Case Report - Pathology

Psammous desmo-osteoblastoma with concomitant aneurysmal bone cyst of mandible

Access this article online
Website: www.amsjournal.com
DOI: 10.4103/2231-0746.161136
Quick Response Code:

S. K. Shruthi, V. V. Kamath, Satish Hegde[1], B. Sreevidya[1]
Departments of Oral and Maxillofacial Pathology and [1]Oral and Maxillofacial Surgery, Dr. Syamala Reddy Dental College Hospital and Research Centre, Bengaluru, Karnataka, India

Address for correspondence:
Dr. V.V. Kamath, Department of Oral and Maxillofacial Pathology, Dr. Syamala Reddy Dental College Hospital and Research Centre, Bengaluru, Karnataka, India.
E-mail: kamathvv2003@yahoo.com

ABSTRACT

Juvenile psammomatoid ossifying fibroma is a gradually progressive, aggressive extragnathic craniofacial tumor of bone. Due to its complex histogenesis, biological behavior, histology, and classification, its nomenclature has always been the focus of debate among pathologists. Based on the clinical behavior and histology, the term psammous desmo-osteoblastoma (PDO) is an apt expression for this lesion. Immunohistochemical investigation with anti-osteonectin shows positivity for spindle cells whereas psammoma bodies are negative. These results shore up the hypothesis of osteogenic differentiation of the undifferentiated mesenchymal cells of the periodontal ligament that may be responsible for the aggressive behavior of the lesion. Aneurysmal bone cysts (ABCs) are known to be secondarily associated with primary osseous neoplasms like ossifying fibroma, giant cell granuloma, etc. We report a rare case of PDO with concomitant ABC in the mandible of a 30-year-old male patient. The present lesion had recurred at the same site of an osteolytic lesion diagnosed 7 years ago as a benign fibro-osseous lesion and treated by conservative surgical curettage. The histological presentation substantiated by special stains and immunohistochemistry point to the diagnosis of psammous desmo-osteoblastoma with a concomitant ABC. Review of the literature revealed the presentation to be rare with very few cases reported till date.

Keywords: Aneurysmal bone cyst, juvenile psammomatoid ossifying fibroma, osteonectin, psammous desmo-osteoblastoma

INTRODUCTION

Psammous desmo-osteoblastoma (PDO) is a benign aggressive fibro-osseous neoplasm of the craniofacial region.[1] According to WHO (2005) odontogenic tumor classification, this lesion has been classified as a subset of ossifying fibroma with the ICD code (9263/0).[2] Juvenile ossifying fibroma is a broad term with two distinct histopathological variants: Trabecular juvenile ossifying fibroma which is distinguished by the presence of trabeculae of fibrillar osteoid and woven bone and juvenile psammomatoid ossifying fibroma (JPOF) that is characterized by the presence of small uniform spherical ossicles that resemble psammoma bodies. The first case was reported in 1983 by Benjamins in the frontal sinus. In 1985, Makek called it as a variant of osteoblastoma and termed it as psammous desmo-osteoblastoma (PDO).[3] This tumor is commonly seen in the extragnathic craniofacial bones, particularly the periorbital, frontal, and ethmoid bones and is extremely uncommon in the jaws. It is an asymptomatic invasive lesion causing erosive destruction of the adjacent bone which has a high tendency for recurrence after surgical treatment.[4]

Aneurysmal bone cyst (ABC) is a pseudocystic lesion that commonly manifests as a secondary lesion with various highly vascularized and fibro-osseous lesions in the vertebrae and long bones but are rarely seen in the jaws.[5,6]

We report a rare case of PDO with concomitant ABC of the mandible occurring as a recurrent lesion in a 30-year-old male. Very few cases of this hybrid lesion have been reported in the orofacial region, the present case being the 5th case reported in the jaw and 8th case in the oro-maxillofacial region [Tables 1 and 2].

CASE REPORT

A 30-year-old male patient reported to the Oral and Maxillofacial Surgery Department with a complaint of swelling in left lower jaw since 1year [Figure 1]. Patient gave a history of similar asymptomatic hard swelling in the same site 7 years ago. Records revealed an unilocular osteolytic lesion in the left mandible extending from the mandibular canine region to the second molar causing expansion of the jaw involving the lower border of the mandible [Figure 2a]. A conservative surgical curettage was listed as having been done under general anesthesia and the pathological diagnosis was reported as a benign fibro-osseous lesion.

Present clinical examination revealed a diffuse extraoral swelling on the left side of the face extending from the midline of the lower lip to tragus of ear anteroposteriorly and extending from ala-tragus line to submandibular region superior-inferiorly. Skin overlying the swelling was normal and tender on palpation. Intraorally it was extending from the lower right lateral incisor to left ramus of the mandible with the obliteration of vestibule. No mobility of teeth was detected and the oral hygiene status appeared to be well maintained. No neurological deficits involving mental or inferior alveolar nerve were elicited.

On radiographic examination, orthopantomograph (OPG) showed a large multilocular radiolucency with well-defined sclerotic borders with the expansion of the cortical plates [Figure 2b]. Serum alkaline phosphatase level was elevated with the value of 209 IU/L.

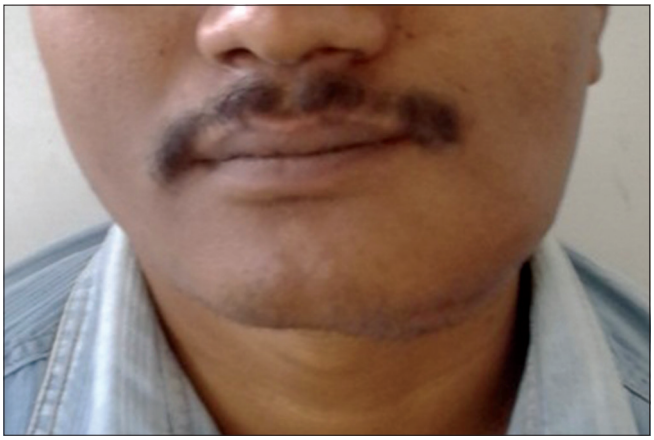

Figure 1: Preoperative photograph showing diffuse swelling of the lower one third of the Face

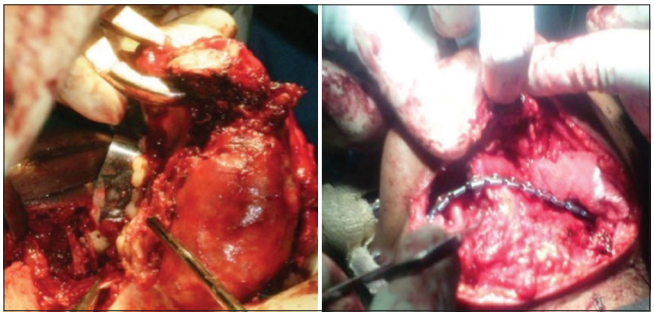

Figure 3: Photograph showing submandibular approach to the mandible, mandibular resection and reconstruction plate placement

Routine preoperative investigations were within normal limits and following a preanesthetic evaluation the patient was posted for resection of the lesion under general anesthesia. Segmental mandibulectomy was done under general anesthesia with the preservation of about 3 mm posterior border of the ramus and it was stabilized using 2.5 mm reconstruction plates [Figure 3]. Adequate stability of the reconstruction plate was ensured and the patient was put in intermaxillary fixation (IMF) immediate postoperative with nasogastric tube feed to ensure adequate stabilization. The patient was discharged on the third postoperative day following uneventful healing. Postoperative OPG revealed adequate fixation and functional occlusion [Figure 2c]. The resected specimen was sent for the histopathological examination. The patient is under follow-up since 6 months and no signs of recurrence have been noticed.

The resected left mandibular segment along with teeth [Figure 4] (41–38) on macroscopic examination measured approximately 10 cm × 7 cm × 5 cm, surface was brownish-white in color and consistency varied from firm to hard.

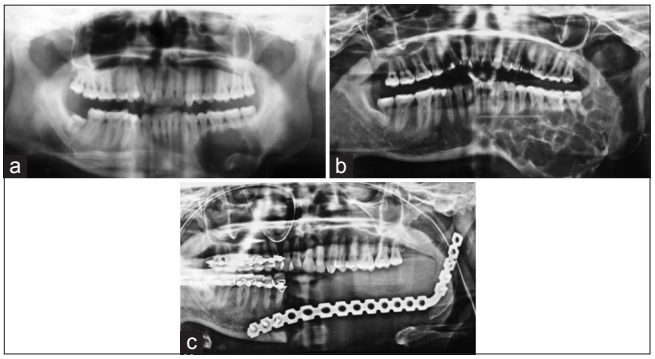

Figure 2: (a-c) Orthopantomograph (OPG) (a) of lesion 7 years ago showing unilocular radiolucency with well-defined sclerotic border. (b) The radiograph of the lesion at the present time shows extensive involvement with the lesion having crossed the midline and involved the ramus right up to the sub-condylar region. Note the multilocularity. (c) Postoperative OPG showing stabilization of mandible with reconstruction plate and intermaxillary fixation

Figure 4: Resected specimen showing the expansion of the buccal cortical plate

Table 1: List of the hybrid cases reported in the literature till date in the jaws

Case number	Author/year	Age and gender of the patient	Site of occurrence	Concomitant lesions	Immunohistochemical investigations
1	El-Mofty *et al.*/2002[7]	15-53 years*	Case 1 - left parietal bone Case 2 - ethmoid sinus Case 3 - mandible	ABC	Nil
2	Sarode *et al.*/2011[8]	17 years/male	Mandible	ABC	Nil
3	El-Mofty *et al.*/2011[9]	17 years/male	Mandible	ABC	Nil
4	Deshigkar *et al.*/2014[10]	18 years/male	Mandible	ABC	Nil

*Gender not mentioned in the article

Table 2: List of the hybrid case reported in the orofacial region in the literature till date

Case number	Author/year	Number of cases	Age and gender of the patient	Site of occurrence	Concomitant lesions	Immunohistochemical investigations
1	El-Mofty *et al.*/2002[7]	3	15-53 years*	Case 1 - left parietal bone Case 2 - ethmoid sinus Case 3 - mandible	ABC	Nil
2	Nasser/2009[4]	1	12 years/male	Frontal sinus and the supraorbital part	ABC	Lesional cells showed strong and diffuse reactivity for vimentin No positivity was seen EMA
3	Sarode *et al.*/2011[8]	1	17 years/male	Mandible	ABC	Nil
4	El-Mofty *et al.*/2011[9]	1	17 years/male	Mandible	ABC	Nil
5	Deshigkar *et al.*/2014[10]	1	18 years/male	Mandible	ABC	Nil

*Gender not mentioned in the article. EMA=Epithelial membrane antigen

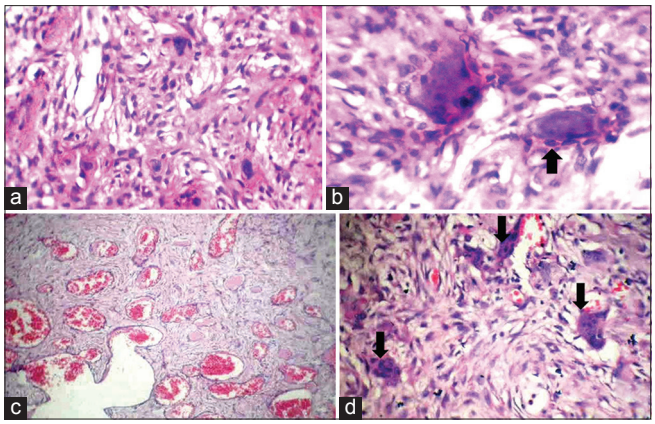

Figure 5: (a and b) Composite photomicrographs of H and E stained section showing the proliferation of spindles cells in fibrous stroma and psammoma bodies with osteoblastic rimming (c and d) showing blood filled spaces and multinucleated giant cells on the periphery of the blood vessels; features indicative of an aneurysmal bone cyst (H and E stained, magnification x10)

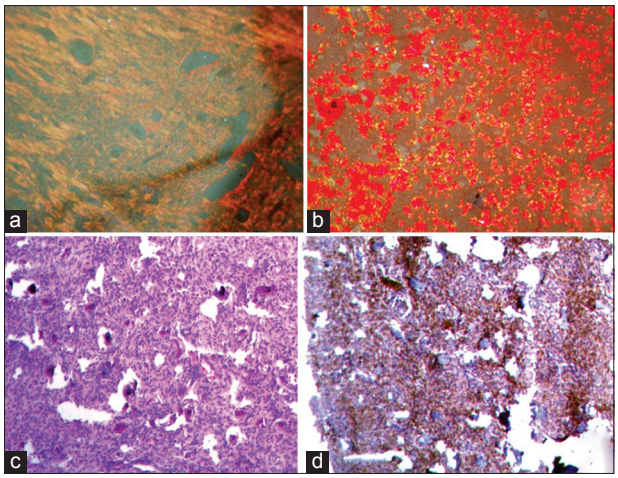

Figure 6: (a-d) Composite photomicrographs of Picrosirius Red stained section (a) under the polarizing microscope showing fibrous stroma which has loose aggregations of connective tissue fibers and (b) the psammoma bodies with densely packed collagen (Picrosirius Red, original magnification ×10). (c) H and E stained section showing spindles cells and psammoma bodies in fibrous stroma and (d) Immunohistochemistry with anti-osteonectin showing positivity for spindle shaped cells and negative for Psammoma bodies. (original magnification x 40)

Microscopic examination of hematoxylin and eosin sections showed various hypercellular fibrous connective tissue stromas consisting of plump hyperchromatic fibroblasts with minimal collagen deposition arranged in the form of sheets and whorls. Numerous psammoma bodies having eosinophilic and fibroblastic rimming were distributed throughout the section [Figure 5a and b]. Pseudocysts of varying sizes filled with erythrocytes were seen, adjacent to which multinucleated giant cells were evident [Figure 5c and d]. This was typical of an ABC and seemed to be present at the distal end of the lesion. To assess the histopathological nature of the collagen stroma

and calcified bodies, Picrosirius red and Alizarin Red stains were employed and the sections visualized under polarized microscope. Tissue stained with Picrosirius Red under polarizing microscope showed compact collagen aggregation in the calcifications and loose collagen in the stroma. Tissue stained with Alizarin Red under polarizing microscope showed varying densities of calcification. The origin and nature of the calcified bodies in a fibro-osseous lesion in the jaws is always a matter of debate. To identify the origin of the cells forming the hard tissue, the sections were stained with anti-osteonectin (Clone 15G12, monoclonal antibody, Novocastra™ Lyophilized antibodies, Leica Biosystems Newcastle Ltd., UK) an immunohistochemical antibody specific to osteoblast lineage of cells. Spindle cells in the stroma showed positivity whereas psammoma bodies were negatively stained [Figure 6a-d]. Based on the constellation of histologic, clinical, and radiographic features final diagnosis of PDO concomitant with ABC was given.

DISCUSSION

Psammous desmo-osteoblastoma is a benign fibro-osseous lesion where fibrous tissue containing mineralized structures replaces the normal bone architecture.[11] Many terminologies have been used in the literature to denote this lesion. Gogl in 1949 called it as psammomatoid ossifying fibroma.[9] Due to the aggressive behavior in the patients below 15 years of age Pimenta *et al*. in 1952 called it as juvenile active ossifying fibroma.[7] WHO (2005) classified it as subset of ossifying fibroma under bone related lesions in odontogenic tumors. It is also called as cementifying fibroma, cemento-ossifying fibroma, and juvenile (active/aggressive) ossifying fibroma with the synonym of juvenile ossifying fibroma.[2,3] Other synonyms such as psammo-osteoid-fibroma, psammous desmo-osteoblastoma, JPOF, and juvenile aggressive psammomatoid ossifying fibroma have been cited in the literature.[12]

The lesion is believed to arise from the undifferentiated mesenchymal cells of the periodontal membrane which serve as multipotential precursor cells capable of differentiating into cementum, osteoid, or fibrous tissue and give rise to a spectrum of fibro-osseous lesions.[13,14] A genetic predisposition may be present based on the observation of a translocation of (X; 2) at the nonrandom chromosome break points of Xq26 and 2q33.[15] Pimenta *et al*., proposed that ossifying fibroma may occur due to haploinsufficiency of HRPT2, which did not hold good in case of PDO.[9,11]

Psammous desmo-osteoblastoma is seen most commonly in the age group between 3 months and 72 years and is seen predominantly in males. It frequently involves paranasal sinus, maxilla, mandible, and calvaria which manifests with proptosis, lateral displacement of the eyeball, progressive blindness, nasal obstruction, headaches, facial swelling, pain, recurrent sinusitis whereas in the jaws it manifests with missing teeth, disturbance of developing sockets and expansion of the cortical plates.[7]

Depending on the degree of calcification and cystic changes this lesion, exhibits sclerotic border which may be radiolucent, mixed or radiopaque. In some cases, ground glass appearance may also be noted.[9]

Histopathologically this lesion consists of a hypocellular and hypercellular areas where plump spindle-shaped fibroblasts with basophilic nuclei are arranged in a fascicular and storiform pattern. Basophilic myxomatous stroma is seen admixed with cellular spindle cell proliferation. Calcifications of varying sizes which are basophilic with peripheral rim around it are called "psammoma bodies" which is derived from a Greek word meaning "sand". Small spicules which are needlelike crystalloids project towards the periphery bestowing it a "brush border" appearance.[9,14] Fibroblastic (preosteoblastic), osteoblastic, and osteocytic cells along with globular mineralization dissimilar to the mineralization of the psammoma bodies are observed on electron microscopic examination.[16]

Sections of the present case were stained with Picrosirius Red and Alizarin Red. The predominance of green-greenish yellow collagen fibers signify an aggressive behavior of the lesion [Figure 3a and b]. The greenish yellow fibrous component under polarizing microscopy indicates an immature stroma that is highly cellular and with a penchant for proliferation. Based on these findings, it was hypothesized that the stroma with poorly packed pathological collagen fibers was induced by proliferating immature cells of PDO.[17]

Immunohistochemical stain with anti-osteonectin showed the positivity of intracellular staining in all tumor cells and extracellular staining in the osteoid, but psammoma bodies were negative. These results are in accordance with previous studies.[16] This renders a unique immunohistochemical trait on this lesion different from other fibro-osseous lesions.[18] The genetic lack of haplotyping with HRPT2, positivity with anti-osteonectin, the aggressive behavior and propensity for recurrence, and the distinctive histopathological features have initiated the debate for incorporating PDO as a separate lesion with its own nomenclature.

Proliferative markers such as Ki-67/MIB-1 did not show any significant staining in the PDO but showed positivity for vimentin, epithelial membrane antigen, SMA, and CD10, with deficient expression of CD34, S-100 protein, and cytokeratins which was similar to that seen in cemento-ossifying fibroma arising from tooth-bearing areas.[4,19-21]

Radiographically PDO should be differentiated from ossifying fibroma, fibrous dysplasia, and mixed odontogenic tumors. PDO presents with the destruction of cortical plate with well-defined sclerotic borders as in our case which is same as ossifying fibroma. Fibrous dysplasia is radiographically ill-defined with blending of the lesion with the normal bone, whereas the mixed odontogenic tumor are usually associated with tooth-bearing areas, impacted tooth, unerupted tooth which are usually well-demarcated and contain coarse opacities that are similar to radio densities to tooth structure.[22]

Psammous desmo-osteoblastoma histopathologically mimics cemental lesions has to be distinguished from the psammomatoid ossicles of fibro-osseous lesions and cementicles in cementum forming lesions. Cementicles are acellular matrix devoid of associated osteocytes, distinguishing basophilic reversal or cemental lines are seen, the periphery is darkly stained with

an eosinophilic fibrillar appearance and finer or more delicate parallel birefringent fibrils are seen under polarized light.[9]

Development of ABC in PDO is frequently reported. They develop initially as a focal myxoid change in the stroma with hemorrhage and osteoclastic giant cells, with gradual expansion and formation of cysts with thin fibrous walls. The cysts tend to occur more Commonly in the younger patients in the first and second decades of life. It is postulated that it may be due to arteriovenous malformation by involutionary changes in the primary lesion which may create hemodynamic changes or reparative or tumor-induced anomalous vascular process.[6,23] Oliveira *et al*. based on his investigations concluded that CDH and USP6 rearrangements in ABCs was negative and thus it characterizes a vague morphologic pattern in different set neoplasms.[24]

This lesion has a good prognosis. Though this lesion has a propensity to invade locally and recur, the reported cases of metastasis are nil.[25]

CONCLUSION

Psammous desmo-osteoblastoma is a lesion arising due to osteogenic differentiation of undifferentiated mesenchymal cells of the periodontal ligament and thus aggressive with high chances of recurrence. Due to its unique biological behavior and nature, there is increasing realization that this lesion can be considered as a separate entity and be classified as such in the WHO list of fibro-osseous lesions.

REFERENCES

1. Guruprasad R, Gupta SD, Dua N, Mehta R. Juvenile psammomatoid ossifying fibroma – A case report. Peoples J Sci Res 2011;4:31-5.
2. Barnes L, Eveson JW, Reichart P, Sidransky D. Pathology and genetics of head and neck tumours. In: Slootweg PJ, El Mofty SK, editors. Odontogenic Tumors. Lyon: IARC Press; 2005. p. 321-2.
3. 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;15:326-9.
4. Nasser MJ. Psammomatoid ossifying fibroma with secondary aneurysmal bone cyst of frontal sinus. Childs Nerv Syst 2009;25:1513-6.
5. Zadik Y, Aktas A, Drucker S, Nitzan DW. Aneurysmal bone cyst of mandibular condyle: A rare case report and review of literature. J Craniomaxillofac Surg 2012;40:e243-8.
6. Kransdorf MJ, Sweet DE. Aneurysmal bone cyst: Concept, controversy, clinical presentation, and imaging. AJR Am J Roentgenol 1995;164:573-80.
7. El-Mofty S. Psammomatoid and trabecular juvenile ossifying fibroma of the craniofacial skeleton: Two distinct clinicopathologic entities. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2002;93:296-304.
8. Waknis P, Sarodeb SC, Dolas RS. Psammomatoid juvenile ossifying fibroma of the mandible with secondary aneurysmal bone cyst: A case report. Asian J Oral Maxillofac Surg 2011;23:83-6.
9. Sarode SC, Sarode GS, Waknis P, Patil A, Jashika M. Juvenile psammomatoid ossifying fibroma: A review. Oral Oncol 2011;47:1110-6.
10. Deshigkar SA, Barpande SR, Bhavthankar JD. Juvenile psammomatoid ossifying fibroma with secondary aneurysmal bone cyst of mandible. Saudi J Dent Res 2014;5:135-8.
11. Pimenta FJ, Gomes DA, Perdigao PF, Barbosa AA, Romano-Silva MA, Gomez MV, *et al*. HRPT2 gene alterations in ossifying fibroma of the jaws. Oral Oncol 2006;42:735-9.
12. Nwizu NN, George MD, Chen F. An aggressive psammomatoid ossifying fibroma presenting as a sphenoethmoidal mass. N Am J Med Sci 2010;3:24-7.
13. Neville BW, Damm DD, Allen CM, Bouquot JE. Oral and Maxillofacial Pathology. 2ed. Saunders, An imprint of Elsevier,11830, Westline Industrial Drive, St. Louis, Missouri; 2002. p. 564-6.
14. Alper S, Yavuzer R, Isik I, Uluoglu O, Atabay K. Juvenile fibroma ossifying fibroma. Report of a case. Gazi Med J 2001;12:67-71.
15. Park S, Jae B, Lee L JH, Cho KJ. Juvenile ossifying fibroma: A clinicopathologic study of 8 cases and comparison with craniofacial fibro-osseous lesions. Korean J Pathol 2007;41:373-9.
16. Störkel S, Wagner W, Makek MS. Psammous desmo-osteoblastoma. Ultrastructural and immunohistochemical evidence for an osteogenic histogenesis. Virchows Arch A Pathol Anat Histopathol 1987;411:561-8.
17. Solomon M, Khandelwal S, Carnelia S. Psammomatoid juvenile ossifying fibroma of the mandible – A histochemical insight. Internet J Dent Sci 2009;7:2.
18. Sakamoto A, Oda Y, Iwamoto Y, Tsuneyoshi M. A comparative study of fibrous dysplasia and osteofibrous dysplasia with regard to expressions of c-fos and c-jun products and bone matrix proteins: A clinicopathologic review and immunohistochemical study of c-fos, c-jun, type I collagen, osteonectin, osteopontin, and osteocalcin. Hum Pathol 1999;30:1418-26.
19. Hasselblatt M, Jundt G, Greiner C, Rama B, Schmäl F, Iglesias-Rozas JR, *et al*. Juvenile psammomatoid ossifying fibroma of the neurocranium. Report of four cases. J Neurosurg 2005;102:1151-4.
20. Noudel R, Chauvet E, Cahn V, Mérol JC, Chays A, Rousseaux P. Transcranial resection of a large sinonasal juvenile psammomatoid ossifying fibroma. Childs Nerv Syst 2009;25:1115-20.
21. Granados R, Carrillo R, Nájera L, García-Villanueva M, Patrón M. Psammomatoid ossifying fibromas: Immunohistochemical analysis and differential diagnosis with psammomatous meningiomas of craniofacial bones. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2006;101:614-9.
22. Sciubba JJ, Fantasia JE, Kahn LB, editors. Atlas of Tumor Pathology: Tumors and Cysts of the Jaw. 3rd series: Washington, D.C: Armed Forces Institute of Pathology; 2001. p. 147-51.
23. Zadik YW, Aktas A, Drucker S, Nitzan DW. Aneurysmal bone cyst of mandibular condyle: A rare case report and review of literature. J Craniomaxillofac Surg 2012;40:e243-8.
24. Oliveira AM, Chou MM, Perez-Atayde AR, Rosenberg AE. Aneurysmal bone cyst: A neoplasm driven by upregulation of the USP6 oncogene. J Clin Oncol 2006;24:e1.
25. Margo CE, Ragsdale BD, Perman KI, Zimmerman LE, Sweet DE. Psammomatoid (juvenile) ossifying fibroma of the orbit. Ophthalmology 1985;92:150-9.

Cite this article as: Shruthi SK, Kamath VV, Hegde S, Sreevidya B. Psammous desmo-osteoblastoma with concomitant aneurysmal bone cyst of mandible. Ann Maxillofac Surg 2015;5:130-4.

Source of Support: Nil, **Conflict of Interest**: None declared.