CASE REPORT

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Aggressive osteoblastoma of the maxilla-a diagnostic conundrum

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ABSTRACT

Osteoblastomas are biologically benign tumors with limited growth potential and may show a wide range of clinical, radiographic and histopathological findings. Osteoblastomas of the jaw may resemble other gnathic lesions including osteoid osteoma, cementoblastoma, cement-ossifying fibroma, cemento-osseous dysplasia and osteosarcoma, offering a unique diagnostic conundrum. Aggressive osteoblastoma, a rare variant of osteoblastoma characterized by the presence of atypical histopathological features like epithelioid osteoblasts and exhibiting locally aggressive behavior is rarely reported in the jaw bones. The accurate diagnosis and delineation of this entity from malignant bone tumors sometimes may be a challenge. The case report emphasizes this entity, which has to be differentiated from low- grade osteosarcoma and highlights the gravity of clinico-pathologic correlation in such lesions.

Key Words: Aggressive osteoblastoma, osteoblastoma, maxilla, epithelioid osteoblasts

Introduction

Osteoblastoma is an uncommon bone forming tumor accounting to 1% of all bone tumors, commonly found in vertebral columns and long bones. It accounts to 15% of cases in maxillofacial skeleton with greater frequency occurring in the mandible (1). The spectrum of osteoblastoma includes lesions characterized by trabeculae of newly formed osteoid lined by plump osteoblasts in a vascular background and a variant with high cellularity, prominent blue osteoid, epithelioid osteoblasts and mitosis designated as atypical osteoblastoma or aggressive osteoblastoma (AO). This subset of osteoblastoma has a locally aggressive behavior with high rate of recurrence but without metastatic potential (2). Dorfman and Weiss described aggressive osteoblastoma as borderline osteoblastic tumor with characteristic epithelioid osteoblasts (3). Microscopic features like focal presence of chondroid matrix, fine lace-like osteoid and mitosis necessitates a detailed clinical, radiographic and histopathological work up to rule out malignancy and is often arduous. Aggressive osteoblastoma is rarely reported in the jaw bones (4-7) with very few cases being reported in the maxilla (8,9). Here, we report a case of aggressive osteoblastoma of the maxilla in a 7-yr old boy.

Case report

A 7-yr old boy presented with a slow growing painless swelling on left upper jaw for one year. Examination revealed a large, non-tender, hard swelling measuring 3x4 cm on the palatal aspect extending from maxillary left deciduous lateral incisor to second molar (Figure 1a). General examination revealed no other abnormality. Orthopantomograph revealed well trabeculated radiopaque mass with radiolucent rim. Computed Tomography revealed a well circumscribed predominantly lytic expansile lesion around 3x4 cm with speckled calcifications (Figure 1b). Though a benign osteoblastic lesion was considered, the concern here was to rule out malignancy. An incisional biopsy disclosed a highly cellular lesional tissue composed of sheets of minimally pleomorphic epithelioid osteoblasts, osteoclast like giant cells with abundant osteoid formation. A striking presence of immature spiculated blue bone was noted with focal chondroid areas. The periphery showed transformation of blue bone to a more organized

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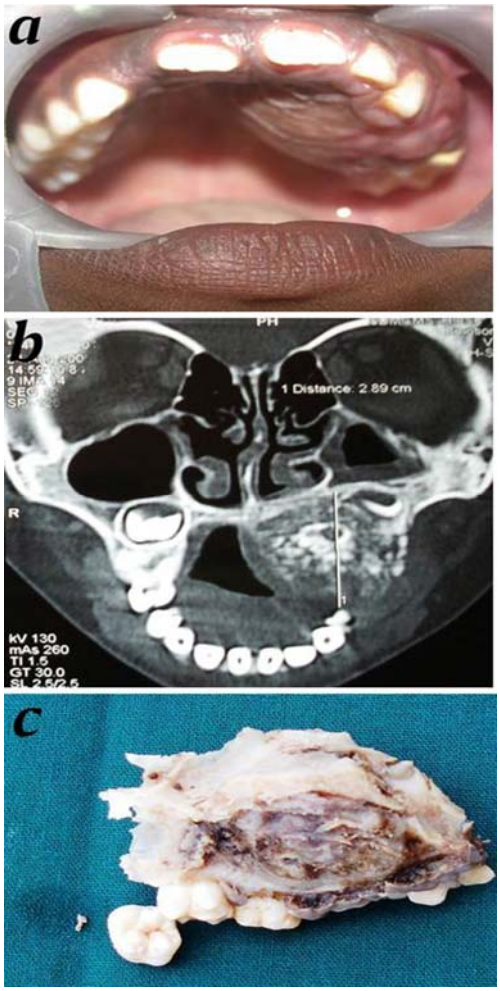

Fig. 1. Clinical photograph depicting the swelling extending from left deciduous lateral incisor to second molar (a); Computed tomogram showing lytic expansile lesion with speckled calcifications (b); photograph showing the resected specimen (c).

trabeculae of woven bone (Figures 2 & 3) with no evidence of permeation into the host bone. The clinical, radiographic and histomorphologic features were most consistent with a diagnosis of aggressive osteoblastoma. The patient underwent a segmental resection and primary reconstruction (Figure 1c). Excisional histopathology confirmed our diagnosis. Patient had an uneventful postoperative course; a two years follow-up showed no evidence of recurrence.

Discussion

The term "Aggressive Osteoblastoma" (AO), was first used by Dorfman to describe a rare bone neoplasm that composed of prominent epithelioid cells, demonstrating locally aggressive growth and high recurrence rate but no metastatic potential (10). In the jaw bones, this entity is extremely rare with only few cases being reported till date to which we add another case (Table 1) (4-9). Extragnathic AO tend to occur in the third to fourth decade, however, most reported cases in the jaws show a predilection for the second and third decades and exhibit a male predominance (4-9,11,12). They demonstrate local destruction, rapid growth and present as expansile osteolytic lesion with focal radiopacities (4). The overlapping histomorphologic features and limited published criteria about this tumor makes differentiating an AO from other osteoid producing lesions a considerable challenge.

The histopathologic differential diagnosis of an osteoblastic lesion includes osteoid osteoma,

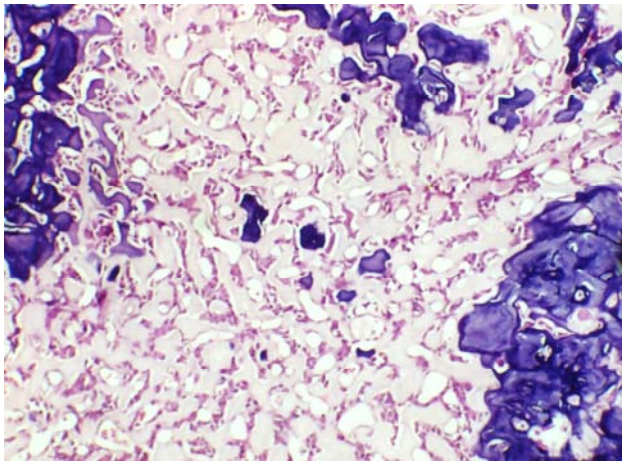

Fig. 2. Photomicrograph showing osteoid areas, spiculated blue bone and epithelioid osteoblasts (hematoxylin and eosin, original magnification X100).

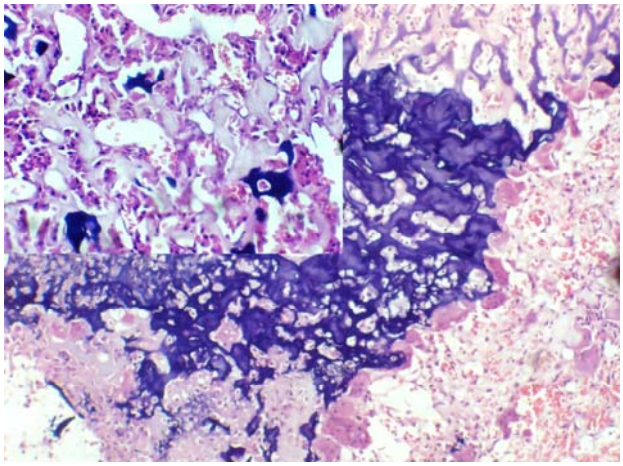

Fig. 3. Photomicrograph showing osteoid areas, spiculated blue bone, osteoclast like giant cells and left lower corner showing chondroid areas. (hematoxylin and eosin, original magnification X100). Inset: Showing osteoid areas and epithelioid osteoblasts (hematoxylin and eosin, original magnification X 250).

Table 1. Clinicopathological features of reported cases of aggressive osteoblastoma of the jaws

Case	Age (years)	Sex	Site	Histopathology	Symptoms and Duration	Treatment	Behavior
Vigneswaran N et al.[4]	21	M	Mandible	Solid sheets of epithelioid cells, amorphous osteoid with foci of calcification	Swelling and Pain , 8 months	Resection with wide surgical margins	No recurrence 2 yr follow up
Vigneswaran N et al. [4]	12	F	Mandible	Solid sheets of epithelioid cells, amorphous osteoid with foci of calcification	Swelling	Resection with wide surgical margins	No recurrence 2 yr follow up
Lypka MA et al.[5]	33	M	Mandible	Sheets of epithelioid cells, calcified matrix	Swelling & pain, 1 year	Segmental resection	No recurrence 1 yr follow up
Ohkubo T et al.[8]	6	M	Maxilla	Sheets of epithelioid cells, partially calcified osteoid material	Painful swelling, 6 months	En bloc resection	No recurrence 4 yr follow up
Harrington C et al. [9]	25	M	Maxilla	Sheets of epithelioid osteoblasts, giant cells, transformation of blue bone to organized eosinophilic trabeculae of woven bone	Asymptomatic, several months	Partial Maxillectomy	No recurrence 8 months follow up
DPV et al.[7]	25	M	Mandible	Woven osteoid and trabeculae of varying degree of mineralization, osteoblasts with eosinophilic cytoplasm and eccentric nuclei, mitotic figures, giant cells	Painful swelling, 8 months	Resection with continuity defect	No recurrence 15 month follow up
Our Case	7	M	Maxilla	Sheets of epithelioid cells, giant cells, spiculated blue bone with focal chondroid area.	Swelling, 1 year	Segmental resection	No recurrence two years follow up

cemento-osseous dysplasia, cementoblastoma, cement-ossifying fibroma and osteosarcoma. The nidus of osteoid osteoma consists of highly vascularized fibrous stroma with interconnected trabeculae of osteoid and woven bone lined by osteoblasts and osteoclasts. The lesion though having a close histological resemblance to osteoblastoma, is usually < 2 cm in diameter exhibiting dull aching pain with an exacerbation at night and being relieved by salicylates (11) Focal cement-osseous dysplasia predominantly involves the body of the mandible and presents as a radioopacity surrounded by a rim of radiolucency. Histologically it is characterized by interlacing trabeculae similar to osteoblastoma with other forms of mineralization resembling cellular cementum with spindled cellular connective tissue that may be well vascularized. Unlike osteoblastoma, plump osteoblasts are not a component of the lesion.[13] Cementoblastoma, being considered a true odontogenic neoplasm appears as a radiopaque mass fused to the roots (1). Histopathologically they reveal interlacing trabeculae of mineralized tissue resembling cellular cementum having an eosinophilic appearance compared to the characteristic

basophilic trabeculae seen in osteobalstoma. Cells are plump, angulated and plasmacytoid similar to osteoblastoma but fusion of calcified component to resorbed root of an adjacent tooth is evident (13). Central ossifying fibroma is another osteoid forming lesion characterized by well-developed connective tissue capsule and relatively uniform cellular proliferation of spindle shaped fibroblastic cells with irregular shaped trabeculae of either woven bone or cellular cementum (9,13). Conventional osteoblastoma exhibits limited growth potential with similar histopathological features, osteoblasts having moderate amount of cytoplasm, absence of mitotic figures and cellular atypia (9). The most significant of the lesions in the differential diagnosis of aggressive osteoblastoma is the low grade osteosarcoma. AO histologically is characterized by non-infiltrative sheets of epithelioid osteoblasts, giant cells of osteoclast type, lace like osteoid production and presence of spiculated blue bone (4). However, in osteosarcoma; tumor cells additionally exhibit extensive cellular anaplasia, deeply stained cytoplasm with hyperchromatic nuclei, prominent nucleoli, abnormal mitotic figures, tumor invasion into host bone or soft tissue and necrosis which are not observed in aggressive osteoblastoma (13, 14). Although the presence of epithelioid osteoblasts is a characteristic feature of AO, they are not a reliable indicator of its biologic behavior, which rather could be dependent on the location and size of the lesion. The presence of epithelioid osteoblasts and its correlation with aggressive behavior and recurrence has not been substantiated (15-17). Presence of chondroid matrix has been infrequently reported in osteoblastoma[17] and was an interesting feature observed in our case. The presence of cartilaginous matrix in an osteoid forming bone tumor does not necessarily rule out the diagnosis of osteoblastoma, nor does it necessarily support the diagnosis of osteosarcoma. Osteoblastomas may reveal areas of metaplastic hyaline cartilage after several recurrences, but in our case it was seen at the first presentation and therefore could be a part of the tumor. No significant difference in the prognosis has been noted in osteoblastomas with or without cartilaginous matrix (17). Surgical resection and reconstruction is the recommended treatment for such lesions (5). Most cases in the jaws showed no evidence of recurrence after a follow up of 8 months-4 years (Table 1).

Aggressive osteoblastoma is a rare entity and due to the overlapping histopathological features in osteoid forming lesions, an accurate diagnosis warrants a thorough clinical, radiological and histopathological workup to prevent over treatment especially in young individuals.

Conflicts of interest: None declared

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