Pakistan Journal of Otolaryngology 2024: 35(1):32-34
DOI 10.70224/pjoh.1.1.2024.10

CASE REPORT

Juvenile Primary Ossifying Fibroma of Maxillary Sinus: A Case Report and Review of the Literature

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ABSTRACT

Fibro-osseous lesions (FOLs) are a group of benign bone disorders, defined by the replacement of functional bone by fibrous connective tissue matrices. Our case report delves into the presentation of a specific FOL, namely the juvenile ossifying fibroma (JOF). This disease's rare occurrence in a clinical setup makes documentation of such presentations vital to otorhinolaryngologists across the globe. Our article highlights the presentation of such a case of JOF in a 15-year-old male child from Pakistan. We discuss differentials, our process towards reaching a diagnosis via biopsy, and the disease's resolution via surgical excision. Due to JOF mimicking a plethora of similar otorhinolaryngology-adjacent diseases, our article wishes to provide insight with regards to approaching patients with relevant presenting complaints, eliminating differentials, and deciding on a particular treatment regimen.

INTRODUCTION: Fibro-osseous lesions (FOLs) are a group of benign bone disorders characterized by the replacement of normal bone by a fibrous connective tissue matrix with varying degrees of mineralization[1]. These lesions include fibrous dysplasia, ossifying fibroma, and cemento-osseous dysplasia. They share some histologic characteristics and are differentiated based on their growth on radiographical presentation[2]. The ossifying fibroma (OF) lesion is a rare, benign, slow growing tumor originating from the periodontal ligament[3] that commonly affects the craniofacial bones, particularly the mandible and maxilla. OF is characterized by the formation of well-circumscribed, mostly radiolucent lesions with radiopaque centers depending on the degree of tissue calcification[4]. It can occur in both juveniles and adults, with a predisposition for females[5]. The juvenile variant is more aggressive and tends to occur in patients under 15 years of age and is responsible for 2% of oral cancers in children[6]. Juvenile ossifying fibroma (JOF) has a propensity to occur in the maxillofacial region, particularly in the jawbones and the paranasal sinuses. Among the paranasal sinuses, the maxillary sinus is the most common site. A cellular fibrous stroma, garland-like bone strands, and cementum particles are histological features of JOF[7]. The diagnosis of JOF can be challenging, but by correlating clinical, linear radiography, CT scan and histopathological results we can make a definitive diagnosis[7,8].

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Case Report: A 15 years old male presented to our Otorhinolaryngology Head and Surgery Department via the OPD with the complaint of Right sided facial swelling since the past one year. The swelling was reportedly initially small in size, but at the time of presentation covered the entirety of our patient's right lower face. The swelling was associated with episodes of epistaxis, nasal congestion and obstruction as well. In addition, sense of smell was absent and the patient also complained of headache, right sided facial pain and numbness and watering in the right eye.

On examination there was a 5x5 cm swelling on the right side of the face causing facial asymmetry with the telecanthus and right eye proptosis (Figure-1) There was a slight discoloration of the skin just below the right orbit with no palpable cervical lymphadenopathy. The nasal patency was decreased in both nostrils with absent sense of smell. A bony, hard enlargement was seen during an intraoral exam in the left maxillary premolar-molar area. The buccal and palatal cortical plates somewhat expanded as the swelling spread from the buccal vestibule to the palatal aspect. Overlying the swelling, the oral mucosa seemed healthy and free of ulcers. The teeth next to it showed no evidence of dental caries or periodontal disease. However, there was malocclusion of upper and lower jaw teeth and a bulge in the right side of the hard palate.

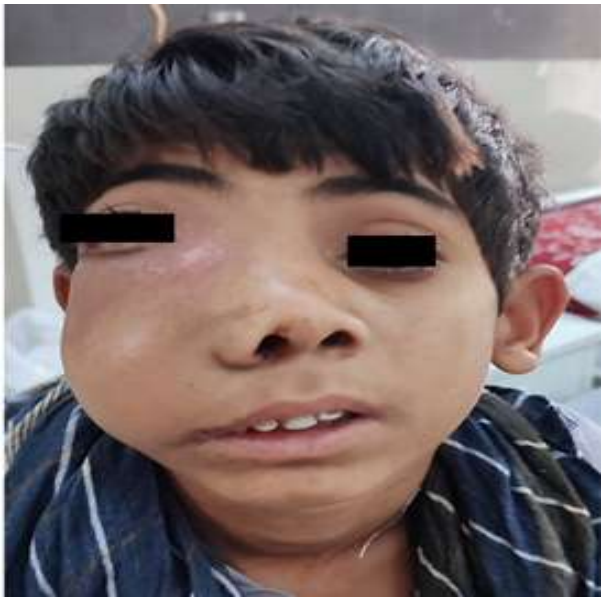

Figure 1: Clinical Photograph Showing Right Sided Facial Swelling With Telecanthus

cavity. In addition, there was an extension to orbit with right eye proptosis. (Figure-2)

From the lesion, an incisional biopsy was collected, and the pathology process was recorded as JOF. The diagnosis was made based on the presence of fibrillar osteoid trabeculae and woven fragments of bone. The entire tumor was excised in Toto as a single mass that is clearly delineated and lobulated with conservative surgical excision. After the lesion was removed, the patient underwent clinical and radiological follow-up to rule out any potential relapses. The patient was also given temporary removable prosthesis advice in the interim.

DISCUSSION: The documentation of cases involving juvenile ossifying fibroma is an important taking, both from a purely academic perspective and, more particularly, for an ENT practitioner's benefit. Addressing the former, since JOF is a disease that is rarely encountered clinically, identifying it may prove

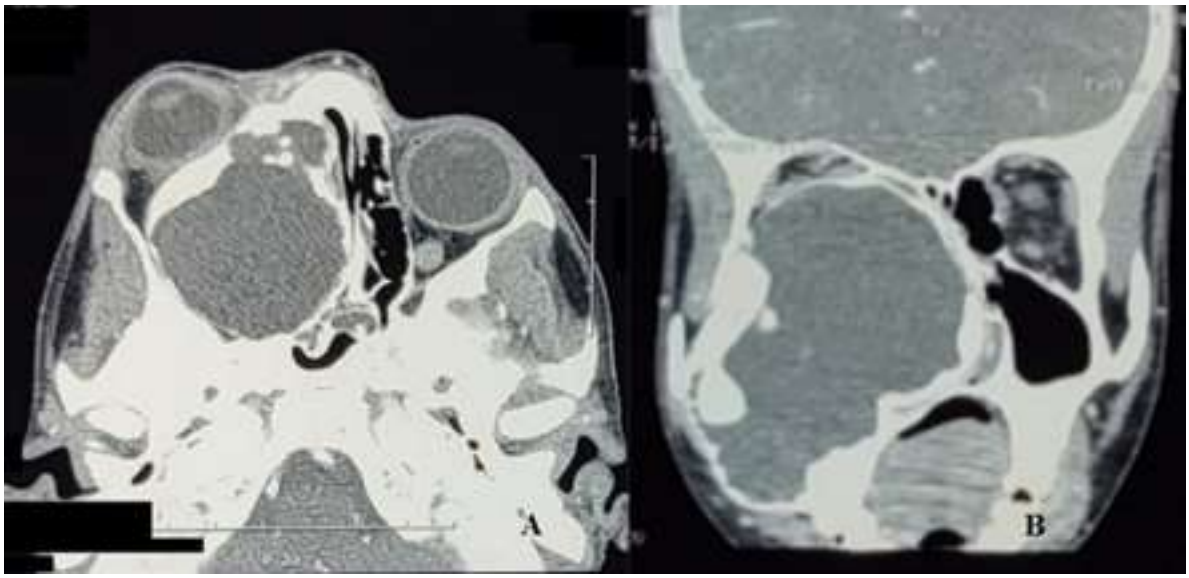

Figure 2: CT Scan of Nose and Paranasal Sinuses Showing (A) Cystic Lesion in the Right Maxillary Sinus With Intra Nasal Extension (B) Expansion of the Lesion in to the Obit With Right Eye Proptosis

As part of investigative proceedings, a CT scan of the paranasal sinuses was conducted with contrast. Findings revealed the presence of a large, expansile cystic lesion, which was seen arising from the maxillary bone on the right side. The lesion showed internal hyperdense areas and measured approximately 7.4x5.8x8.5 cm (craniocaudal x anteroposterior x transverse) and is associated with obliteration of the right maxillary sinus, intranasal extension, and obliteration of the right nasal

challenging to physicians. This dilemma is further compounded upon by the elusive nature that JOF as a disease inherently possesses. Many cases of JOF remain clinically silent, and are only identified via incidental clinical findings, such as dental X-rays[9]. Patients also present with a variety of non-specific symptoms, which provide an array of differentials, further clouding a clear and efficient diagnosis. Unfortunately, a quick diagnosis is imperative to a positive patient's clinical

outcome, as the neoplasm's aggressive progression can lead to higher rates of mortality.

From an ENT physician's perspective, identifying JOF early is not only important to impede the disease's own pathogenesis, but also to prevent the occurrences of other metastatic disease. 2% of all JOF cases are known to progress towards oral cancers, making for a complicated clinical course for patients[6]. The need for multiple surgeries not only puts patients under physical peril and mental stress, but also contributes towards a decrease in compliance. At any rate, prior research into JOF has provided future surgeons with a set minimum of parameters needed to either identify the disease, or at least have it taken into consideration as a differential. JOF patients will present as individuals under the age of 15, with tumors typically involving the maxilla, paranasal sinuses, the orbit, the frontoethmoid complex and, to a lesser degree, the mandible[9].

CONCLUSION: Our patient's presentation with a notable swelling, distorting facial features and associated with symptoms of nasal congestion, gave rise to suspicion of many other conditions before JOF, with fungal invasion of the sinuses being one such differential. Ultimately, however, a heavy emphasis on confirming differentials via imaging techniques and biopsy proved to guide our approach towards an unconventional disease. JOF may not be the most common of otorhinolaryngological diseases, but we postulate that keeping an open mind and trusting a more explorative process led to a quicker resolution for our patient.

The identification of JOF, and that too at as early a stage as possible, remains imperative for any and all successful treatment regimens. The disease is known to progress towards more insidious metastatic conditions, which only further complicate an effective surgical outcome. Therefore, keeping in mind the few tenets of a patient presenting with JOF (i.e., an individual under the age of 15 presenting with tumors involving the maxilla or paranasal sinuses), relying on CT imaging for any such presenting complaints, and ruling out other diseases in a fast and efficient manner will remain the cornerstones of improving patient outcome.

Declaration: Conflict of interest: Authors declare no conflict of interest.

Funding disclosure: Nil

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