

Recurrent juvenile psammomatoid ossifying fibroma with secondary aneurysmal bone cyst of the maxilla: a case report and review of literature

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Abstract

Juvenile ossifying fibroma is a benign fibro-osseous lesion commonly affecting the extra-gnathic craniofacial skeleton of the young individuals. The psammomatoid and trabecular variants are its two histopathological subtypes having distinctive clinico-pathological characteristics. Secondary aneurysmal bone cysts are frequently reported to arise in the pre-existing fibro-osseous lesions but rarely reported in the psammomatoid variant of the juvenile ossifying fibroma. Such hybrid lesions, especially massive in size, tend to exhibit a greater aggressive growth potential and higher recurrence rate and mandate complete surgical removal of the lesion along with a long-term follow-up. The objective of this case report was to present a rare incident of recurrent psammomatoid ossifying fibroma associated with a secondary aneurysmal bone cyst in the maxillary jaw bone of a young patient and review the similar published reports in the English literature.

Introduction

Benign fibro-osseous lesions (BFOLs) are characterized by a fibro-cellular stroma with varying degree of mineralized material leading to the replacement of physiological bone architecture with fibro-osseous tissue. Fibrous dysplasia, ossifying fibroma, and cemento-osseous dysplasia are the noted BFOLs of the jaws.^{1,2} In 2005, ossifying fibroma was included under the bone-related lesions in the WHO classification of odontogenic tumors. In the recent 2017 WHO classifications, cemento-ossifying fibroma was re-classified under the odontogenic tumors to distinguish it from the juvenile categories of ossifying fibroma.³

Juvenile ossifying fibroma (JOF) is a variant of ossifying fibroma and described as a benign neoplasm commonly affecting the extra-gnathic craniofacial skeleton of the young individuals. The psammomatoid (PsJOF) and trabecular (TrJOF) variants are the subtypes of JOF.¹ PsJOF and TrJOF are indeed distinct clinico-pathologic entities, affecting different age groups, location predispositions, and with distinguishing histopathological features. PsJOF is observed in a wider age range and predominantly found in the sinonasal and orbital bones, whereas TrJOF affects patients of a younger age group and mainly affects the jaws.²

The unusual presentations of BFOLs associated with other pathological entities are occasionally reported in the literature. The rare association of aneurysmal bone cyst (ABC) with PsJOF is one such example. ABC is described as an osteolytic lesion comprising of variable sized blood-filled spaces separated by connective tissue septa containing trabeculae of osteoid tissue and osteoclasts. It can occur *de novo* or secondary to other existing bone lesions. In the jaws, the incidence of secondary ABC is reported to be 14.8% and frequently associated with ossifying fibromas.^{4,5} The fact that JOF associated with secondary ABC cases presents with greater aggressive growth potential and higher recurrence rate adds to its clinical significance.⁶ In view of that, the present report describes a case of recurrent PsJOF with secondary ABC in a young patient along with a comprehensive review of similar published case reports in the English literature.

Case Report

A 10-year-old male patient reported to our institution with the chief complaint of painless progressive swelling in the anterior

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region of the maxillary jaw since one and a half year. The preliminary case history revealed that the patient had undergone surgical excision of a small swelling in the maxillary anterior region of the jaw 2 years back at a private hospital. On histopathological examination, the lesion was diagnosed as PsJOF. After one and a half years, the patient developed swelling in the same region, which was rapidly increasing in size. At the time the patient reported to our department, the lesion was of massive size. Elicited medical and family history did not reveal significant findings. Extra-orally, upon inspection, the swelling extended anteriorly up to the right canine region, posteriorly up to the left side zygomatic bone, superiorly up to left infraorbital margin and inferiorly up to the alveolus and measured 6x8 cm in size. The overlying skin appeared normal (Figure 1A). Palpatory findings revealed the swelling was hard in consistency and non-tender. On intra-oral inspection, a huge swelling extending from the maxillary right canine up to the maxillary left first molar obliterating the nasolabial fold with no visible secondary surface changes was noted (Figure 1B). On palpation, the swelling was hard in consistency, non-tender, and had no palpable pulsations. Tooth mobility was noted in relation to 21, 22, C, 11 and 12.

The orthopantomogram revealed a ground glass appearance with ill-defined borders over the anterior region of the maxilla extending to the left quadrant (Figure

2A). The maxillary central and lateral incisors showed displacement of the crowns as well as roots. Furthermore, the un-erupted maxillary right canine and left premolar showed evidence of displacement. The computed tomography scan showed an expansile lesion involving the alveolar arch of the left maxilla and extending into the pre-maxillary region and hard palate. The central lesional area showed focally distributed radio-dense areas. The lesional soft tissue was found to be extended into the left nasal cavity (Figure 2B). Fine needle aspiration cytology showed numerous red blood cells and occasional spindle-shaped cells suggesting benign nature of the lesion. The histopathological examination of incisional biopsy showed a highly cellular lesion predominantly composed of fibroblast-like spindle cells interspersed with varying degree of mineralization in the form of spherical ossicles. The spherical ossicles were acellular and had a basophilic center and an eosinophilic peripheral fringe resembling psammoma bodies (Figure 3A). Large vascular and sinusoidal spaces engorged with red blood cells and devoid of endothelial cell lining were also noted (Figure 3B). Based on the clinico-pathological correlation, a final diagnosis of recurrent PsJOF with secondary ABC was made.

Under general anesthesia, infiltration of 2% xylocaine with 1:100,000 adrenaline was given after the marking the Weber Ferguson's incision. Lateral rhinotomy incision with horizontal infraorbital component and midline lip-split was done. The sublabial incision was performed after splitting upper lip in the midline to facilitate elevation of the flap from the anterior wall of the maxilla. The incision was extended through the entire buccogingival sulcus up to maxillary tuberosity. The horizontal component of Weber Ferguson's incision was performed 1 mm below the infraorbital rim and the flap was reflected. The bilateral palatal cut was given after the reflection of palatal mucosa and zygoma cuts were given to disarticulate the bilateral anterior maxilla and the complete tumor was removed. The palatal prosthesis was inserted and fixed to posterior maxilla at the permanent molar region bilaterally. The closure was done in layers. The histopathological evaluation of the surgically excised specimen was consistent with the diagnosis of PsJOF with secondary ABC.

The obturator placed intra-operatively in the maxillectomy cavity was removed after 5 days. Following the first week after the surgery, healed facial incision was noted. A transient obturator with artificial denture was placed in the cavity with the support of the remaining teeth after 3 weeks

of surgery. After six weeks of surgery, the patient was given an artificial denture as an obturator with missing anterior teeth. Thereafter, the patient is in regular follow-up for past 3 years with no recurrence.

Discussion

To best of our knowledge, the present report is the seventh case of PsJOF with secondary ABC and the first case to be reported in the maxillary jaw. A thorough

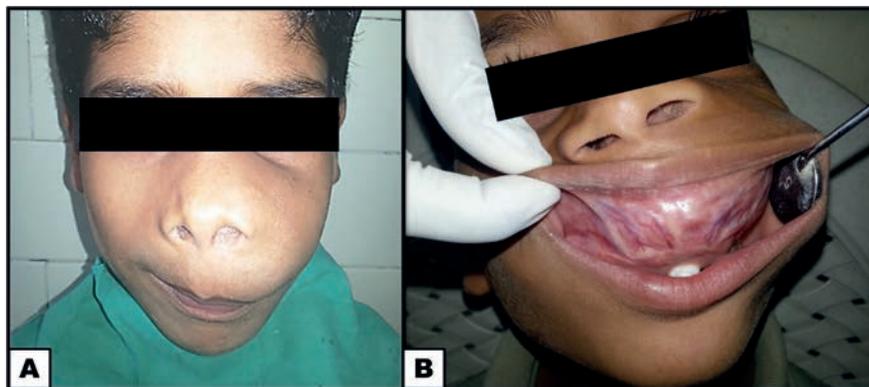


Figure 1. A) Extraoral appearance of the swelling. B) Intraoral presentation of the swelling involving the maxillary jaw.

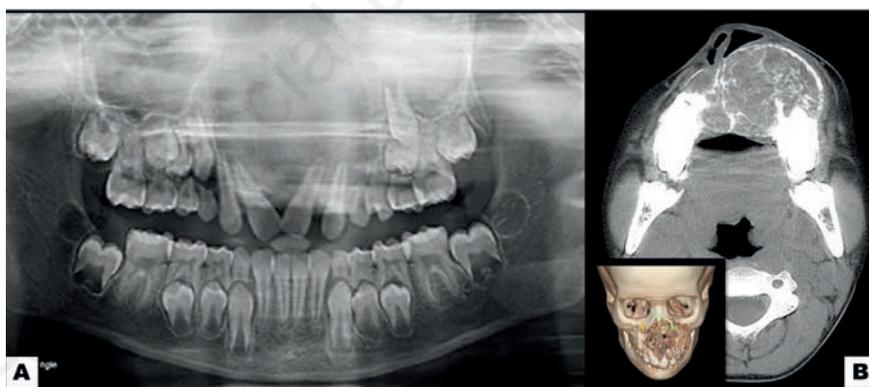


Figure 2. A) Orthopantomogram showing an expansile lesion with ground glass appearance, ill-defined borders and displacement of maxillary anterior teeth. B) Computed tomography scan revealing extensions of the lesion into the alveolar arch of the left maxilla, pre-maxillary region, hard palate and the left nasal cavity.

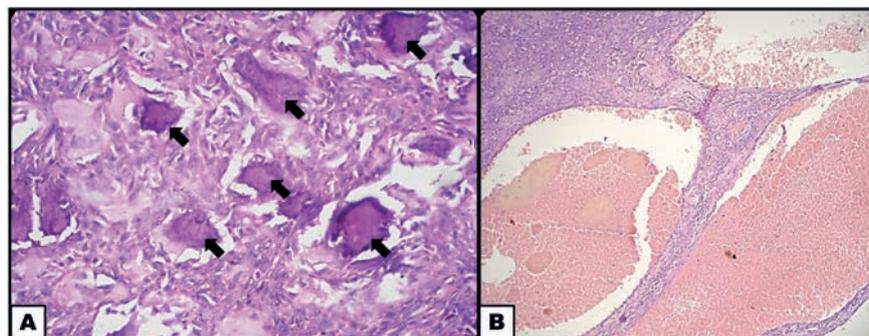


Figure 3. A) Dense fibro-cellular stroma with psammomatoid bodies (arrows) and focal areas of hyalinization. B) Large cystic spaces (devoid of epithelial lining) engorged with extravasated RBCs and separated by connective tissue septae.

search of published literature via PubMed, SCOPUS and Web of Science revealed only six previously reported cases of PsJOF with secondary ABC (in English literature) and all of which were located in the mandibular jaw. A detailed description of the aforementioned cases is depicted in Table 1.^{1,7-11}

JOF is hypothesized to originate from the overproduction of the myxo-fibrous cellular stroma which is otherwise involved in the physiological growth of the septae in the paranasal sinuses as they enlarge and pneumatize. The hyaline material produced by the stromal cells ossifies and the cystic changes are initiated by the connective tissue mucin.^{12,13} Massive aggressive maxillary JOF are frequently found to be associ-

ated with secondary ABC.⁸ The origin of secondary ABC is probably attributed to trauma or to a local vascular abnormality in a pre-existing bone lesion.^{4,5} It is proposed that there is an initial focal myxoid change in the stroma of JOF followed by hemorrhage and amassing of osteoclastic giant cells which leads to gradual expansion and formation of a cyst.⁷ Another proposed notion states that intercellular edema in the primary bone lesion with loose unsupported stroma might initiate microcyst formation, into which blood is pooled under hemodynamic pressure via rupture of blood vessels, leading to secondary ABC formation.¹⁴

It is evident from Table 1 that the reported cases occurred in young individu-

als and shows a male predisposition. Likewise, the present case was reported in a young male patient. PsJOF is frequently found to involve the sino-naso-orbital regions (ethmoidal followed by the frontal, maxillary, and the sphenoid sinuses), and in contrast to TrJOF rarely manifests in the jaws.¹⁵ Distinctively the present case was found to involve the nearly entire region of the maxillary arch in comparison to all the six cases reported exclusively in the mandibular jaw. The location in maxilla may lead to several complications such as nasal obstruction, epistaxis, displacement of orbital floor and intracranial extension.¹³ In the present case, the lesional tissue was found to deform the inferior orbital wall and

Table 1. Review of published case reports of psammomatoid juvenile ossifying fibroma with secondary aneurysmal bone cyst.

S. No.	Author	Age (years)	Sex	Location	Chief complaint	Duration	Radiographic features	Other	Treatment	Recurrence
1.	Gotmare <i>et al.</i> ⁷ 2017	8	Male	Mandibular	Painless gradually increasing swelling	1 year	Multilocular, mixed radiolucent-radiopaque lesion	Thinning of the inferior border and anterior displacement of 37 tooth bud construction plate (recurrent case)	- Enucleation and curettage (primary case) - Hemi-mandibulectomy with the fixation of	- Primary case: Yes after 1 years - Recurrent case: No (6 months follow-up)
2.	Tamgadge <i>et al.</i> ¹ 2014	7	Male	Mandible (body, angle, ramus, condyle and coronoid)	Painless gradually increasing swelling	1 year	Multilocular, mixed radiolucent-radiopaque lesion	Inferior margin showed multiple septae with thinning of the cortical plates, anterior displacement of the tooth bud 37	Enucleation, curettage and chemical cauterization	No (2 months follow-up)
3.	Deshingkar <i>et al.</i> ⁸ 2014	18	Male	Mandible (body and ramus)	Painless swelling	8 months	Multilocular, mixed radiolucent-radiopaque lesion	In part wispy radiopacities, with endosteal scalloping and a narrow transitional zone with the adjacent normal bone	Standard hemi-mandibulectomy with disarticulation of the condyle and reconstruction	Not available
4.	Tolentino <i>et al.</i> ⁹ 2012	12	Male	Mandible (body angle and ramus)	Painless gradually increasing swelling	-	Expansive, multilocular osteolytic lesion	Wispy mineralized material foci in the interior of the lesion, 1 st molar roots partially reabsorbed and the 3 rd and 2 nd molars displaced	Excision and vigorous curettage followed by reconstruction with a 2.4-mm titanium plate, to prevent fracture of the remaining bone	No (15 months follow-up)
5.	Waknis <i>et al.</i> ¹⁰ 2011	17	Male	Mandible (body, angle, ramus, coronoid and condyle)	Painless swelling	1 year	Multilocular expansile osteolytic lesion	In part a ground glass appearance, with endosteal scalloping and a narrow transitional zone with adjacent normal bone	Standard hemi-mandibulectomy with disarticulation of the condyle and reconstruction	No (24 months follow-up)
6.	Smith <i>et al.</i> ¹¹ 2009	12	Female	Mandible	Slowly enlarging non-tender swelling	2 years	Multi-locular expansile lesion	In part a ground glass appearance, with endosteal scalloping and a narrow transitional zone with adjacent normal bone	Segmental resection of the jaw followed by reconstruction	No (16 months follow-up)

extend into the left nasal cavity of the patient.

Clinically the lesions tend to be massive, painless, expanding swelling with an aggressive growth pattern and high recurrence potential (especially cases dealt with inadequate surgical interventions). The recurrence rate for PsJOF is reported to range from 30% to 56%, with few cases described to recur for more than once.¹² The present case was an example of recurrence with a massive and aggressive growth pattern leading to deformation of the inferior orbital wall. The recurrence was further associated with development of ABC as secondary lesion.

Radiographically, a multilocular expansile lesion with scalloped borders and destruction and displacement of adjacent structures are noted. Depending upon the extent of the presence of mineralized material, the lesion may appear completely radiolucent to mixed radiolucent-radiopaque in nature.¹ In the present case, an expansile lesion with ground glass appearance, irregular borders, displacement of teeth and extensions into the left nasal cavity was observed.

Microscopically, PsJOF is a non-encapsulated lesion characterized by the presence of concentric or laminated ossicles called psammoma bodies. The stroma comprises of proliferating plump fibroblastic cells, which are arranged in fascicular to the storiform pattern. Mitotic figures and cellular pleomorphism are apparent in the stroma. Negligible extracellular collagen deposition is noted. Myxomatous areas admixed with cellular spindle cell proliferation are often identified.¹² The microscopic features of ABC include the presence of several blood-filled spaces that may show signs of thrombosis and separated by septae of fibrous connective tissue. Abundant osteoclasts and immature fibroblasts along with the variable content of osteoid, hemosiderin and bone formation are noted as well.⁵

The management of smaller tumors may be achieved successfully by enucleation and curettage, but the massive aggressive tumors with infiltrating borders and where the inferior border of the mandible is compromised mandates surgical resection with 5-mm margin clearance. The present case was treated with the total maxillectomy procedure followed by reconstruction

and rehabilitation. Nonetheless, long-term follow-up of PsJOF patients especially associated with ABC is highly recommended to report late recurrences.¹¹ Since PsJOF is reported to have high chances of recurrence, an immediate reconstruction is cautioned. The prognosis of PsJOF is good since malignant transformations have not been reported.⁸ The present case was followed-up for a duration of 3 years with no recurrence.

Conclusions

The present report is the seventh case describing the rare occurrence of a recurrent PsJOF with secondary ABC in the maxillary jaw of a young male patient with subsequent description of its clinical course and management. A case study of the similar published reports was done and allied with the present case findings. To conclude, PsJOF with secondary ABC are rare benign aggressive expansile lesions of the jaws and warrants apt surgical management with long-term follow-up.

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