

Recurrent Psammomatoid Juvenile Ossifying Fibroma with Aneurysmal Bone Cyst: An Unusual Case Presentation

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Abstract

Juvenile ossifying fibroma (JOF) is a rare, benign, locally aggressive entity of the extragnathic craniofacial bones with a high tendency towards recurrence. Two distinctive microscopic patterns of juvenile ossifying fibroma have been described: a trabecular juvenile ossifying fibroma (TrJOF) and a psammomatoid juvenile ossifying fibroma (PJOF). Psammomatoid variant is predominantly a craniofacial lesion and occurs rarely in the jaws. The pathognomonic histopathologic feature is the presence of spherical ossicles, which are similar to psammoma bodies. Aneurysmal bone cyst exists as a secondary lesion arising from another osseous condition such as fibrous dysplasia, ossifying fibroma and giant cell granuloma. Very few cases of juvenile psammomatoid ossifying fibroma in association with the secondary aneurysmal bone cyst formation have been reported in the literature. Treatment consists of complete surgical removal; the incomplete excision has been associated with a high local recurrence rate. The authors report a case of recurrence of psammomatoid juvenile ossifying fibroma with aneurysmal bone cyst in an 8-year-old boy emphasizing the point that concomitant occurrence of these locally aggressive lesions requires adequate surgical removal and long-term follow-up.

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Keywords • Fibroma • Ossifying • Mandibular neoplasms

What's Known

- Literature reveals that psammomatoid juvenile ossifying fibroma in association with aneurysmal bone cyst is very rare.
- Very few cases are documented with recurrence.

What's New

- Authors document a unique case of recurrent psammomatoid juvenile ossifying fibroma with aneurysmal bone cyst involving the entire ramus of the mandible in a young patient.
- A large lesion shows an aggressive nature with recurrence occurring within a year after treatment with surgical enucleation and curettage.

Introduction

Psammomatoid juvenile ossifying fibroma (PJOF) and trabecular juvenile ossifying fibroma (TrJOF) are the two histopathological variants reported according to the WHO classification of odontogenic tumours 2005.¹ The pathognomonic histopathologic feature of the psammomatous type is the presence of spherical ossicles, which are similar to psammoma bodies.² PJOF is a slowly progressive lesion with a tendency to invade surrounding and recur after surgical excision. Aneurysmal bone cyst (ABC) is a benign cystic lesion of bone associated with rapid growth pattern that results in bony expansion. ABC occurs in two clinicopathological forms as a primary (de novo) or as a secondary lesion arising from another osseous condition, which includes giant cell granuloma, ossifying fibroma, and fibrous dysplasia.³ The importance of such presentation lies in the fact that cases of PJOF associated with secondary ABC tend to show a more aggressive growth pattern

and greater recurrence potential. This paper documents a rare case of PJOF of the mandible in association with ABC with recurrence.

Case Presentation

An 8-year-old male child presented with a painless swelling on the left side of his face since 1 year in March 2015. The patient had already visited the School of Dentistry (Dr. D.Y. Patil University, Nerul, Navi Mumbai) for the same problem one year ago, with the complaint of gradually increasing swelling. The patient was diagnosed with PJOF with ABC affecting the mandible, following which he was treated with enucleation and curettage in the same site. Extra-oral examination showed the presence of diffused swelling extended from the corner of the mouth to the posterior border of the ramus of the mandible. On palpation, the swelling was hard in consistency. An orthopantomogram showed a large multilocular radiolucent lesion on the left side of the mandible with thinning of the inferior border of the mandible. Anterior displacement of the tooth bud 37 was noted. The computed tomography (CT) sections showed a large expansile lesion in the left ramus of the mandible. Gross examination of the resected mandibular specimen showed brownish black, multiple cystic spaces in the tan white coloured solid areas. Microscopic examination revealed a fibrocellular stroma with oval to spindle-shaped cell proliferation surrounding the areas of ossifications and calcifications. Calcifications are basophilic in centre surrounding eosinophilic border resembling psammoma bodies. It also showed multiple sinusoidal blood-filled spaces devoid of endothelial lining and surrounded by fibrocellular stroma. These sinusoidal spaces were bordered by few multinucleated giant cells. Fibrocellular stroma shows plenty of small cystic spaces filled with blood elements and extravasated blood elements (figures 1-6). On compilation of histomorphologic, radiographic and clinical features of this lesion supported an interpretation of recurrent PJOF with ABC. As previously, the patient was treated with enucleation and curettage. The treatment was done with standard hemi-mandibulectomy along with the fixation of construction plate that showed satisfactory condition postoperatively. There was no sign of recurrence after six months follow-up. As the patient was minor, consent was obtained from the patient's guardians.

Discussion

WHO in the classification of odontogenic tumours (2005) defined JOF as "an actively growing lesion

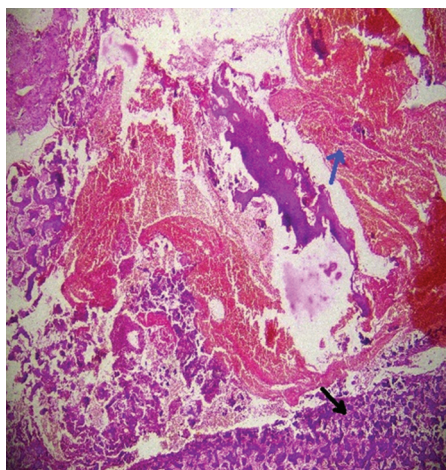


Figure 1: Photomicrograph (H&E, 4x) showing sinusoidal spaces filled with RBC's (blue arrow) surrounding connective fibrocellular connective tissue stroma with psammoma bodies (black arrow).

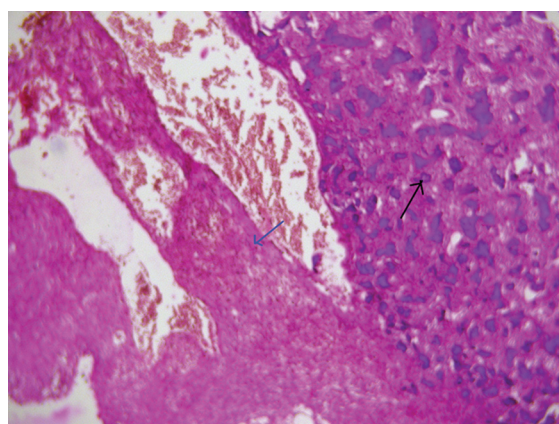


Figure 2: Photomicrograph (H&E, 4x) showing psammoma bodies (black arrow) in a cellular connective tissue stroma adjacent to aneurysmal cystic changes filled with red blood cells (blue arrow).

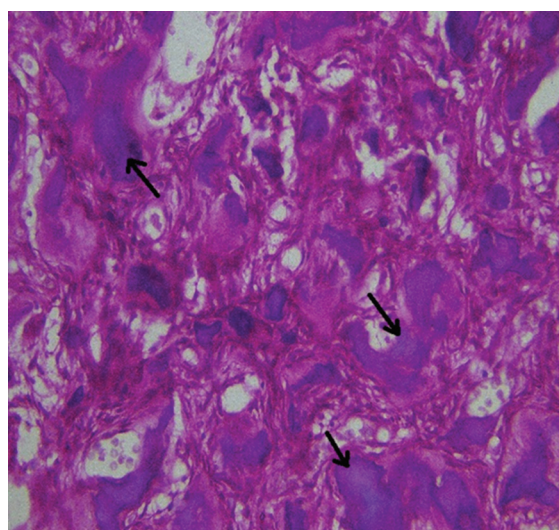


Figure 3: Photomicrograph (H&E, 40x) showing psammoma bodies (black arrow).

well demarcated from surrounding bone that is composed of cell-rich fibrous tissue containing

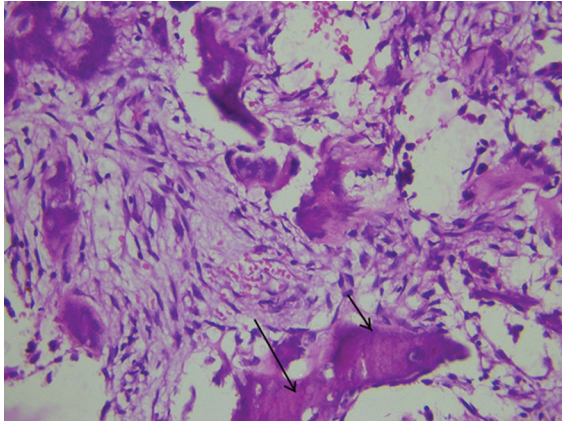


Figure 4: Photomicrograph (H&E, 40×) showing basophilic psammomatoid bodies.

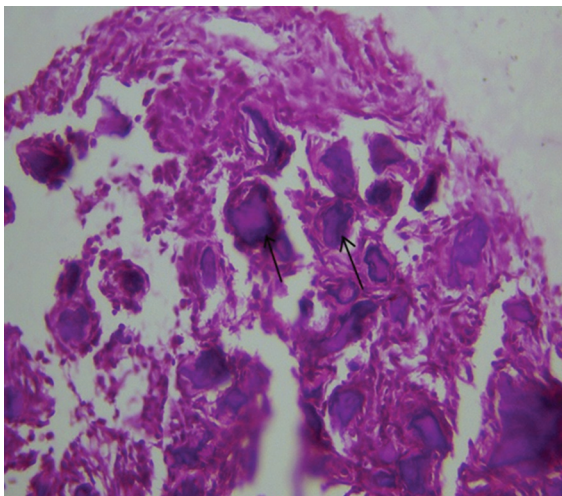


Figure 5: Photomicrograph (H&E, 40×) showing ossifications with osteoblastic rimming.

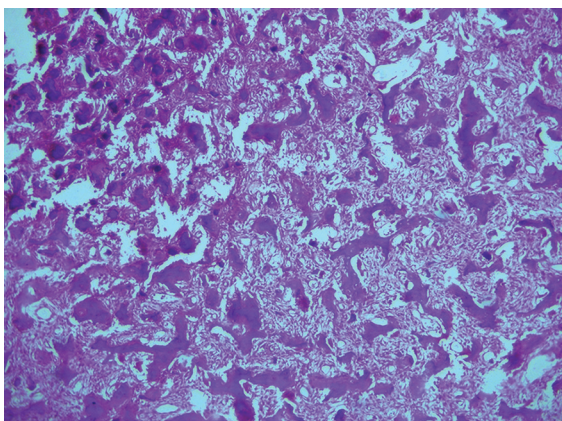


Figure 6: Photomicrograph (H&E, 10×) showing ossifications with osteoblastic rimming and psammoma bodies.

bundles of cellular osteoid and bony trabeculae without osteoblastic rimming. Giant cells may also be present". The lesion well demarcates from the surrounding bone and is non-encapsulated.⁴ On the basis of histopathological criteria, two variants of ossifying fibroma were reported (TrJOF and PJOF), which has distinct

predilection for specific age groups (TrJOF: 8.5-12 years and PJOF: 16-33 years). It signifies that PJOF observed in a few years older age group than those with TrJOF.^{4,5} The literature reveals that most of the cases involve the sinonasal areas and jaws (90%), out of which 10% of the cases involve the mandible with a slight male predominance (1.2:1).⁴ Clinically, it appears as painless swelling involving the jaws; commonly affecting the ramus of the mandible in case of the lower jaw due to bone expansion.^{1,4,5} Radiographically, PJOF appears as a radiolucent, mixed or radiopaque depending on the degree of calcification and the extent of cystic changes. Furthermore, cortical thinning and perforation were observed in case of aggressive lesions.^{4,6} Contrary to conventional ossifying fibroma, JOF is characterised by early onset that is under 15 years of age, location of tumour, rapid growth, radiological appearance, and ability to recur.⁷ The present report shows a massive involvement of the body, angle, and ramus of the mandible extending to the coronoid and condylar process. The present case shows multilocular expansile lesion, a displacement of tooth bud 37, and a thin inferior border of the mandible. Most of the features described in our case are more or less similar to articles in the literature, such as patient's age, rate of growth (within 4 months), radiographic features, and histologic features. The histogenesis of JOF is not clearly understood. The literature states that mandibular lesions can arise from myxoid dental papilla of developing tooth, cell nests of primitive mesenchymal cells or undifferentiated cells of periodontal ligament and remaining cells after incomplete migration of medial part of nasal anlage.¹ Histopathologically, PJOF shows the presence of numerous small round ossicles called psammoma bodies in a cellular fibrous stroma. The ossicles show basophilic centre with a periphery of pink rim. The cellular fibrostroma consists of plump fibroblastic spindle cells arranged in fascicular to storiform pattern with basophilic nuclei. In PJOF, osteoblastic and osteoclastic activities are observed at the periphery of the lesion.⁴ Cytogenetic changes were done only in few cases of ossifying fibroma. Deletions at 2q31-32 q35-36 were detected in one case of COF of the mandible. Randomly, chromosome breakpoints were observed at Xq26 and 2q33 resulting in X;2 translocations in a study of 3 cases of PJOF of the orbit.⁴ Aneurysmal bone cyst is an expansile, multilocular, osteolytic lesion with blood-filled spaces separated by fibrous septa containing multinucleated giant cells and peripheral reactive bone. This lesion usually

affects the long bone in the jaw, more often the mandible, predominantly in the posterior regions and can develop as a secondary change in the number of benign and malignant bone lesions.⁸ Aneurysmal bone cyst developments in PJOF have been previously reported. It is postulated that there is initially a focal myxoid change in the stroma with haemorrhage and osteoclastic giant cells with gradual expansion and formation of cyst with thin fibrous wall.³ The cyst tends to occur in the first and second decades of life and more commonly in younger patients with a high rate of recurrence (30-56%) and is aggressive in nature.^{3,5} Our patient supports these findings. This case histologically shows the presence of PJOF with secondary changes of aneurysmal bone cyst. The present case was earlier treated with enucleation and curettage. It showed recurrence within a year, which may be related to the incomplete removal of the lesion and the infiltrative nature of the tumour borders.

Conclusion

Juvenile psammomatoid ossifying fibroma involving mandible is a rare benign locally aggressive lesion. ABC is also an aggressive lesion. The recommended treatment is complete surgical excision. Partial or complete resection results in recurrence. Both lesions are of the aggressive nature and show high recurrence rate, hence a standardized follow-up protocol is mandatory for such mixed lesions.

Conflict of Interest: None declared.

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