Case Report

Juvenile psammomatoid ossifying fibroma with aneurysmal bone cyst in the mandible: A report of a rare case

ABSTRACT

Aneurysmal bone cysts (ABCs) are rare benign lesions seen as locally destructive, rapidly expansile, and mostly affecting the long bones and vertebrae. The association of ABCs with juvenile psammomatoid ossifying fibroma (PsJOF) is predominantly seen in the extra gnathic region, and it is extremely rare with only a few cases reported so far in the mandible. Here, we report one such case of a hybrid lesion in a 30-year-old male, who presented with a solitary swelling of the right mandible showing partial obliteration of lingual vestibular sulcus, which was histologically confirmed as PsJOF as a preexisting lesion, transforming into an ABC. Such hybrid lesions are usually misdiagnosed and have been sparsely reported in the literature.

Keywords: Aneurysmal bone cyst, fibro-osseous lesions, juvenile psammomatoid ossifying fibroma

INTRODUCTION

All fibro-osseous lesions consist of the replacement of normal bone architecture with benign fibrous tissues composed of fibroblast and collagen. They also show varying amounts of mineralized material. This includes a broad group of several entities such as ossifying fibroma, juvenile ossifying fibroma (JOF), and fibrous dysplasia. Out of these, JOF is a benign bone-forming lesion, and it is defined as a variant of the ossifying fibroma in the extra gnathic craniofacial skeleton of young patients. JOF commonly occurs in the facial bones (85%), calvarium (12%), and mandibular region (10%). Very rarely, it has been reported extracranially (3%). JOF has two subtypes: psammomatoid ossifying fibroma and trabecular ossifying fibroma of which the trabecular type commonly involves the jaws. Juvenile psammomatoid ossifying fibromas (PsJOF) are unique lesions that occur commonly in children. Psammoma-like bodies are the hallmark of this lesion. An aneurysmal bone cyst (ABC) can occur as a secondary change in association with a number of benign and malignant bone lesions. Mandibular lesions are uncommon and can be mistaken for an odontogenic cyst. To the best of our knowledge, the number of PsJOF cases converting into ABC has not been reported in the literature so far, but Makek in his study found that out of the 69 cases of PsJOF, only three cases showed ABC transformation. Based on this study, the estimated percentage would be 4.3%. Here, we report a case of PsJOF that occurred in the mandible, an uncommon site, and was associated with ABC, which is also a rare entity.

CASE REPORT

A 31-year-old male patient reported to the department of oral and maxillofacial surgery with a chief complaint of a painless swelling on the right lower jaw region for the...
past 8 months. Extraorally, there was no presence of any swelling in the involved region. On palpation, it was bony hard, and there was no appreciable tenderness. Intraorally, a 2 cm × 2 cm, oval-shaped swelling is seen on the buccal and lingual aspect extending from 41 to 46 mediolaterally and from crest of alveolar ridge to lower border of the mandible superioinferiorly; there was expansion of lingual cortical plates with lingual vestibular obliteration in relation to 41–46 and displacement of first and second premolars [Figure 1]. Mild tenderness was appreciated on palpation over lingual swelling with no crepitations or egg shell crackling on buccal aspect. No mobility of involved teeth was appreciated. Class I dental caries with pulp involvement was seen in relation to 46, which showed tender on percussion. On pulp vitality test, nonvital response was seen in relation to 44–46 on electrical pulp tester. Fine needle aspiration cytology (FNAC) was performed which showed blood-tinged fluid on aspiration.

Radiographically, a large multilocular, mixed radiolucent radiopaque lesion was seen on the right side of the mandible extending from the mandibular right central incisor to the mesial root of mandibular right second molar. Superiorly, it extended up to the alveolar border. The inferior margin showed multiple septae with thinning of the cortical plates [Figure 2]. Anterior displacement of tooth 44 was noted. Based on the preliminary findings of an orthopantomogram (OPG), the lesion looked like a benign odontogenic tumor-like ameloblastoma, as well as nonodontogenic tumors such as JOF, ossifying fibroma, and ABC. The computed tomography (CT) sections showed a large severely expansile lesion in the right side of the mandible extending up to the cortical plates, and ballooning of the intermedullary space was seen. The lesion showed a heterogenously enhancing soft tissue mass with multiple bony septae.

An incisional biopsy was performed from the involved site which revealed highly cellular and vascular connective tissue stroma with few ossifying areas, numerous endothelial lined blood capillaries confirming it as ABC. To evaluate the presence of vascular feeders, CT angiography study of the bilateral external carotid artery was performed which showed no vessel feeders for the lesion. Preoperative root canal therapy was done in relation to 44-45-46. Further, enucleation was planned and performed under general anesthesia by creation of a labial window from 41 to 45 [Figures 3 and 4]. Histopathological examination of the excised mass revealed highly cellular connective tissue stoma consisting of fibroblastic proliferation. The connective tissue stroma shows the presence of irregular ossifying areas and also few round to oval ossicles (resembling like psammoma bodies). The connective tissue also indicates the presence of collagen fibers, endothelial-lined blood vessels, and hemorrhagic areas [Figure 5]. The constellation of histomorphologic, radiographic and clinical features of this lesion supported an interpretation of JPOF with secondary ABC.

**DISCUSSION**

Slootweg *et al.* identified two distinct groups, the JOF-WHO type and JOF-psammomatoid type; based on the age of
occurrence, the mean age for JOF-WHO is 11.8 years and for JOF psammomatoid type is 22.6 years. El-Mofty suggested two categories: Juvenile TOF (JTOF) and JPOF, based on histopathologic criteria. The two categories have a distinct predilection for specific age groups: The average age for JTOF is 8½–12 years, whereas that for JPOF is 16–33 years. Recently, nonrandom chromosome break points at Xq26 and 2q33 resulting in (X;2) translocation have been identified. Unfortunately, no studies have been performed on JTOF which would enable the identification of cytogenetic differences between two variants.

JOF has been separated as a distinct entity from central ossifying fibroma due to its early-onset and aggressive behavior. JOF has two well-defined histopathological variants, psammomatoid JOF and trabecular JOF (TJOF). These two entities differ in their clinical presentation and age with the involvement of sinonasal and orbital bones of the skull. However, TJOF is predominantly a gnathic lesion with a predilection for maxilla in the age group of 8.5–12 years, perhaps to the development of an ABC.

Struthers and Shear postulated that the initiation of an ABC is related to microcyst formation which is the result of intercellular edema in a primary lesion with loose unsupported stroma. Rupture of vessels in the microcysts introduces blood under hemodynamic pressure. With little resistance provided by the stroma, the blood spaces resorb the surrounding bone and lift the periosteum, which produces a thin shell of new bone. Kransdorf and Sweet have proposed that an ABC should only be considered as a secondary lesion. They believe that the preexisting lesion has often been overlooked, or the morphologic changes into ABC have erased traces of the preexisting lesions and hence have been described as primary lesions.

Boysen et al. stated that the differential diagnosis of fibrous dysplasia versus ossifying fibroma rests on a radiographic criterion after the histopathologist has verified the fibro-osseous nature of the lesion. In our case, the differential diagnosis based on clinical manifestations and conventional radiographic studies was controversial. Lesions such as periapical abscess and radicular cyst were ruled out as blood aspiration was obtained in FNAC. Histologic interpretation was critical and led to correct treatment.

The pathogenesis of ABC is controversial. An ABC may have juxtacortical or intramedullary location. Juxtacortical ABCs are found in a subperiosteal location and are primarily due to traumatic origin. Intramedullary lesions reflect secondary change within preexisting lesions. It is suggested that the cysts may result from a vascular disturbance in the form of sudden venous occlusion or the development of an arteriovenous shunt. PsJOF has to be distinguished from extracranial meningioma with psammoma bodies, which demonstrates epithelial membrane antigen positivity. Moreover, the psammomatoid ossicles in PsJOF are clearly different from the acellular spherical true psammoma bodies.

The treatment for JOF varies from surgical excision to radical surgery. JOF is associated with a very high recurrence rate of 30%–56%. The propensity of JOF to perforate cortical bone may explain its high rate of recurrence following enucleation as the tumor typically has infiltrative borders. Small-sized lesions with intact cortical plates can be successfully treated with enucleation without any postoperative recurrence which was preferred in the present case scenario. The present case was kept under close follow-up, and no recurrence was seen even after the time period of 2 years which was confirmed.
clinically and radiographically by OPG and cone beam computerized tomography showing rapid bone formation in the defect [Figures 5 and 6].

CONCLUSION

JOFs represent a unique subset of fibro-osseous lesions of the maxillofacial region. They have distinctive histomorphologic features and a tendency toward locally aggressive behavior, including invasion and destruction of the adjacent anatomical structures. PsJOF with secondary ABC is a rare benign aggressive expansile lesion of the jaws and warrants appropriate surgical management with long-term follow-up. A careful clinical, radiological, and histopathological diagnosis helps us to make out the nature of the lesion and its treatment outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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