CASE REPORT

Juvenile psammomatoid ossifying fibroma with secondary aneurysmal bone cyst of mandible

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Received 3 October 2012; revised 22 December 2013; accepted 22 December 2013
Available online 16 February 2014

Abstract Juvenile psammomatoid ossifying fibroma (JPOF) is a rare, slowly progressive tumor of the extragnathic craniofacial bones, with a tendency towards locally aggressive behavior and recurrence. The pathognomonic histopathologic feature is the presence of spherical ossicles, which are similar to psammoma bodies. Very few cases in association with the secondary aneurysmal bone cyst (ABC) 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 prognosis is good because malignant change and metastasis have not been reported. We report a case of JPOF of the mandible with secondary ABC in an 18-year-old male patient.

1. Introduction

Juvenile ossifying fibroma (JOF) is a fibro-osseous lesion that occurs in the facial bones. It is also called aggressive ossifying fibroma due to its aggressiveness and high tendency to recur. Due to its distinct histological features, it is recognized as a separate histopathological entity among the fibro-osseous group of lesions. El-Mofty identified two histopathological variants; juvenile trabecular ossifying fibroma (JTOF) and juvenile psammomatoid ossifying fibroma (JPOF). JPOF is a rare tumor of the extragnathic craniofacial bones. It is a slowly progressive lesion with a tendency to invade surrounding tissue, and recur after surgical excision. The pathognomonic histopathologic feature is the presence of spherical ossicles, which are similar to psammoma bodies. Here we report a case of JPOF of the mandible associated with aneurysmal bone cyst (ABC) in an 18-year-old male patient. To our knowledge, very few cases of JPOF of the mandible in association with ABC have been reported in the literature.

2. Case report

An 18-year-old male presented with a painless swelling on the right side of his face since 8 months. Extra-orally the...
swelling extended from para-symphyseal region to the posterior border of the ramus of mandible and superioinferiorly from the preauricular region to the inferior border of the body of mandible (Fig. 1A). Overlying skin was normal. No cervical lymphadenopathy was noted and mouth opening was normal. Intra-oral examination revealed a swelling in retromolar region with obliteration of lower buccal vestibule from distal of left mandibular first premolar to the anterior border of ramus of the mandible (Fig. 1B). The labial cortex was expanded with no apparent mucosal changes. On palpation, the swelling was hard in consistency with no fluctuation elicited.

An orthopantomogram showed a multilocular expansile lesion on the left side of body and ramus of the mandible without invasion of the coronoid and condylar processes (Fig. 1C). The lesion had in part wispy radiopacities, with endosteal scalloping and a narrow transitional zone with the adjacent normal bone. Routine hematological and urine investigations ascertained values within normal limits.

Histopathological examination of incisional biopsy revealed a cellular proliferation of uniform, cytomorphologically bland fibroblastic spindle cells with no evidence of mitotic figures (Fig. 2A). Interspersed between the fibroblastic cells were numerous irregular and spherical ossicles exhibiting varying degrees of calcification with minimal extra-cellular collagen deposition. These psammoma body-like ossicles were relatively acellular, with peripheral eosinophilic rimming and distributed homogeneously throughout the section (Fig. 2B). Large vascular and sinusoidal spaces devoid of the epithelial lining and engorged with red blood cells (Fig. 3A) were evident. Osteoclastic giant cells were focally demonstrated in the lining of these spaces establishing the presence of associated ABC (Fig. 3B). The constellation of histomorphologic, radiographic and clinical features of this lesion supported an interpretation of JPOF with secondary ABC.

Due to patient’s poor economic status, a primary reconstruction with a bone graft could not be done. Standard hemi-mandibulectomy with disarticulation of the condyle was done. A titanium reconstruction plate with condylar head which was pre-adapted on cadaveric mandible was prepared to give the best patient profile. Arch bars were fixed to the maxilla and right mandible and maxillo-mandibular fixation (MMF) was done. The reconstruction plate was fixed with three screws in the anterior mandible. The MMF was released and condylar movement was observed. Suture removal was done after 8 days and MMF was done with elastics for 3 weeks to maintain the occlusion.

The post-operative facial appearance and oral function was satisfactory. Due to the economic reason patient refused further reconstructive surgery and did not turn up for follow-up.

3. 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 of for JOF-WHO is 11.8 years and for JOF Psammomatoid type is 22.6 years.\textsuperscript{6} El-Mofty suggested two categories, JTOF and JPOF, based on histopathologic criteria.\textsuperscript{4} The two categories have a distinct predilection for specific age-groups: the average age for JTOF is 8.5–12 years, whereas that for JPOF is 16–33 years.\textsuperscript{6,13} Recently, non-random chromosome break points at Xq26 and 2q33 resulting in (X;2) translocation have been identified.\textsuperscript{13} Unfortunately, no studies have been performed on JTOF which would enable the identification of cytogenetic differences between two variants.

The age of the patients ranges from 3 months to 72 years, with mean age of occurrence 17.7 years.\textsuperscript{6,7,13} It is most commonly seen in first and second decade of life.\textsuperscript{4,6,8} It shows slight male predominance with the male:female ratio of 1.2:1.\textsuperscript{4,6,8} The age and gender of the present case is in confinement of the data from literature. Most cases affect sinonasal area and jaws (90%), out of which 10% cases involve mandible.\textsuperscript{6,13} Clinical manifestations include proptosis, visual
disturbances, progressive blindness, airway obstruction, and progressive craniofacial deformities. Local expansion may extend into paranasal sinuses, nasal cavity, nasopharynx, palate, and cranial cavity. In the jaw, painless swelling with expansion of cortical plates is present. Mandibular ramus is commoner site than body of the mandible. The present case shows involvement of body, angle and ramus of the mandible (Fig. 1).

The imaging study of present case showed a multilocular expansile lesion with a narrow transitional zone with the adjacent normal bone (Fig. 1C). The internal structure can be radiolucent, mixed or radiopaque depending on the degree of calcification and presence of cystic areas. Root displacement is common and resorption, though rare, can occur. The present case demonstrates resorption of the roots of second molar (Fig. 1C). The lesion can cause expansion as well as perforation. The radiographic features can resemble that of fibrous dysplasia and cemento-ossifying fibroma. JPOF is not capsulated but is separated from the surrounding bone by radiopaque borders, and this finding is helpful in differentiating it from fibrous dysplasia. A ground glass appearance is also reported. It usually has a centrifugal growth pattern, which can lead to an erroneous clinical diagnosis of cemento-ossifying fibroma.

The characteristic histologic feature is the presence of numerous small, round ossicles or “psammomatoid” bodies embedded in a cellular fibrovascular stroma. The ossicles are mineralized collagenous foci that vary from small, smoothly contoured round-to-oval patterns to larger, irregular shapes, with concentric layering like that of psammoma bodies. A marginal osteoid rim surrounds the ossicles. The ossicles have a thick irregular collagenous rim that may include multiple ossicles. The ossicles may fuse together to form trabeculae showing reversal lines. Sometimes, JPOF contains basophilic concentrically lamellated particles with irregular thread-like or thorn-like calcified strands in hyalinized background.

The present case was characterized by large cystic areas of ABC showing giant cells and osteoid formation. Development of the ABC in JPOF has been previously reported. It develops initially as a focal myxoid change in hemorrhagic stroma. Osteoclastic giant cells appear along with gradual expansion and formation of cysts having thin fibrous walls. The cysts

Figure 2  (A) Histopathological examination showing cytomorphologically bland fibroblastic spindle cells with interspersed numerous ossicles (H and E stain; Original magnification x100). (B) Psammomatoid ossicles with a smooth contour and thin peripheral eosinophilic rim (arrows) set in cellular fibroblastic stroma (H and E stain; Original magnification x400). Figure 3  (A) Aneurysmal bone cyst like changes showing large cystic areas filled with red blood cells (arrows) (H and E stain; Original magnification x100). (B) Cystic areas devoid of epithelial lining and presence of giant cells (black arrows) and psammomatoid ossicles in the wall (H and E stain; Original magnification x400).
tend to occur commonly in the younger patients in the first and second decades of life. Large aggressive maxillary lesions are commonly associated with ABC.\textsuperscript{7}

The aggressive nature of this entity with high rates of recurrence (30–56\%)\textsuperscript{6,7} suggests that JPOF should be treated with surgical resection, rather than conservative curettage.\textsuperscript{4,13} Fatal consequences are extremely rare and are caused by complications arising from direct intracranial extension.\textsuperscript{7} Recurrence may be attributed to difficulty in proper resection caused by the location of the lesion and the infiltrative nature of the tumor borders. Complete resection results in no recurrence for a period of 6 months to 7 years\textsuperscript{8} or more than 7 years.\textsuperscript{4}

There is no standardized follow-up protocol in the literature. Because of high recurrence rate, immediate reconstruction is not advised. Secondary reconstruction may be undertaken sooner for slow-growing lesions, and be delayed for fast-growing lesions. Although there are no cases of malignant transformation, rapidly growing lesions should raise suspicion and add a reason for delaying reconstruction.\textsuperscript{12}

The prognosis is good because malignant changes and metastases have not been reported.\textsuperscript{4,13,14} Radiotherapy is contraindicated because of the risk of malignant transformation\textsuperscript{13} and potential harmful late effects in children. Thus appropriate recommended treatment is aggressive surgical approach followed by clinical and radiological follow-up.

Conflict of interest

None.

Acknowledgement

None.

Reference