

Juvenile Psammomatoid Ossifying Fibroma – A Case Report

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Abstract:

Juvenile ossifying fibroma (JOF) is a rare fibro-osseous neoplasm in young children. JOF is defined as a variant of the ossifying fibroma, and latter includes juvenile psammomatoid ossifying fibroma (JPOF) and juvenile trabecular ossifying fibroma (JTOF). Both variants involve the craniofacial bones with the trabecular variant being more common in the jaws and the psammomatoid variant being more common in the paranasal sinuses. This lesion is locally aggressive and spreads quickly. As it has a very high recurrence rate, complete excision is essential. A case of large JPOF involving right maxilla and other cranial bones in a 12 year old female patient with clinical, radiographic and histopathological features are presented. Surgical management and follow up is also emphasized.

Key Words: Fibroma, Ossifying; Juvenile.

Introduction:

The juvenile ossifying fibroma is also known as “Aggressive ossifying fibroma or Active ossifying fibroma” (Neville et al, 2002). It has been distinguished from the larger group of ossifying fibroma on the basis of age of occurrence, most common site of involvement and clinical behavior (Granados et al, 2002). Most of them affect extra-gnathic bones; some affect maxilla and craniofacial bones but rarely mandible (Lawton et al, 2002). Juvenile ossifying fibroma was first described by Benjamins in 1938 as “osteoid fibroma with atypical calcification.” (Khoury et al, 2002). Later on in 1952 Johnson coined the term “juvenile active ossifying fibroma” (Neville et. al 2002). Histologically, It has two distinct microscopic patterns; trabecular and psammomatoid. It is an aggressive lesion mimicking malignancy such as osteosarcoma. So, it is important to accurately recognize JOF for making the diagnosis and managing the disease (Park et al, 2007).

Case report:

A 12 year old girl reported to department of Oral Medicine & Radiology, Guru Nanak Dev Dental College & Research Institute, Sunam with complaint of swelling on right side of face for last 3 years. History revealed a small painless swelling first appeared at the age of 6 years which gradually increased in size within the span of 2 years. She underwent surgery for that

and remained asymptomatic for one year. Swelling recurred again at same site and gradually increased within the span of 3 years to attain the present size. There was history of difficulty in breathing, swallowing and watering from right eye. Extraoral examination (Fig. I & II) revealed gross facial asymmetry on the right



Fig.I: Extraoral photograph (frontal view) showing facial asymmetry due to swelling

middle 1/3rd of face. A solitary, roughly oval in shape and well defined swelling measuring 8 × 9 cm in dimension was noticed which was extending superiorly from infraorbital ridge to inferiorly about 2cm above the lower border of mandible, medially starting from left ala of nose crossing the mid sagittal line and extending laterally 3cm in front of right tragus. Overlying skin was stretched with focal areas of hyperpigmentation.

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Fig.II: Extraoral photograph (lateral view) showing swelling in right middle 1/3 of face.

There was deviation of nose towards left side, right eye was displaced upward and laterally, drooping of right commissure of mouth with obliteration of inferior orbital and nasolabial fold. On palpation, swelling was non tender, hard in consistency with egg shell crackling was felt. There was no local rise in temperature.

Intraoral examination revealed a solitary well-defined swelling obliterating the right buccal vestibule involving major portion of the hard palate except on left lateral margin (Fig. III). The swelling was 2 mm away from palatal gingival margin of maxillary anterior teeth and extending posteriorly up to the junction of hard and soft palate. Laterally extending from right buccal vestibule, crossing the alveolar ridge and midpalatal raphae, and extending 1.5 cm in front of palate-gingival margin of left maxillary posterior teeth medially. Overlying mucosa was stretched. Swelling was non-tender and hard in consistency with egg-shell crackling in the center. Hard tissue examination revealed patient was in mixed dentition stage. Teeth 12 and 16 were clinically missing and multiple retained primary teeth were present.

Provisional diagnosis of Tumour of Right Maxilla with differential diagnosis of Fibrous dysplasia, Cementoossifying fibroma, Juvenile ossifying fibroma, Ameloblastoma, Odontogenic keratocyst, Osteosarcoma and peripheral giant cell granuloma were



Fig. III: Intraoral photograph showing palatal swelling.

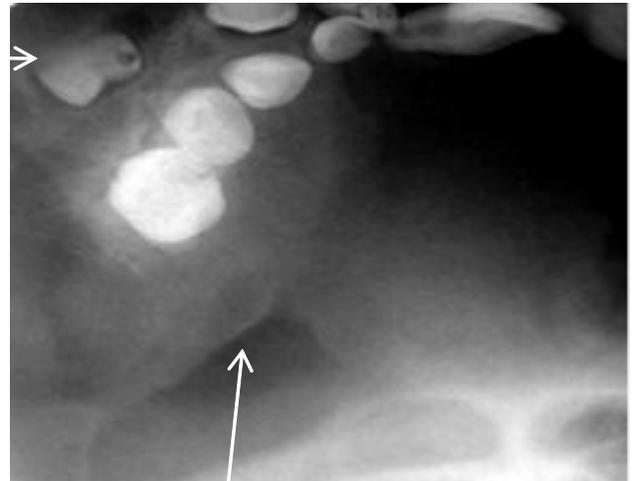


Fig. IV: Maxillary occlusal radiograph showing cortical expansion with radiopaque internal mass & buccal displacement of 12,13,14 (arrows).

considered. All teeth in the vicinity of lesion were vital. Haematological and serological investigations were within normal limits except elevated alkaline phosphatase level (244 IU/l).

Maxillary occlusal cross-sectional radiograph (Fig.IV) revealed dense homogeneous well demarcated radiopacity with focal areas of radiolucency on right side with expansion of buccal cortex and palatal bone extending from right premaxilla to beyond the right maxillary tuberosity. Teeth 12,13, 14 were buccally placed.

Panoramic radiograph (Fig. V) revealed that on right side homogeneous radiopacity involving maxilla encroaching maxillary sinus, zygomatic bone, nasal cavity, pterygoid plates and downward slanting of occlusal plane with drifting of maxillary teeth of right side.



Fig.V: Panoramic radiograph showing diffuse radiopacity at right maxilla obliterating maxillary sinus (arrow).

Paranasal sinus view (Fig. VI) also revealed homogeneous radiopacity medially crossing midline, superiorly extending about 0.5 cm above right infra orbital rim and Inferiorly approaching the alveolar process of maxilla.



Fig. VI: Para-nasal sinus view showing large radiopaque mass obliterating maxillary sinus & surrounding areas.

CT scan (Fig.VII & VIII) showed grossly expanded right maxillary sinus with distorted contours and marked thinning of all walls and expansile mixed attenuation mass with well demarcated outline filling the right maxillary sinus, frontal sinus, ethmoidal sinus, right nasal cavity medially crosses midline, superiorly extending toward inferior orbital ridge, Inferiorly encroaching the palate on right side and extending toward the sphenoid sinus. Incisional biopsy of the lesion was taken and sent for histopathological examination (Fig. XI) which

revealed evidence of cellular proliferation of bland appearing spindled cells of apparent fibroblastic nature with prominent associated cementum-like calcifications (psammoma bodies).

Final diagnosis of juvenile psammomatoid ossifying fibroma was made. Surgical excision of lesion was carried out. Patient was recalled regularly every 3 months interval. No evidence of recurrence was seen even after one year. (Fig. X)



Fig. VII: CT Coronal view showing expansile mixed attenuation mass with grossly expanded maxillary sinus (arrow).

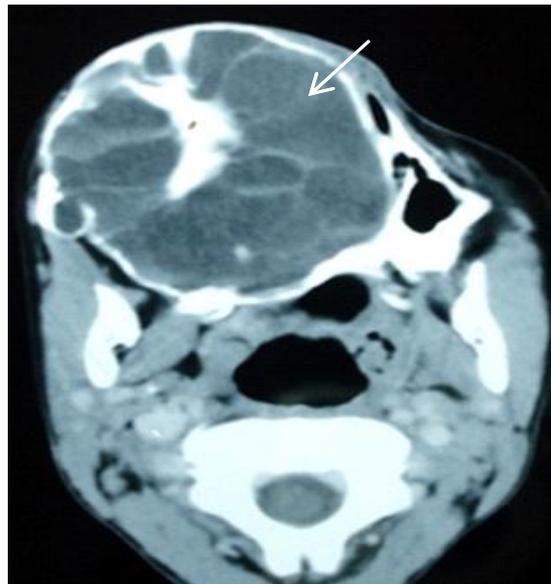


Fig. VIII: CT Axial view showing expansile mixed attenuation mass (arrow).

Discussion

The fibro-osseous lesions are those in which the normal bone architecture is replaced by fibroblasts and collagen fibres that contain various amounts of

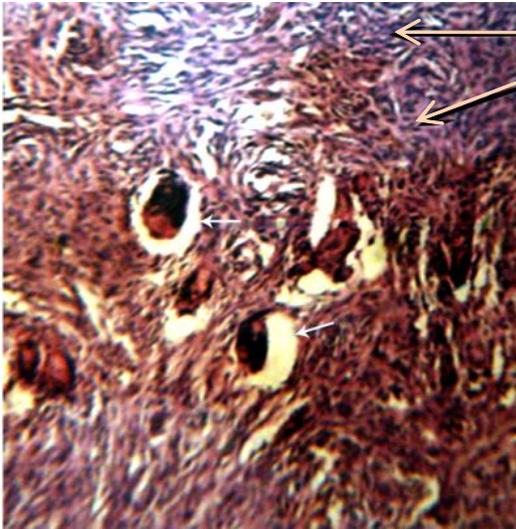


Fig. IX : Photo micrograph showing proliferation of fibroblasts in a densely arranged stroma (arrows-orange) with prominent associated cementum-like calcifications i.e. psammoma bodies (arrows), (40X, H&E).



Fig. X: Post-surgical photograph showing significant recovery of facial esthetics.

mineralized material. Juvenile psammomatoid ossifying fibroma is one of the types of fibro-osseous lesion. The main characteristics are: patient under 15 years of age, the location of the tumour, the radiologic pattern and tendency to recur. The orbit and paranasal sinuses are the most common site, accounting for over 72% of reported cases, followed by the calvarium 11%, maxilla 10%, and mandible 7%. In reviews published by Hamner et al and Sloop et al. (Saiz-Pardo-Pinos et al, 2004), the mean age of onset was 11.5 and 11.8 years. Most cases of maxillary JOF are asymptomatic. In large lesion, the clinical manifestations are swelling of the maxilla, exophthalmos, bulbar displacement nasal

obstruction, root resorption and displacement of teeth in the tooth-bearing region. The radiologic features show varying degrees of radiodensity. On CT, aggressively progressing ossifying fibroma appears as expansile mass covered by a thick shell of bone density with a multiloculated internal appearance and a content of varying density (Sarwar et al, 2008). Histologically, Psammomatoid type of JOF shows highly cellular fibrous stroma often with whorled pattern containing closely packed spherical ossicles resembling psammoma bodies. The treatment for JOF is conservative excision or curettage; some lesions may necessitate more aggressive management. Because the recurrence rate for JOF ranges from 30% to 58%, continued follow-up is essential. Despite the aggressive nature of the lesion and high rate of recurrence, malignant transformation to sarcoma has not been reported (Sun et al, 2007).

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