

# Epithelioid osteoblastoma of maxilla: A rare and aggressive variant of a benign neoplasm at an uncommon site

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## Abstract

Osteoblastoma (OB) is an uncommon benign bone-forming tumor accounting for <1% of all bone neoplasms. Unlike conventional OB, its small subset variant “Epithelioid osteoblastoma (EO)” is characterized by its propensity for local invasion and recurrent behavior. This rare variant of an uncommon tumor when occurs in an atypical site can lead to diagnostic problems more so due to ambiguous clinico-radiologic presentation. This was what faced in the present case of 18-year-old female with a swelling in upper jaw. OB is usually more common in males and involves primarily the posterior element of the spine and the sacrum (40–55%). Less frequently, long bones of limbs are involved. Clinical, radiological and histopathological correlation in this case guided us to reach at right diagnosis of EO which helped the patient in getting correct treatment which involves surgical excision over conventional curettage. The purpose behind this case presentation is to improve the awareness about this recurrent tumor variant which has many close differentials including well-differentiated osteoblastic osteosarcoma.

**Key Words:** Aggressive, epithelioid cells, maxilla, osteoblastoma, osteosarcoma

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## INTRODUCTION

Epithelioid osteoblastoma (EO) is an uncommon histologic variant of osteoblastoma (OB) and derives its name from the characteristic presence of plump epithelioid osteoblasts. EO is also named as aggressive osteoblastoma (AO) due to its propensity for local invasion and high rate of recurrence.<sup>[1,2]</sup> Conventional osteoblastoma (CO) is an uncommon benign bone-forming tumor accounting for <1% of all bone neoplasms. It most commonly affects males during the second decade of life with vertebral column and pelvis being common sites and involves long bones of limbs infrequently.<sup>[3,4]</sup> About 10–12% of CO can occur in maxillofacial skeleton with mandible being the most common site.<sup>[4,5]</sup> Less than hundred cases of OB of jaw

have been reported in literature, with very few being of AO.<sup>[1]</sup> We report an interesting and rare case of EO involving maxilla in an 18-year-old female patient which challenged us and threw many differentials. It needed close clinical, radiological and histological correlation to reach at correct diagnosis.

## CASE REPORT

An 18-year-old girl reported to our institution with a complaint of slowly progressive palatal swelling for last 1 year on right side of her upper jaw in maxillary molar region. There was a history of gradual loosening of teeth from #13 to #17 along with mild pain. There was no history of trauma or tooth

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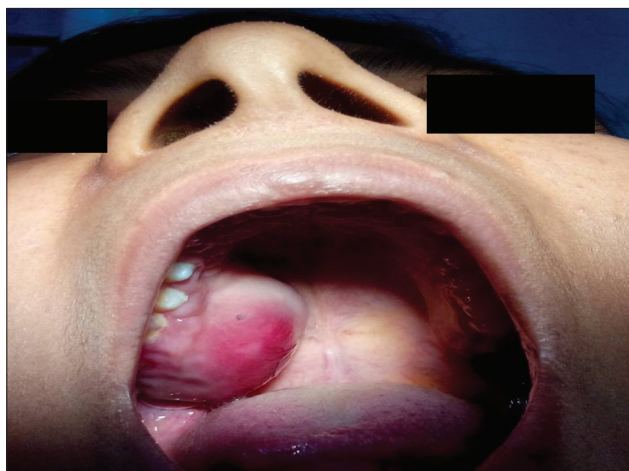
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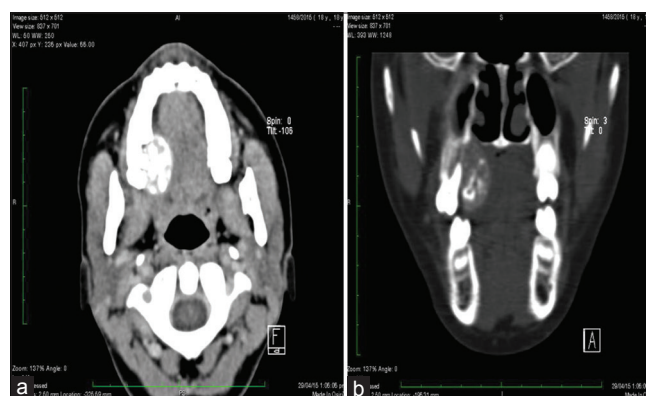
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extraction. On examination, a diffuse purplish-red growth measuring approximately 4.2 cm in diameter was seen which was mildly tender, nonreducible, nonpulsatile and did not bleed on touch [Figure 1]. There were no associated nasal, ocular or neurological symptoms. Rest of the systemic examination was normal. Computed tomography scan revealed a well-defined, smoothly margined, lobulated expansile lytic lesion involving the right side of maxilla at the level of right first, second and third molar along the upper inner alveolar margin. The lesion was seen to partially erode and encase the posterior root of the second molar tooth. The lesion had coarse, pleomorphic calcific matrix with regions of ossification within [Figure 2]. Mucosal bulge in maxillary sinus and oral cavity was present with no involvement or ulceration. Posteriorly, the lesion was seen abutting the inferior opening of sphenopalatine foramen. The pterygoid plates were uninvolved. Possibilities considered

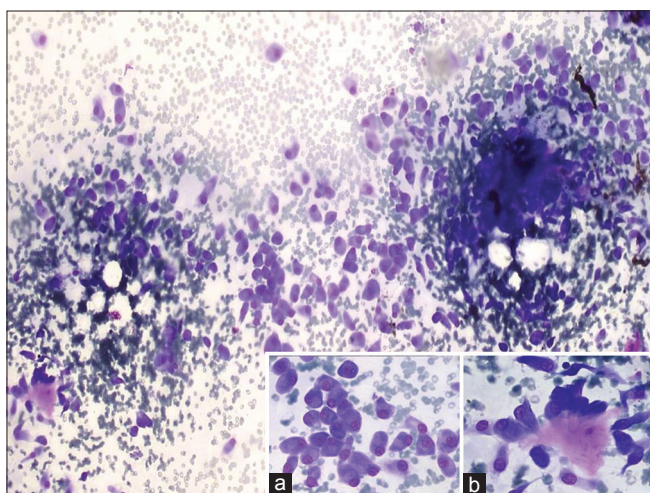
on imaging include ossifying fibroma and central giant cell granuloma. Fine-needle aspiration cytology (FNAC) of the lesion was done and smears revealed monomorphic population of discohesive plasmacytoid epithelial cells with abundant cytoplasm, well-defined cell borders and eccentric round to oval nucleus with occasional nucleoli. Few cells also showed presence of perinuclear huff consistent with Golgi apparatus. Some cells were more pleomorphic with coarse chromatin and prominent nucleoli. At few places, these cells were seen lining eosinophilic acellular material [Figure 3]. Multinucleated osteoclasts such as giant cells were also appreciated. No inflammatory cells, necrotic material or mitotic figures were seen. FNAC pointed towards benign proliferative lesion and an osteoblastic neoplasm was favored. Excision of the lesion was preferred over curettage. Cut section of the lesion on gross was red brown with gritty consistency [Figure 4]. Histopathology revealed sheets of



**Figure 1:** Clinical image shows intraoral swelling in the right side of upper jaw in maxillary molar region



**Figure 2:** (a) Contrast-enhanced computed tomography axial scan shows an eccentric expansile lytic mass arising from the medial aspect of right maxillary alveolus in the posterior molar region with foci of ossification within. The mass involves both the medial cortex and the medullary cavity. (b) Bone window in contrast-enhanced computed tomography coronal reformat shows the superior surface of the right maxillary alveolar mass abutting the right lateral aspect of the hard palate and inferior wall of ipsilateral maxillary sinus



**Figure 3:** Fine-needle aspiration cytology smear shows population of plasmacytoid cells, which at places are surrounding eosinophilic acellular material (LG stain,  $\times 100$ ). [Inset: a: High power view of plasmacytoid cells (LG stain,  $\times 400$ ), b: High power view of eosinophilic acellular material (LG stain,  $\times 400$ )] LG stain: Leishman Giemsa stain



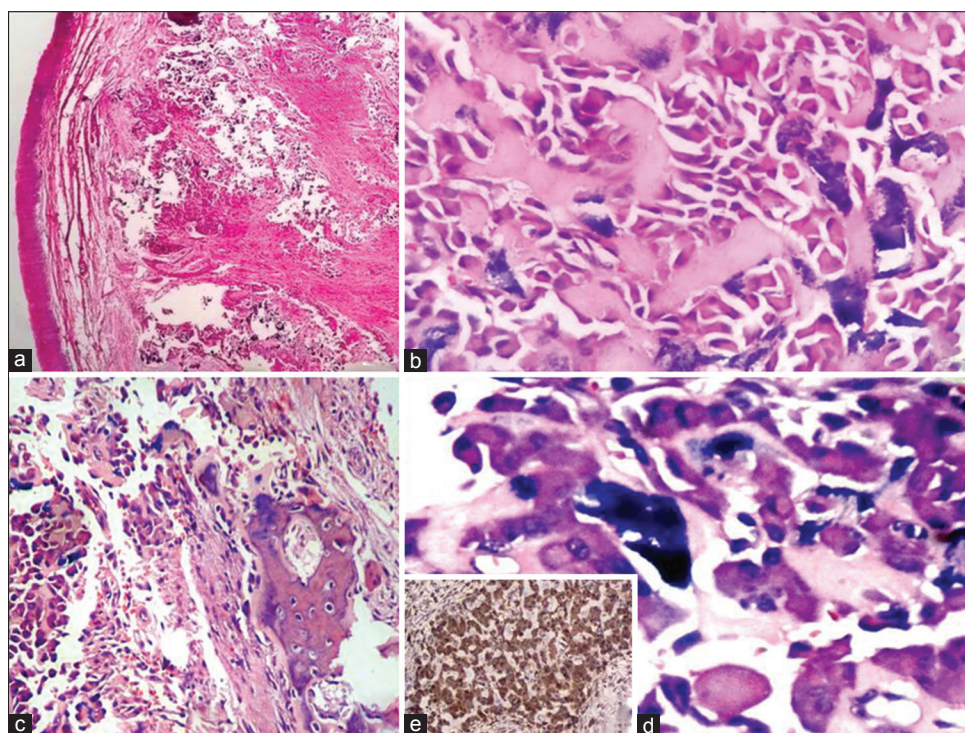
**Figure 4:** Gross image of excised specimen showing red-brown and gritty cut surface

large epithelioid cells with eccentric nucleus and abundant cytoplasm, rimming the irregular deposits of osteoid and broad bony trabeculae [Figure 5]. Few osteoclast-like multinucleated giant cells were present. Peripheral maturation of “blue bone” to more organized eosinophilic trabeculae of woven bone was seen in the margins of lesion. No abnormal mitotic figures or area of necrosis were seen. The host lamellar bone was devoid of any invasion. The diagnosis of EO was given and patient was kept in close follow-up due to aggressive nature of lesion.

## DISCUSSION

OB is a benign bone-forming tumor. The lesion was first described by Jaffe and Mayer in 1932 and has had many synonyms including giant osteogenic fibroma and giant osteoid osteoma. In 1956, Jaffe and Lichtenstein separately proposed the term “benign OB” for an osteoblastic osteoid-forming lesion similar to osteoid osteoma, but with a greater growth potential.<sup>[2]</sup> OB accounts for <1% of all primary bone tumors, with a male:female ratio of 2:1. It usually occurs in young adults, with mean age of 20 years and primarily involves the posterior element of the spine and the sacrum (40–55%). Less frequently long bones of limbs are involved.<sup>[3,4]</sup> Dorfman and Weiss in 1984 described a subset of OB which was characterized by locally destructive pattern, recurrence and presence of epithelioid osteoblasts and labeled them as “AO.”<sup>[6]</sup>

Approximately 10–12% of CO occurs in maxillofacial skeleton with mandible being the commonest site as reported by Alvares Capellozza *et al.* while reviewing largest series of OB of jaw.<sup>[5,7]</sup> In this series, 7.2% cases were noted as recurrent however, no confirmatory histological features for AO were described.<sup>[7]</sup> Reports of gnathic AO are extremely rare. Our case had involvement of upper jaw with swelling of 4.2 cm in diameter. AO lesions tend to be clinically and radiographically larger ( $\geq 4$  cm) than lesions of CO ( $< 4$  cm).<sup>[5]</sup> The radiologic appearance of AO can be similar to CO, consisting of circumscribed lytic defect sometimes surrounded by sclerotic rim although a more aggressive appearance with cortical expansion and local destruction can be seen.<sup>[4,8]</sup> Imaging of our case revealed expansile lytic lesion involving the cortex and medullary cavity of right maxillary alveolus, eroding and encasing the posterior root of the second molar tooth. Excision with partial maxillectomy was preferred over curettage because of expansile and lytic nature of lesion and keeping in mind the possibility of osteogenic sarcoma. Few authors in literature have quoted that patients of AO are usually in older age bracket than CO and are seen in the third or fourth decade.<sup>[2,4]</sup> However, our patient was 18 years old at time of presentation. Differential usually considered in cases of jaw tumor includes fibro-osseous lesions, bone tumors and odontogenic tumors. The differential offered on imaging in our case included ossifying fibroma and central giant cell granuloma, these lesions being



**Figure 5:** Histopathological image shows (a) mucosa covered unencapsulated circumscribed lesion comprising of sheets of epithelioid osteoblasts (H&E stain,  $\times 100$ ) (b) rimming irregular, broad osteoid (H&E stain,  $\times 400$ ) (c and d) Peripheral maturation and presence of blue bone. (c) H&E stain,  $\times 100$ , (d) H&E stain,  $\times 400$  and (e) cells showing S-100 positivity

more common in this site and age group. Juvenile ossifying fibroma can have aggressive behavior.<sup>[9]</sup> OB being rare in this region was kept low on differentials. Osteoid osteoma which could be another close differential is usually smaller in size, seldom exceeding 1 cm in greatest diameter and are associated with characteristic pain and surrounding sclerotic bone reaction.<sup>[10]</sup>

The plasmacytoid cells on aspiration smears pointed toward osteoblastic origin and had plasma cells and myoepithelial cells of salivary gland origin as close morphologic differentials. The possibilities were narrowed to osteoid osteoma, OB and low-grade osteosarcoma after close clinicopathological and radiological correlation. Myoepitheliomas of salivary gland origin do not present with chondroid or osteoid formation.<sup>[11]</sup>

Histopathological examination is the gold standard investigation for a definitive diagnosis. Surgical excision with wide local margins and reconstruction was done in our case. The presence of sheets of characteristic large epithelioid osteoblasts rimming the osteoid and irregular broad bony trabeculae helps us in reaching the diagnosis. Peripheral maturation or zonation to organized trabeculae of eosinophilic woven bone was also seen. OB-like osteosarcoma is the main differential of AO. The absence of lace-like osteoid, bizarre cells and prominent mitotic activity helped in excluding diagnosis of osteosarcoma.<sup>[2]</sup> According to Bertoni *et al.*, the chief microscopic feature separating osteosarcoma from OB is lack of tumor maturation at the margins of osteosarcoma, with permeation of tumor in adjacent host tissue.<sup>[12]</sup>

In a review of 306 cases of OB, Lucas *et al.* concluded that the clinically aggressive behavior in OB did not seem dependent totally on the presence of epithelioid osteoblasts but appear more related to precise location and size of the tumor which impact the surgeon's ability to completely remove the lesional tissue.<sup>[2,4,8]</sup> Considering the recurrence rate of 13.6% for OB and nearly 50% in AO, surgical resection should be preferred over curettage.<sup>[1,13]</sup> Our patient was treated with surgical resection and is on close follow-up with no complaints of recurrence.

## CONCLUSION

AO is a rare tumor in the oral and maxillofacial region.

Proper diagnosis and management of AO requires close and careful correlation of clinical, radiographic and histopathological findings.

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## Conflicts of interest

There are no conflicts of interest.

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