



Juvenile ossifying fibroma of the maxilla in a 6-year-old male: case report

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Fibro-osseous lesions encompass a category of histopathology characterized by the presence of a fibrous stroma with varying amounts of mineralized material resembling bone or cementum.^{1,2} One such fibro-osseous lesion is the rarely occurring juvenile ossifying fibroma (JOF).¹ No clear-cut criteria exist for separating this tumor from the more common forms of ossifying fibroma; however, these juvenile tumors behave differently from the common ossifying fibroma.¹ The tumor can grow rapidly, expanding in the affected area; thus they are also referred to as "active" or "aggressive" ossifying fibromas.^{1,3} JOF usually occurs between the ages of 5 and 15 years, although cases in older patients have been reported.¹⁻⁵ Usually JOF involves the maxilla, paranasal sinuses, orbital and frontoethmoid bones. It is believed that the majority of these tumors arise in the paranasal sinuses. As the JOF expands it may involve the orbital, nasal, and cranial cavities. Mandibular lesions have been reported.^{1,3}

Radiographic features are highly variable depending on the amount of calcification. Varying degrees of radiolucency and opacity are commonly seen, along with fair demarcation of the tumor. Erosion and invasion of the surrounding bone may be noted in more aggressive tumors.² The JOF is a neoplasm composed of polyhedral and spindle cells in a relatively sparse collagen stroma. In addition, immature cellular osteoid forms within the lesion and multinucleated giant cells may be present, and sometimes may be very numerous. As the lesion matures, the bone formation may take on a more woven appearance.^{2, 4, 5} This case report details



Fig 1. 6-year-old male presents with left side maxillary swelling.

the diagnosis and management of JOF that resulted in significant facial destruction in a 6-year-old child.

Case report

History and chief complaint

A 6-year-old black male came to the emergency room of the Medical University of South Carolina Hospital with facial swelling of the left maxilla. His mother reported that she had noticed a slight swelling of the area 3 months earlier, but over the 2 weeks prior to visiting the MUSC Hospital she had noticed an increase in the size of the swelling. There was no history of pain. The child's past medical history included hyperactivity currently treated with Ritalin™ (CIBA Pharmaceutical Co, Summit, NJ) and impetigo successfully treated with penicillin 3 months earlier.

The clinical exam, performed by emergency room pediatricians, revealed left maxillary swelling, no pain or tenderness on palpation, normal temperature, and moderate nontender lymphadenopathy in the anterior cervical region. A preliminary diagnosis of gingivitis was made by the pediatricians without dental consultation. The child was placed on penicillin VK 250 mg

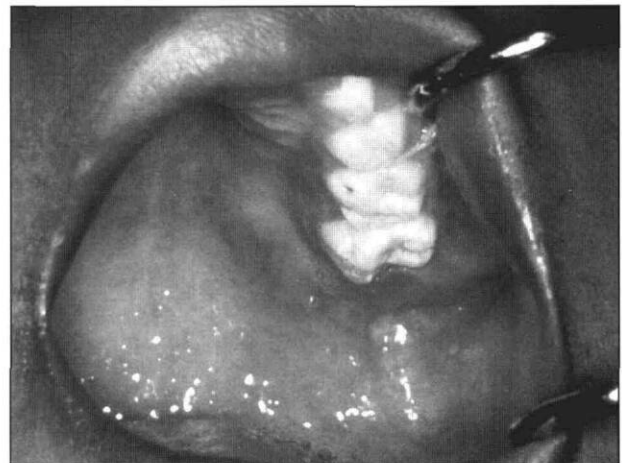


Fig 2. Intraoral view exhibiting buccal and medial expansion.

qid and given a follow-up appointment for a week later.

The child returned as scheduled with no appreciable decrease in the size of the swelling, which was recorded as indurated and nontender. CBC and sedimentation rate were within normal limits except for elevated MCV 86.9 (normal range 75.0–85.0), MCH 30.0 (normal range 24.0–28.0), segmented white cells 35 (normal range 40–60), and lymphocytes 57 (normal range 20–50). These lab values were interpreted as inconclusive by the medical staff and the patient was referred to the pediatric dentistry department for routine consultation.

Clinical examination by a pediatric dentist 2 days later revealed noticeable extraoral swelling of the left maxillary/midface area (Fig 1). A left side maxillary swelling adjacent to the posterior teeth was noted primarily in the buccal vestibule, but extended posteriorly toward and encompassed the tuberosity (Fig 2). The swelling was diffuse, indurated, nontender, and approximately 3 cm in diameter clinically. The maxillary

posterior teeth exhibited class III mobility. No gingival inflammation, caries, or previous restorations were noted in the region. The teeth were not sensitive to heat, cold, or percussion. The periodontal exam was unremarkable for bleeding or pockets. The neck exam revealed bilateral moderate anterior cervical lymph nodes, which were nontender and slightly mobile. The child reported no history of pain or discomfort.

A panoramic radiograph revealed a diffuse expansion of the left maxilla, approximately 4 cm in diameter. The expansile area had a radiographic consistency similar to alveolar bone and extended into the sinus superiorly, displacing the maxillary left permanent second molar into the left maxillary sinus (Fig 3). In addition, there was marked expansion distal to the tuberosity. Periapical and bite-wing radiographs revealed no evidence of decay or periapical pathology associated with the posterior teeth in that quadrant.

The differential diagnosis initially included primary

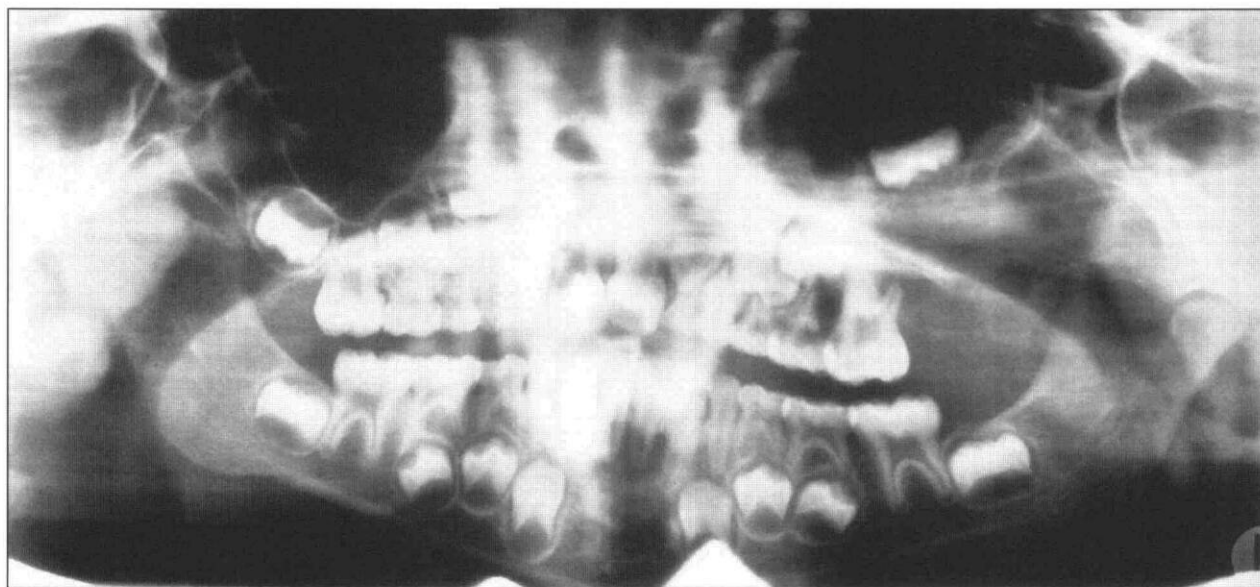


Fig 3. Panoramic radiograph displays superior repositioning of tooth #15 nearly to the orbital floor.

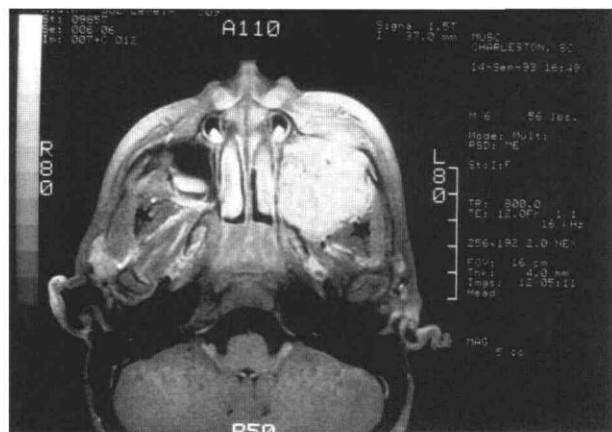


Fig 4. MRI showing mass nearly filling the left maxillary sinus.

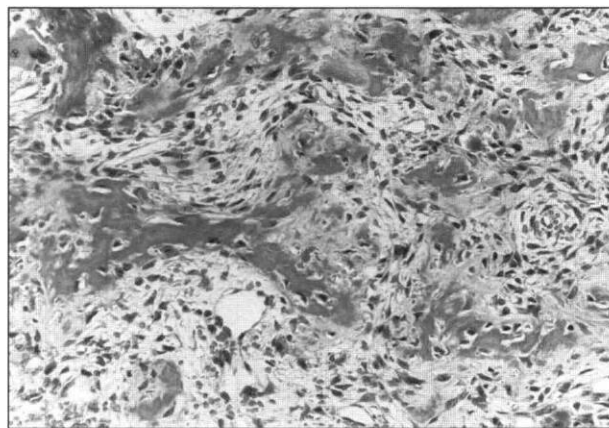


Fig 5. High-power photomicrograph showing irregular spicules of immature bone within a stroma of spindle-shaped cells. (Hematoxylin & eosin stain, original magnification X200)

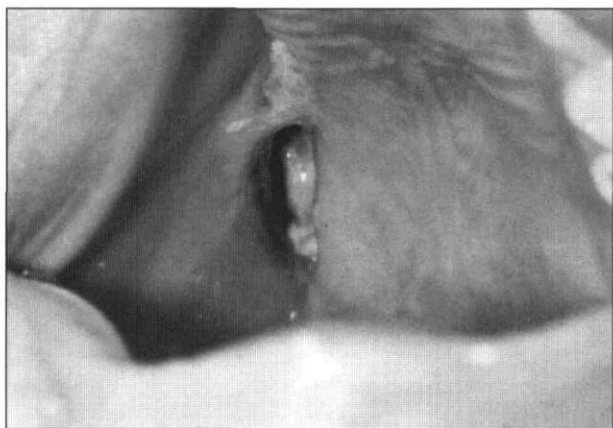


Fig 6. Intraoral view 3 months postoperative showing oral-antral fistula.

tumors of the bone such as osteoblastoma and JOF. In addition, the possibility of a locally aggressive odontogenic lesion such as an ameloblastic fibroma or a calcifying odontogenic cyst was considered — although deemed less likely.

An MRI confirmed an expansile mass involving the left maxilla (Fig 4). The mass nearly filled the left maxillary sinus, extending upward through the orbital floor, expanding the sinus laterally against the masticator muscles and nearly contacting the coronoid process. Expansion of the lesion was observed medially to the medial aspect of the maxillary sinus. The mass extended anteriorly through the anterior wall of the sinus and involved the alveolar ridge to the level of the left maxillary canine. Posteriorly, the tumor perforated the posterior antral wall. There was no evidence of an infiltrative pattern and the mass maintained relatively sharp margins.

One week later an incisional biopsy determined the lesion to be a juvenile ossifying fibroma. Microscopically, the tumor appeared as a matrix of spindle cells with some collagen, showing scattered early osteoid production with scattered collections of multinucleated giant cells (Fig 5). The tumor was subsequently removed *in toto* via a modified Weber-Ferguson approach by the department of otorhinolaryngology and oculoplastic surgery. A prosthesis to cover the intraoral defect was completed by maxillofacial prosthodontics.

Discussion

The tumor discussed in this case report exhibited the features typical for JOF as previously described in the literature.^{1,2,5,6} This particular case resulted in a significant facial defect due to the large size of the lesion. Clinical management and prognosis of JOF varies according to clinical presentation. Some lesions may be slow in formation and nonaggressive, while others, particularly in younger children, may be quite destructive and aggressive.¹ Management has most frequently been conservative local excision and curettage, although some cases have been treated with more radical resection. The aggressive nature in this particular case with involvement of the maxillary sinus to the

orbital floor and antral wall necessitated *in toto* removal. Due to the sparsity of JOF articles, controlled studies to evaluate conservative versus more aggressive forms of treatment have not been documented. Recurrence rates between 30 and 58% have been reported; therefore, close follow-up is required.^{1,3,4,6} Malignant transformation has not been shown.¹

A retrospective analysis of the history and clinical presentation suggests that the diagnosis may have been achieved sooner under different circumstances. The child had never been to a dentist before the referral from pediatrics. A radiographic survey, in addition to a thorough orofacial exam, likely would have revealed the tumor much sooner if the child had been under the routine care of a dentist. The child's mother stated that she had noticed the swelling several months before presentation to the emergency room; unfortunately, she didn't seek dental or medical care earlier. In addition, the initial emergency room presentation, which resulted in a diagnosis of gingivitis, clearly shows the importance of a close working relationship between the medical and dental communities. The absence of inflammation and bleeding on probing, coupled with the history and presentation of a nontender indurated swelling likely would have led a dentist to rule out an inflammatory process such as gingivitis. A dental referral at the time of presentation would have likely resulted in an appropriate oral and radiographic evaluation with resultant proper diagnosis.

The child is doing well and is being periodically evaluated by otorhinolaryngology. He is scheduled to undergo a soft tissue graft to close a postsurgical oral-antral fistula (Fig 6). In addition, the child now receives regular dental care in the pediatric dental clinic.

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