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Case Report
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Ossifying Fibroma of the Left Ethemoidal and Sphenoidal Sinus: A Case Report

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ABSTRACT

Ossifying fibroma (OF) is a rare benign lesions included in a heterogeneous group of bone disorders affecting the craniofacial bones. We would like to report a case of a 10-year-old female with a cemento-ossifying fibroma of the left sinonasal region, expanding into the orbit and causing proptosis of the left eye arising from sphenoid sinus. Complete surgical resection via an endoscopic nasal approach was performed after a preoperative work-up comprising head and neck computed tomography.

A review of the literature has revealed that treatment of this aggressive benign tumor must comprise of complete surgical resection since it is notorious for recurrence. Patients with juvenile ossifying fibroma tumours require long-term follow-up due to the high recurrence rate.

Keywords - *Ossifying fibroma; Sphenoidal sinus; Computed tomography.*

INTRODUCTION

Ossifying fibroma (OF) is a rare benign fibro-osseous tumor, located mainly in the head and neck region.[1] Seldom involving the maxillary, the tumor occurs with a unilateral centrifugal growth pattern, evolving slowly and asymptomatically, therefore, consultation and discovery can be delayed unless it causes an aesthetic or functional problem. OF is divided into the conventional (classical) form which frequently targets the mandible[2], is female predominant with peak incidence in the second and third decades of life,[3,4] whilst the juvenile form (JOF) typically occurs in children and young adults with an equal sex distribution, occurring principally in the extragnathic craniofacial bones, particularly the paranasal sinus and periorbital bones, with the tendency for maxillary involvement.[5] It is an aggressive variant with bone destruction and tumor recurrence. Histopathologically, it is not encapsulated, although it is well demarcated from the surrounding bone[6] and can be further divided into two variants: psammamatous and trabecular pattern.[7,8]

The term "cemento-ossifying" is a generic term used synonymously with (central) ossifying fibroma (of the head and neck) related to histological compositions of OF with calcified tissue resembling bone and/or cementum in addition to fibrous tissue.[10,11] Presumptive diagnosis is established by clinical examination and radiological data, however the definitive diagnosis is founded on histopathological examination (psammomatous bodies, osteoblastic rim, trabecular bone).

Case Report

We report the case of a 10-year old girl who experienced painless, progressive facial swelling, nasal obstruction and proptosis of the left eye, resulting in deformation of the left side of the face in the maxillary region since three months of age. She had no family history or past medical history of OF. Clinical examination revealed a hard non-tender nasal mass covered with normal mucosa. Ophthalmological examination revealed non axial proptosis with no signs of oculomotor paralysis and no loss of visual acuity or visual field abnormalities (Figure1).

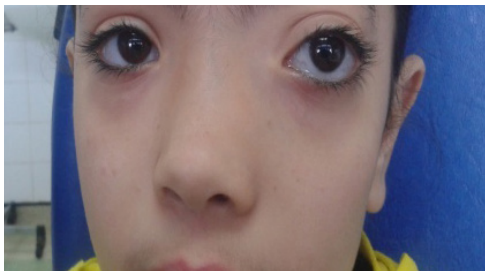

Figure 1: Left facial deformity, proptosis of left eye

The computed tomography scan of the nose and sinuses showed an expansile lesion with ground glass matrix with a less dense fibrous centre of the tumor in left sinonasal region, with regular shape that contained microscopic calcifications, extending into the ipsilateral lamina papyrasea and orbital floor which was pushing the surrounding soft tissues forward and laterally without invading them.

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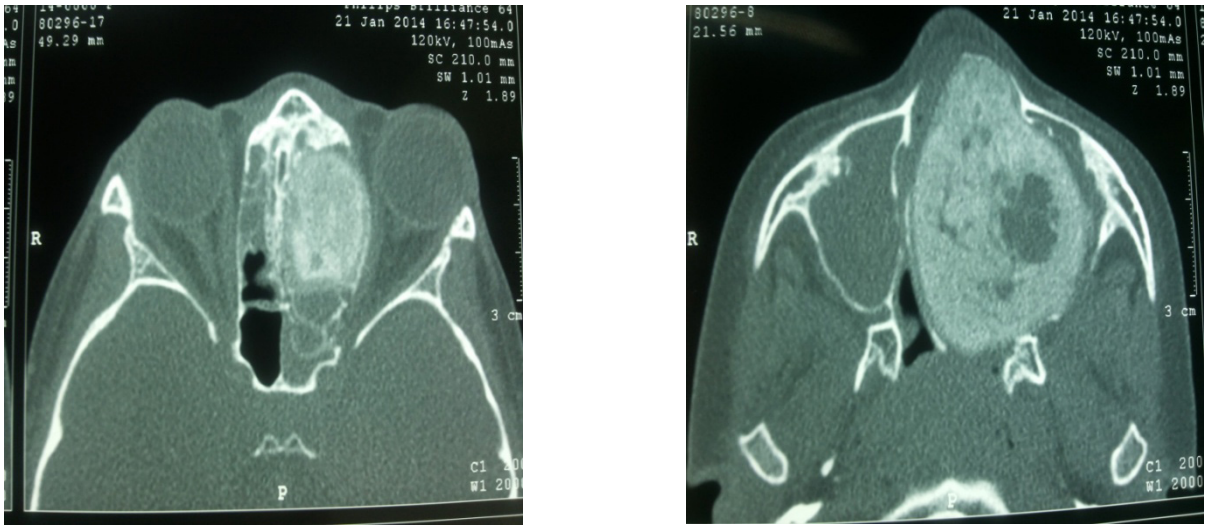
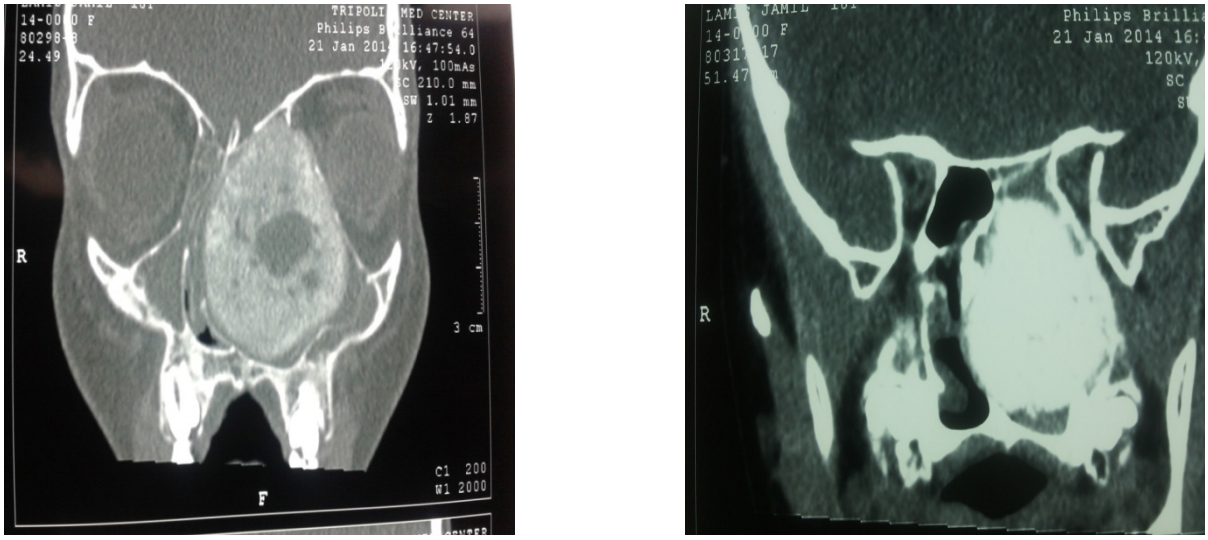


(a) **(b)**

Figure2 a,b: Show axial CT of the head demonstrating an ossifying fibroma of the left maxilla.



(a) **(b)**

Figure 4, 5: Coronal CT of the head demonstrating an ossifying fibroma of the left ethemoidal and sphenoidal sinus with destructing sinus floor invading nasal cavity - soft tissue window setting.

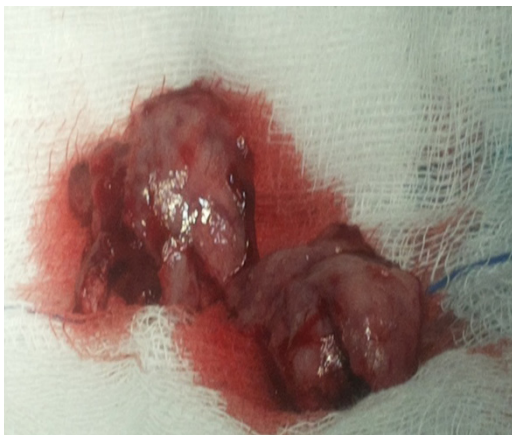


Figure 4: Tissue removed from left nasal cavity

A differential diagnosis:

Fibrous dysplasia; osteofibrous dysplasia; osteoblastoma; low-grade osteosarcoma and chronic osteomyelitis.

The histopathological examination of the tumor confirmed the diagnosis of cemento-ossifying fibroma. Microscopic examination of tissue showed benign spindle stromal cells arranged in a storiform pattern with discrete round masses of cementum-like material.

Total resection was performed on an endoscopic nasal approach. The excised tissue was dome shaped and firm in consistency measuring 4cm × 4cm × 3cm (Figure 4) with an irregular surface. The patient was free of clinical or radiological follow-up imaging performed nine months after the operation recurrence at follow-up.

DISCUSSION

Ossifying fibroma is one of the benign fibro-osseous lesions (BFOL). The term (BFOL) is subsequently used in the literature to describe a spectrum of lesions ranging from fibrous dysplasia to ossifying fibroma. Common to all BFOL is the replacement of normal bone with a tissue composed of collagen fibers and fibroblasts that contain varying amounts of mineralized substance. The etiology and pathogenesis of OF is still unknown. The proposed theories of trauma and developmental abnormality as a cause of OF had been suggested[14,15], involving the craniofacial bones, first reported by Menzel in 1872[16,17] and described as a clinical entity by Montgomery in 1927.[5,16]

Based on literature review, the most common site is the mandible accounting for 75-89% of cases[18], followed by maxilla and, less commonly, paranasal sinuses.

Clinical presentation of these tumors is variable, depending on the site and rate of growth. When encountered in the calvaium, the presentation is typically with bone expansion, usually painless. However, several cases reports[1,10,14,19,20] Midline skull base lesions, and may present with symptoms due to mass effect, such as nasal obstruction, headache or anosmia or ocular symptoms including visual loss, diplopia, and exophthalmos. There have been no reports of malignant transformation of this benign bone tumor, although some case reports describe local aggressiveness with extensive, rapid growth that may mimic malignancy and associated with neurological signs due to an intracranial mass effect.[14,15] Radiographic features of OF vary from case to case. Most authors described the usual radiological characteristics of OF as a well-defined lesion[3,14-17] surrounded by a uniformly radiolucent periphery or occasionally by peripheral sclerosis.[17]Treatment of sinus OF is surgical, aiming at complete resection.[5,16] Recurrence ranges from 1% to 63%.[1] Adjuvant radiation therapy is contraindicated due to risk of malignant transformation[3] and lack of proven efficacy. Spontaneous malignant transformation is rare but has been reported[18], whereas metastasis has not.

CONCLUSION

Sinonasal ossifying fibroma is a rare benign tumor. Treatment is surgical and preferably functional endoscopic sinus surgery, indicated for symptomatic lesions or ophthalmic or infections complications. Long-term clinical and radiological surveillance is required due to high incidence of recurrence.

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