Annals of Clinical Case Reports

Case Report
Published: 08 May, 2017

Gnathic Psammomatoid Ossifying Fibroma: Exceptional Cases

Praveen BN, Shubhasini AR, Shubha G, Keerthi G, Shiladitya Sil and Swathi Mani*

Department of Oral Medicine and Radiology, K L E Society's Institute of Dental Sciences, Bangalore, India

Abstract

Juvenile Ossifying Fibroma (JOF) is a rare fibro-osseous neoplasm of the craniofacial bones, having locally aggressive behaviour and recurrence. The psammomatoid juvenile ossifying fibroma (PsJOF) is a variant of JOFs, occurring as extragnathic lesions in the sino-nasal and orbital bones. PsJOF is rarely seen in the jaw bones, with slight male predominance and mean age of occurrence between 16-33 years. PsJOF are central lesions manifesting radio-graphically as a radiolucent, mixed or radiopaque lesion which depends on the degree of calcification giving a ground glass appearance. PsJOF is histologically characterised by the presence of psammoma bodies which are concentric, lamellated calcified structures. We hereby report two unique cases of PsJOF, one occurring as a combined lesion, in association with Aneurysmal Bone Cyst in the maxilla and another case PsJOF presenting as soft tissue growth in an elderly female patient.

Keywords: Fibro-osseous lesions; Juvenile ossifying Fibroma; Psammoma bodies; Aneurismal bone cyst

Introduction

The World Health Organization (WHO) in 2005 defined Ossifying Fibroma (OF) as "a well-demarcated lesion composed of fibro-cellular tissue and mineralized material of varying appearances." [1] OF is subdivided into conventional and juvenile forms. Juvenile Ossifying Fibroma (JOF) is a rare fibro-osseous neoplasm of the craniofacial bones having locally aggressive behaviour and recurrence [2,3]. The most characteristic feature of JOF is its occurrence in children and young adults [4].

JOF is further distinguished into Trabecular Juvenile Ossifying Fibroma (TrJOF) and Psammomatoid Juvenile Ossifying Fibroma (PsJOF) [4]. TrJOF are gnathic lesions affecting the jaws with predilection for the maxilla, with the average age of occurrence between 8.5-12 years [5,6]. Whereas, PsJOF are extragnathic lesions occurring in the sino-nasal and orbital bones [6], usually occurring between 16-33 years [4,7]. PsJOF is distinguished from the other fibro-osseous lesions by its age of onset, clinical presentation, aggressive behaviour and high recurrence rate. PsJOF is histologically characterised by the presence of psammoma bodies which are concentric, lamellated calcified structures [6,8].

We hereby report two cases of PsJOF which rendered a diagnostic challenge because of their varied clinical presentation, unusual site and age of occurrence.

Case Presentation

Case 1

A 17 year old male patient reported with the complaint of swelling on the left side of the face since 5 years. The swelling was gradual in onset, slowly progressing and was not associated with pain, discomfort, difficulty in breathing, vision, speech or chewing food.

On examination (Figure 1 a and b) a solitary well defined swelling on the left middle one-third region of the face was appreciated measuring approximately 4 cm x3 cm, extending antero-posteriorly from the ala of the nose up to 3 cm short of the tragus of the ear; superior-inferiorly from the infraorbital margin to the ala-tragal line. Skin over the swelling appeared stretched with no change in colour; the surrounding skin appeared normal. The swelling was hard and non-tender, on palpation.

On intraoral examination (Figure 1c) solitary well defined swelling was seen in the left buccal

OPEN ACCESS

Correspondence:
Praveen BN, Department of Oral Medicine and Radiology, K L E Society's Institute of Dental Sciences and Research Centre, No. 20, Yeshwantpur Suburb, Tumkur Road, Bangalore, 560022, India, Tel: 919845136960;
E-mail: praveen.birur@gmail.com

Received Date: *09 Feb 2017*
Accepted Date: *06 May 2017*
Published Date: *08 May 2017*

Citation:
Praveen BN, Shubhasini AR, Shubha G, Keerthi G, Sil S, Mani S. Gnathic Psammomatoid Ossifying Fibroma: Exceptional Cases. Ann Clin Case Rep. 2017; 2: 1357.

ISSN: 2474-1655

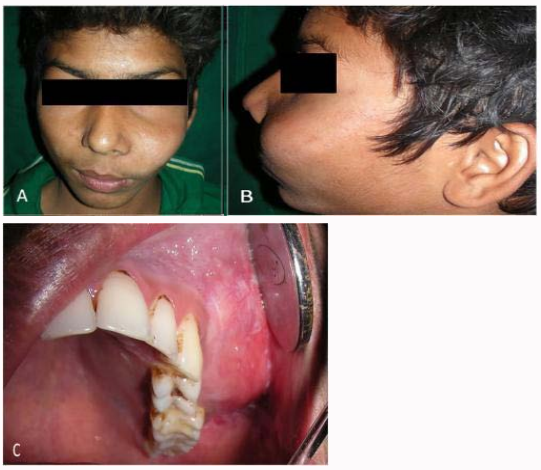

Figure 1: a and b) Extra-oral examination reveals a well-defined bony hard swelling in the left side of the face. c) Intraoral examination reveals a solitary bony hard swelling in the upper left buccal vestibule.

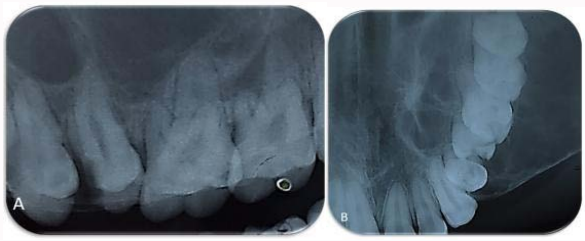

Figure 2: a and b) Intraoral periapical and maxillary lateral occlusal radiographs reveal a multi-locular lesion with ground glass appearance in 24, 25, 26 &, 27 region. Left buccal cortical plate expansion is appreciated.

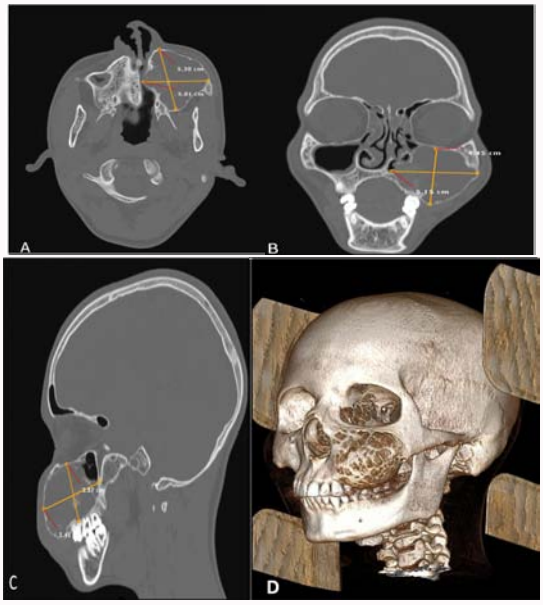

Figure 3: a) Axial section reveals a well-defined multi-locular hypodense area of 5.3 cm x5 cm. Expansion and thinning of the left buccal cortex and lateral wall of the nasal cavity is appreciated. B) Coronal section reveals the perforation of the floor and lateral wall of the nasal cavity. Complete obliteration of the left maxillary sinus and mild mucosal thickening in the right maxillary sinus. C) Sagittal section reveals a lesion measuring 4.5 cm x4.6 cm with no evidence of any calcifications. The maxillary sinus appears displaced postero-superiorly. D) The 3D reconstructed image showing the complete obliteration of the buccal vestibule.

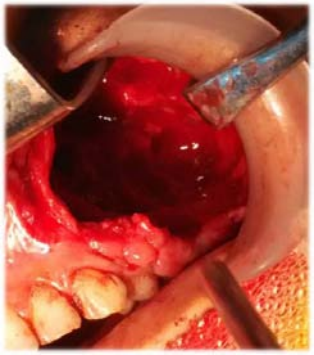

Figure 4: Intra-operative curetted surgical site.

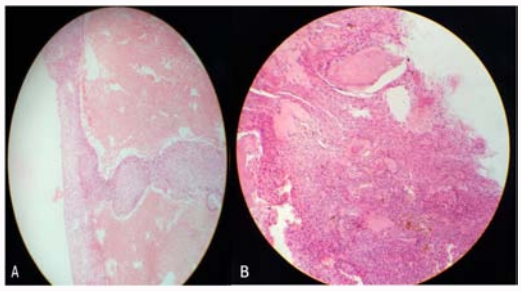

Figure 5: a, b). Shows the H and E stained section (10x) reveals psammoma bodies and blood filled cavities with few giant cells.

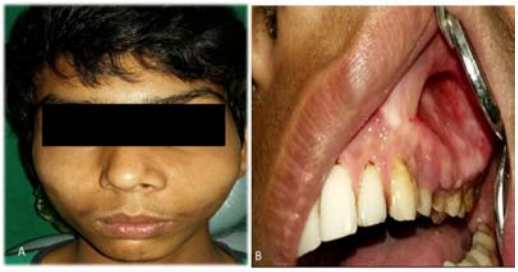

Figure 6: a and b) Reveals post-operative extra-oral and intra-oral healing surgical site after 2 months.

vestibule and on the palatal aspect of the region corresponding to 23 and extending up to the distal aspect of 28, measuring approximately 3.5 cm x 2 cm. Obliteration of the buccal vestibule was appreciated. The mucosa over the swelling and the surrounding mucosa appeared normal. The swelling was hard in consistency and non-tender. Buccal cortical plate expansion was appreciated with no evidence of decortication. 24 and 25 were grade I mobile. With the above findings, a provisional diagnosis of fibrous dysplasia of left maxilla was given. A differential diagnosis of Cemento-ossifying fibroma, cemento-osseous dysplasia ameloblastoma and maxillary sinus pathology were considered.

Radiographic investigations such as intra oral periapical radiograph (Figure 2a), Maxillary lateral occlusal radiograph (Figure 2b) and computed tomography were done (Figure 3 a,b,c and d). Excisional biopsy with enucleation, curettage and recountoring was done (Figure 4). Histopathological diagnosis of psammomatoid ossifying fibroma with aneurysmal bone cyst was made (Figure 5 a,b). Post-operative assessment (Figure 6 a,b), with regular follow-up was done.

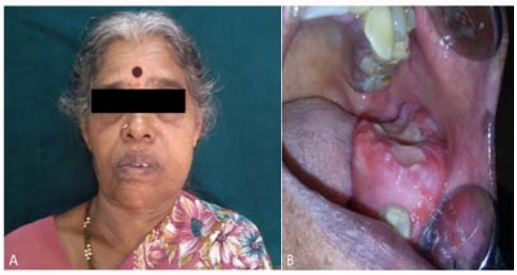

Figure 7: a) Extra oral appearance of the patient showing gross facial asymmetry due to a diffuse swelling on the left side of the face. b) Intra oral examination reveals a growth in 35-37 region measuring 5x3cm, with indentations of opposing teeth.

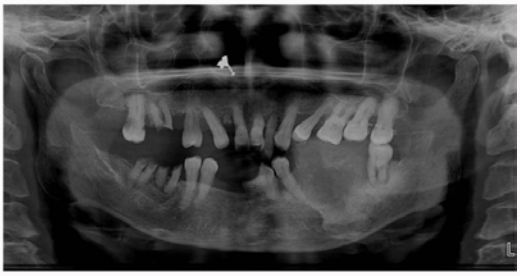

Figure 8: Panoramic view reveals a well-defined homogenous radiopacity, measuring about 5x3 cms in relation to the 35, 36 and 37 region. The internal structure shows altered trabecular pattern with ground glass appearance. Root displacement seen in relation with 35 and 37, with no evidence of root resorption.

Case 2

A 57 years old female patient reported with complaint of pain in lower left back tooth region since 1 year. She gave a history of a decayed tooth in the lower left back tooth region that was extracted 2 years back followed by uneventful healing. She was apparently healthy for 1 year after which she noticed a peanut sized swelling arising in the same region, which eventually increased over the last 2 months (Figure 7a). She gave history of was difficulty in chewing, not associated with pain, loss of weight or loss of appetite. Her medical history was non- contributory.

On examination, three left submandibular lymph nodes were palpable, measuring about 0.5 cm to 1 cm in diameter, which were circular, firm, mobile and non-tender. Gross facial asymmetry was observed on the left lower third of the face. On intraoral examination (Figure 7b), a well-defined solitary, exophytic, sessile growth on the mandibular alveolar mucosa in 35, 36 & 37 region, measuring approximately 5 cm x 3 cm and extending antero-posteriorly from distal aspect of 34 to 37 region. The overlying mucosa has occlusal indentations, surrounded with erythema and was covered with slough. Obliteration of the buccal vestibule was observed (Figure 7b). The growth was firm in consistency, non-tender, non-indurated and was fixed to the underlying structures. Buccal cortical plate expansion was appreciated.

Based on these findings, a provisional diagnosis of Peripheral Ossifying Fibroma was given and differential diagnosis of Central Giant Cell Granuloma and Primary Intra osseous Malignancy was considered.

Radiographic investigations such OPG (Figure 8), mandibular true occlusal radiograph and PA skull view was taken (Figure 9 a and b). An incisional biopsy was performed and histopathological diagnosis of Juvenile Ossifying Fibroma (psammomatoid type) was given (Figure 10). Excision of the lesion followed by curettage followed by reconstruction was performed. Post-operative assessment with follow-up of the patient showed no evidence of recurrence (Figure 11).

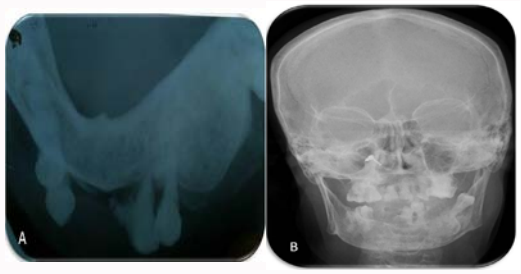

Figure 9: a and b) Mandibular true occlusal view and PA skull view reveals a homogenous radiopaque lesion in the 35, 36 regions, giving a ground glass appearance. Buccal cortical plate expansion is appreciated.

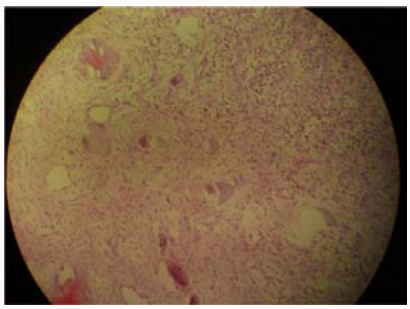

Figure 10: H & E stained smear (20x) reveals multiple psammoma bodies of varying sizes.

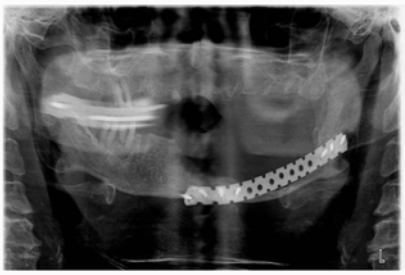

Figure 11: Post- treatment OPG reveals marginal mandibulectomy with reconstructing plate.

Discussion

Benjamin in 1938 designated the term "osteoid fibroma with atypical ossification" of the frontal sinus [6]. Later, Gogl coined the term "Psammomatoid juvenile ossifying fibroma" of the nose and paranasal sinuses [9,10]. PsJOF was first classified as a benign neoplastic fibro osseous lesion by WHO in 1995. Though, the exact aetiology of the disease is unknown, it has been demonstrated the presence of non-random chromosome break points at Xq26 and 2q33 resulting in translocation is responsible for this swelling to develop [3].

PsJOF is an extragnathic variant, commonly occurring in sinus, orbit and fronto-ethmoid bone and rarely seen in the jaw bones [6]. The average age of occurrence for PsJOF is between 16-33 years with

a slight male predilection [4,6]. On the contrary, one of our case reports the occurrence of PsJOF in the mandible of an elderly female patient. PsJOF manifests as an asymptomatic bony-hard swelling, the duration and extent of which may vary depending on the site and aggressiveness of the lesion [11,12] but our case presented clinically, as a chronic asymptomatic extensive soft tissue growth in the mandible. Another case reports, a unique gnathic variant in a young adult occurring in maxilla in combination with aneurysmal bone cyst. Simultaneous occurrence of the two lesions in the maxilla makes the case distinct. Though previous literature states similar case report in the mandible [11], none has so far been reported in the maxilla**.**

Radio-graphically PsJOF may manifest as radiolucent, mixed or radiopaque lesion which depends on the degree of calcification giving a ground glass appearance [9,13]. Root displacement is appreciated in benign lesions [4,13]. Whereas, aggressive lesions cause root resorption, expansion of bone, cortical thinning and perforations [4]. Thus our cases represent the aggressive variants of PsJOF. Radio-graphically, Fibrous Dysplasia and Ossifying fibroma can be distinguished, in which the former has a poorly defined margin whereas the latter has a well-defined margin. However PsJOF is not capsulated, but is separated from the surrounding bone by radiopaque borders, and this finding is helpful in differentiating it from fibrous dysplasia [14,15].

Though histopathologic diagnosis is confirmatory, it is based on the presence of the psammoma bodies, which are spherical cementum like bodies present in abundance [16,17]. Soopta "et al."[16] suggested that longer the duration of PsJOF, more will be the number of psammoma bodies in the histopathological examination [16].

Enucleation and curretage is the preferred modality of treatment, for benign lesions. For tumours with aggressive nature, resection is preferred with a 5 mm margin. Maxillary lesions are more difficult to remove completely because of the quality of bone and larger size of the lesion at the time of presentation [17].

PsJOF are aggressive lesions with varied clinical presentations and high recurrence rate (30-58%) [17]. Thus, earlier the diagnosis, lesser will be the degree of bone destruction and better will be the prognosis.

References

1. Khan SA, Sharma NK, Raj V, Sethi T. Ossifying fibroma of maxilla in a male child: Report of a case and review of the literature. Natl J Maxillofac Surg. 2011;2(1):73-9.
2. Pirana S, Zerati F, Voegels R, Maia R. Psammomatoid ossifying fibroma. Rhinology. 2003;41(4):250-2.
3. Sarode SC, Sarode GS, Waknis P, Patil A, Jashika M. Juvenile psammomatoid ossifying fibroma: a review. Oral Oncol. 2011;47(12):1110-6.
4. Ramakrishna T, Anuradha P, Shesha P, Shubha G, Kishen P. Gnathic Juvenile Psammamatoid Ossifying Fibroma: report of a rare case and review. J Mich Dent Assoc. 2013.
5. Gupta S, Goel S, Ghosh S, Singh A. Psammomatoid Type Juvenile Ossifying Fibroma of Mandible: A Rare Entity. Internat J Sci Study Case Reports and Review. 2014;1(7):18–21.
6. Ranganath K, Kamath SM, Munoyath SK, Nandini HV. Juvenile psammomatoid ossifying fibroma of maxillary sinus: case report with review of literature. J Maxillofac Oral Surg. 2014;13(2):109-14.
7. Kubbi J R, Kumar N, Reddy V. Juvenile Ossifying Fibroma, J Dent Sci Research. 2011;2(2):1–5.
8. Nasser MJ. Psammomatoid ossifying fibroma with secondary aneurysmal bone cyst of frontal sinus. Childs Nerv Syst. 2009;25(11):1513-6.
9. Slootweg PJ, Panders AK, Koopmans R, Nikkels PG. Juvenile ossifying fibroma. An analysis of 33 cases with emphasis on histopathological aspects. J Oral Pathol Med. 1994;23(9):385-8.
10. Malathi N, Radhika T, Thamizhchelvan H, Ravindran C, Ramkumar S, Giri G, et al. Psammomatoid juvenile ossifying fibroma of the jaws. J Oral Maxillofac Pathol. 2011;15(3):326-9.
11. Deshingkar S, Barpande S R, Bhavthankar J D. Juvenile Psammomatoid Ossifying Fibroma with Secondary Aneurysmal Bone Cyst of the Mandible, The Saudi J Dent Res. 2014;5:135–8.
12. Malathi N, Radhika T, Thamizhchelvan H, Ravindran C, Ramkumar S, Giri G, et al. Psammomatoid juvenile ossifying fibroma of the jaws. J Oral Maxillofac Pathol. 2011;15(3):326-9.
13. El-Monty S. Psammomatoid and Trabacular Juvenile Ossifying Fibroma of the craniofacial skeleton. Two distinct clinic-pathologic entities. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2002;93:296–304.
14. Ravikumar R, Raghavendra K, Kumar S. Aggressive Juvenile Ossifying Fibroma of the anterior mandible. J Dent Sci Res. 2011; 2:26–34.
15. Slootweg PJ, Müller H. Differential diagnosis of fibro-osseous jaw lesions. A histological investigation on 30 cases. J Craniomaxillofac Surg. 1990;18(5):210-4.
16. Sopta J, Dražić R, Tulić G, Mijucić V, Tepavčević Z. Cemento-ossifying fibroma of jaws-correlation of clinical and pathological findings. Clin Oral Investig. 2011;15(2):201-7.
17. Smith SF, Newman L, Walker DM, Papadopoulos H. Juvenile Aggressive Psammomatoid ossifying fibroma: an interesting, challenging and unusual case report and review of literature. J Oral Maxillofac Surg. 2009;67(1):200–6.