementifying fibroma or ossifying fibroma respectively while lesions with mixture

of both cementum and bone matrix are known as cemento-ossifying fibroma. According to the WHO classification in 1992, ossifying fibroma is classified within nonodontogenic lineage tumors.² The mesodermic origin of the tumor has been widely accepted, and it is thought to be originating from the periodontal ligament.³ It contains multipotential cells capable of forming cementum, lamellar bone and fibrous tissue. Under pathological conditions neoplasms containing any or all of the components may be produced.⁴ Ossifying fibroma is a locally destructive, deforming tumor that can occur almost anywhere in the craniofacial region. Most osseous lesions have been reported in the premolar-molar region of the mandible; other sites include the maxilla, zygoma, paranasal sinuses, orbital and petromastoid regions.^{5,6} Trauma is widely accepted as the predisposing factor for these lesions.^{3,5} Ossifying fibroma occurs in patients of a wide age range, but most cases are encountered during the third and fourth decades of life and more commonly in women.⁵ They manifest themselves as asymptomatic, slow-growing and well-circumscribed intraosseous masses. These lesions can produce sinus obstruction, infection, facial deformity, proptosis and intracranial complications, leading to considerable esthetic and functional deformities. The clinical behavior and radiological appearance of fibro-osseous lesions is variable, based on evolution time and development phase. Understanding the nature of fibro-osseous lesions facilitates appropriate management. Aggressive lesions require a radical surgical approach to ensure complete excision, despite an increase in associated morbidity. Complete excision is dependent on the correct surgical approach. Incomplete excision of aggressive lesions may result in recurrence with severe morbidity or mortality. In contrast, a slow progressive lesion often does not warrant extensive surgical excision. Here we present a case of ossifying fibroma of the frontal bone in a 9-year-old boy.

CASE REPORT

A 9-year-old male child with no prior ocular or systemic problems developed slowly progressive proptosis and downward displacement of the left eye with forehead swelling over an 8 months period. He was referred to the otorhinolaryngology department for further management. On examination, a diffuse nontender bony hard swelling fixed to underlying bone with normal overlying skin over left forehead with irregular left supraorbital margins was seen.

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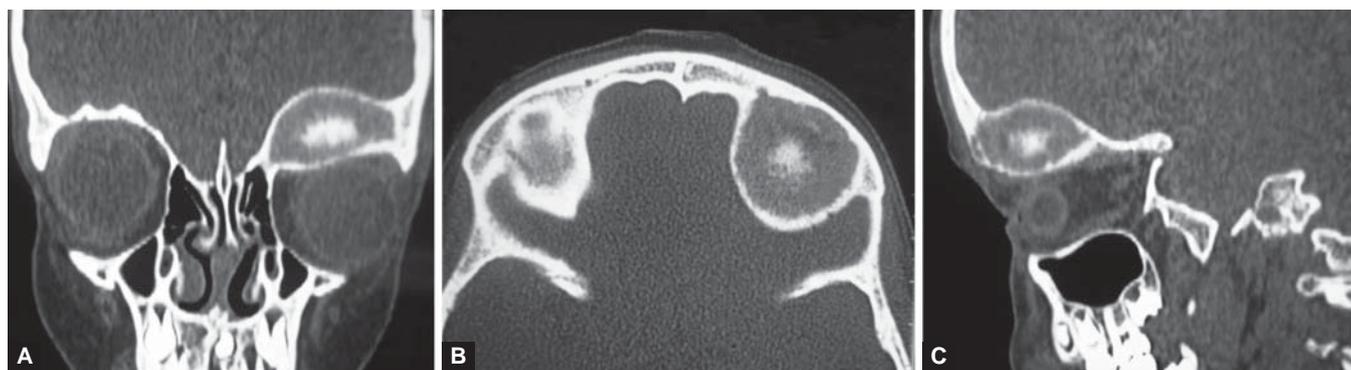
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The roof of orbit was bulging and on palpation a smooth, nontender hard swelling was found. External examination demonstrated mild facial asymmetry and left proptosis. Ocular examination disclosed a normal visual acuity in both eye. There was obvious proptosis, with downward ward displacement of the left eye. There was a bilateral normal eye movement. Remaining ENT examination was unremarkable. Computed tomography (CT) without contrast material in axial, sagittal and coronal planes showed a large, well circumscribed mass in the left frontal bone with left orbit and intracranial expansion (Fig. 1). The lesion was nonhomogeneous owing to foci of ossification in the center. The frontal bone was displaced around the edge of the lesion. The lesion was well-defined and showed both radiolucent and radiopaque features. On the basis of the clinical and CT findings our clinical diagnosis was a fibro-osseous lesion, most likely ossifying fibroma. The patient was managed by surgical resection via a Lynch Howarth approach. Under general anesthesia a Lynch Howarth incision was made and extended superiorly to expose the roof of orbit. The periosteum was elevated and the thinned inferior frontal wall was exposed (Fig. 2). The large size and unyielding nature of the mass made removal in one piece impossible, therefore the gritty tumor was removed piecemeal. During the dissection, the lesion was easily separable from adjacent structures. The 0° endoscope was used for facilitating the complete removal of the tumor from the frontal bone. After complete removal of the mass, the cavity was irrigated and a corrugated drain was put for 24 hours. Blood loss was less than 200 ml and there were no surgical complications. The excised surgical specimen (Fig. 3) was sent for histopathological examination which revealed a diagnosis of ossifying fibroma of bone. The postoperative period was uneventful and the patient was advised regular follow-up. The patient is doing well, and has no ocular problems approximately 6 weeks after the surgery.

DISCUSSION

The World Health Organization lists four kinds of fibro-osseous lesions: cemento-ossifying fibroma, fibrous

dysplasia, ossifying fibroma and cementifying fibroma.² Ossifying fibroma tends to be asymptomatic in the initial phases and is frequently diagnosed in advanced stages. There is localized increase in volume which is usually painless and leads to considerable esthetic and functional deformity. When this tumor arises in children, it is called the juvenile aggressive ossifying fibroma, which presents at an earlier age and is more aggressive clinically and more vascular at pathologic examination.⁷ The common clinical manifestations are proptosis, diplopia, epiphora, airway obstruction, facial swelling, epistaxis, headache, secondary sinusitis, meningitis and intracranial extension through the cribriform plate when arising from the sinonasal tract. Although ossifying fibroma is a slow growing tumor, different authors coincide in stating that the ossifying fibroma of the middle facial line and paranasal sinuses have a more aggressive behavior than those having a mandibular location.⁸ The ossifying fibroma that arises in infancy or early childhood tends to be aggressive and spreads rapidly. Our patient also had a giant lesion, which was involving the frontal bone, expansion of frontal bone leading to forehead swelling and proptosis. Short history was suggestive of aggressive behavior of lesion in our case. Most pathologists feel that cementifying fibromas and ossifying fibromas arise from the same progenitor cell but produce variable amounts of bone and cementum within any one lesion. From a histologic perspective, it contains a fibrous stroma made up of fusiform cells that are mixed with calcified trabecular and/or spherical foci that resemble, respectively, bone and cementum. The calcifications are extremely variable in appearance and represent various stages of bone and cementum deposition. Radiologically, ossifying fibroma appears as a well-circumscribed, solitary radiolucency with scattered radiopaque foci. It shows distinct stages during its development. In the early stages; the ossifying fibroma appears as a radiolucent lesion with no evidence of internal radiopacities. As the tumor matures, there is increasing calcification so that the radiolucent area becomes flecked with opacities until ultimately the lesion



Figs 1A to C: (A) Coronal (B) axial (C) and Sagittal CT scans showing expansile, heterogeneous density lesion in the left frontal bone



Fig. 2: Intraoperative photograph showing the bony lesion

appears as an extremely radiopaque mass. The lesion in our patient had mixed density mass with central radiopacities. One additional important diagnostic feature is that there is a centrifugal growth pattern rather than a linear one and therefore the lesion grows by expansion equally in all directions and expands the surrounding cortical bone without perforation. Surgery is the mainstay of treatment for ossifying fibroma, it usually shells out easily at surgery. Treatment of aggressive lesion requires radical surgical approach. Growth of the facial skeleton is an important consideration in the pediatric age group.⁹ Extensive tumors in this age group are more difficult to manage since the tumors here tend to be more invasive, aggressive and have more chances of recurrence. Mandibular tumors can be treated with conservative surgery but aggressive surgery is warranted for midface and paranasal sinuses because of their more aggressive behavior.¹⁰ Even though small tumors can be removed by simple curettage and enucleation, they are better avoided for the fear of recurrence especially in juvenile ossifying fibroma. Our patient was managed by endoscopic assisted open surgical approach. One study showed a recurrence rate of 28% after curettage.¹¹ Endoscopic excision has also been tried successfully and has the advantage of tumor excision under direct vision without any external scar.⁸ It could be used as adjuvant for other approaches especially when the tumor is extensive and has to be removed piecemeal. It is also useful to assess the surgical cavity for bleeding and tumor remnants after the excision of the tumor.¹² Since conservative techniques and piecemeal approaches could make histological interpretation more difficult, especially in cases of hybrid lesions, hence wherever possible, an open surgical technique is advocated for adequate visualization and complete excision.¹³ Larger lesions with intracranial extension may require craniofacial resection. In our patient the lesion was extensive, aggressive



Fig. 3: The excised surgical specimen (gross appearance)

in behavior and located in frontal bone, hence we excised the tumor by open surgical technique through Lynch Howarth approach and to ensure the complete excision in the cavity endoscope was used. Potential advantages of endoscopic techniques include enhanced visualization for preserving vital structures and determining tumor margins. Additionally, endoscopic approaches generally result in less collateral damage, offer superior cosmetic results and postoperative surveillance. There are still areas for future research to study the role of sole endoscopic surgery in ossifying fibroma treatment and needs to be defined by multicenter outcome studies. Recurrence after surgery is common and reported to be in the range of 30 to 56%.¹⁴ Treatment of recurrences by revision surgery is difficult again in the pediatric group. The overall prognosis of ossifying fibroma appears to be good. Despite their tendency for local invasion and recurrence, there are no reported instances of metastatic disease.

CONCLUSION

Ossifying fibroma of the frontal bone should be considered in the differential diagnosis when a child presents with proptosis. We believe that clinical awareness of this tumor, combined with further refinement of its CT features will lead to more accurate clinical diagnosis and more effective management of patients with juvenile ossifying fibromas in the frontal bone. Complete surgical excision of the tumor is possible when surgery is based on preplanned criteria. The radiological interpretation will be helpful for selecting appropriate surgical approach in growing child.

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