Aggressive osteoblastoma of the proximal humerus

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ABSTRACT

A nine-year-old boy presented with increasingly worsening right shoulder pain of 18 months' duration. On physical examination, there was a tender firm swelling over the right upper arm. Radiographs showed a large osteolytic lesion in the proximal humeral diaphysis, with prominent mixed acute-on-chronic periosteal reaction in a lamellar fashion. There was a pathological fracture. The lesion appeared to be radiographically aggressive in nature. Bone scintiscan showed solitary marked uptake. On-table frozen section histopathological examination of the lesion showed an osteoblastic lesion with aggressive features. Completion curettage and high speed burring of the cavity was performed. In view of the patient's young age, which required a biological solution, and potential for local recurrence, that necessitated a radiopaque filler, the lesion was packed with a calcium phosphate cement paste. The final diagnosis was osteoblastoma with aggressive features. The patient remained well on follow-up to date. The filler continues to be remodelled to native tissue and there is no evidence of local recurrence. Osteoblastoma is a relatively rare benign tumour that typically occurs in the posterior elements of the vertebral column. The humerus is a very rare site of disease in the appendicular skeleton, and poses a diagnostic dilemma which implicates the possibility of osteogenic sarcoma.

Keywords: bone tumours, osteoblastoma, osteogenic sarcoma, paediatric tumour

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INTRODUCTION

Osteoblastoma is a rare, benign, bone-forming tumour. The average age of presentation of osteoblastoma is between 15 and 20 years, with approximately 90% of patients diagnosed before the age of 30 years.[1,2] It typically occurs in the posterior elements of the vertebral column (40%–55% of cases). In the appendicular sites, the proximal femur, distal femur and proximal tibia are the most frequent sites similar in distribution to osteoid osteomas. While histologically identical to the latter, there is however little diagnostic dilemma between these two entities as lesions larger than 2 cm are considered osteoblastomas and those smaller than 2 cm are considered osteoid osteomas. Based on this criteria,

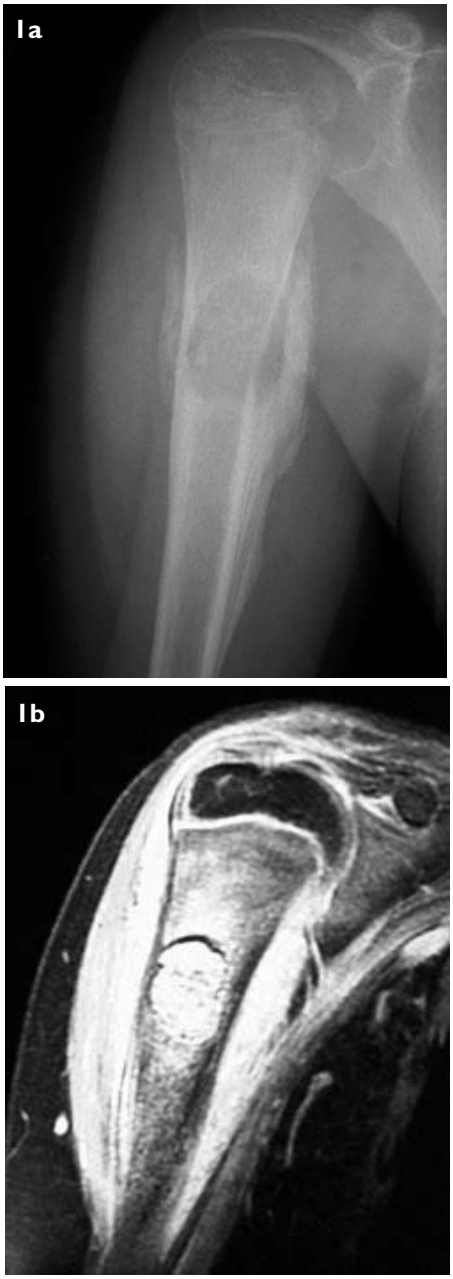

Fig. 1 (a) AP radiograph of the right proximal humerus shows a large osteolytic lesion in upper humeral diaphysis with prominent periosteal reaction. There is cortical destruction of the medial cortex and a pathological fracture. (b) Enhanced fat-saturated coronal T1-weighted MR image shows the added information of a markedly-enhancing lesion in the diaphysis with extensive bony and soft tissue oedema.

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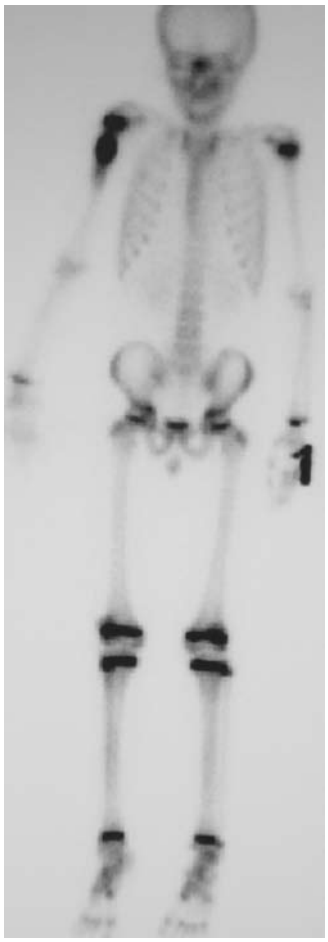

Fig. 2 99m Tc-MDP bone scan shows abnormal uptake at the right proximal humeral shaft. There is no other site of abnormal uptake.

osteoid osteomas are considerably more prevalent than osteoblastomas. The humerus is a very rare site. Despite its benign nature, the tumour may sometimes exhibit aggressive behaviour and is therefore typically treated with curettage and packed with a bone or a bone substitute. We present an unusually aggressive-appearing osteoblastoma located at the rare site of the proximal humeral diaphysis.

CASE REPORT

A nine-year-old boy presented with right shoulder pain of 18 months' duration, with recent exacerbation of pain. On physical examination, there was a tender firm swelling over the right upper arm. Shoulder abduction was painful. Radiographs showed a large osteolytic lesion located in the proximal humeral diaphysis, with prominent mixed acute-on-chronic periosteal reaction in a lamellar fashion, reminiscent of onion skin. There was a pathological fracture through the lesion but this was undisplaced (Fig. 1a). Given its aggressive radiographical appearance, the tumour was diagnosed to be an osteogenic sarcoma. A differential diagnosis was osteomyelitis. Tc-99m methylene diphosphonate (MDP) bone scintiscan showed a solitary marked uptake in the proximal humerus. Magnetic resonance (MR) imaging showed a central enhancing lesion of 3 cm diameter with a high signal on T2-weighted sequences (Fig. 1b). There was extensive oedema around the lesion. Lamellar periosteal reaction was confirmed. The overall appearance was consistent with an aggressive osteoblastic lesion in the proximal humerus most consistent in this age group with an osteogenic sarcoma. Computed tomography (CT) of the thorax was normal. Bone scintiscan showed solitary increased uptake over the proximal humerus (Fig. 2).

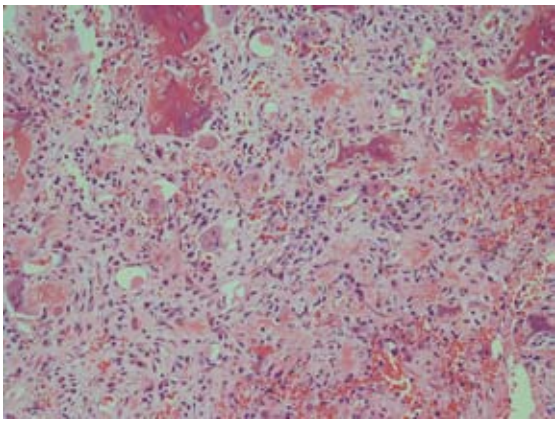

Fig. 3 Photomicrograph of the curettage specimen shows irregular to uniform, delicate trabeculae of woven bone and a lacy filigree pattern of osteoid. The bone trabeculae were rimmed and separated by layers of osteoblast-like cells with enlarged atypical nuclei and occasionally distinct nucleoli. Intermingled with the tumour cells are a variable number of osteoclasts. In some areas, the stroma appeared loose, oedematous and fibrovascular. (Hematoxylin & eosin, × 200).

Biopsy of the lesion was performed. On-table frozen section histology showed a lesion which formed irregular to uniform, delicate trabeculae of woven bone, as well as lacy filigree pattern of osteoid. Rimming and separating the bone trabeculae were layers of osteoblast cells with enlarged atypical nuclei and occasionally distinct nucleoli. Mitotic figures were present but rare and there were no atypical mitosis. The stroma appeared loose, oedematous and fibrovascular. There appeared to be a permeative growth pattern and cartilaginous differentiation. The features were felt to be suggestive of osteogenic sarcoma. At the time of surgery, this posed a dilemma – if this was a malignancy, then any attempt at an extensive currettage could disseminate the disease locally and possibly systemically. Insertion of a bioactive filler like an autogenous bone graft could result in florid growth of the remnant osteogenic sarcoma. The standard of care would be to fill such a defect with cement or a haemostatic substitute with the intention of resecting the tumour *en bloc* at a later date if this was an osteogenic sarcoma. On the other

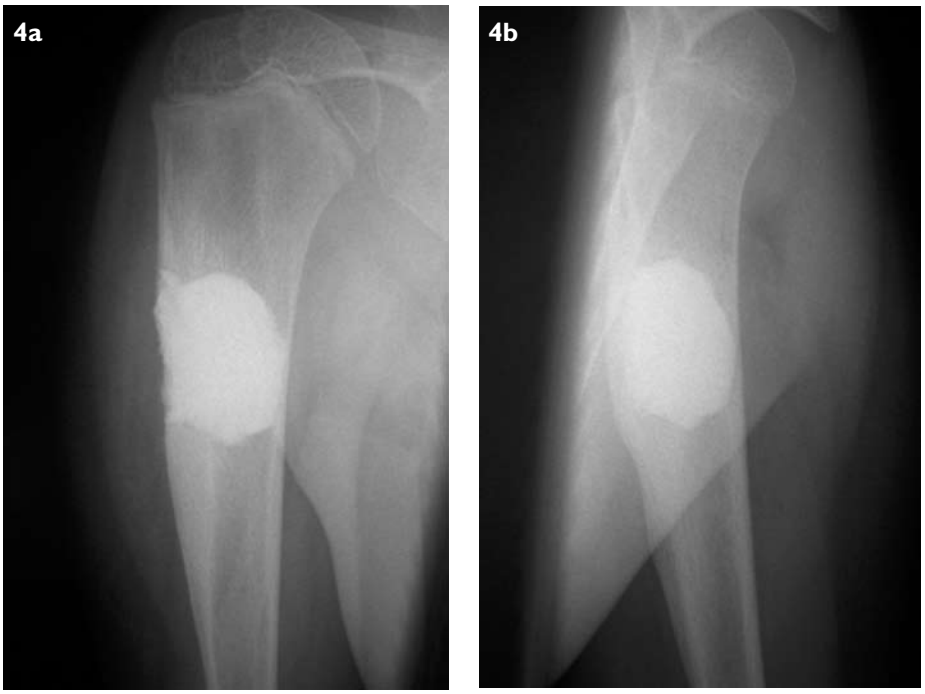

Fig. 4 Follow-up (a) AP and (b) lateral radiographs of the right proximal humerus show calcium phosphate cement filling the previous tumour site and no evidence of recurrence.

hand, if this were a benign lesion, it was aggressive at the very least and would require an extensive curettage and filling with a bioactive material. As this was a child, it would not be desirable to fill a benign defect with polymethlmethaacrylate (PMMA) as this might need to be removed at a later date. Nevertheless, the PMMA also offered a radiopaque interface on which recurrence of disease can readily be determined. With these considerations in mind, we elected to fill the defect with a calcium phosphate paste (Norian, Synthes, USA). This would provide the biological filler in the defect and still provide a radiopaque interface on which local recurrence could be determined. In the event of a diagnosis of osteogenic sarcoma, the entire lesion and filler could be resected *en bloc*.

Despite the likelihood of osteogenic sarcoma, atypical mitotic activity was not seen and there was no definite evidence of malignancy, including the permeation of pre-existing lamellar bone, when paraffin sections of the specimen were reviewed. Following an overseas consultation, the final diagnosis was that of an osteoblastoma with aggressive features (Fig. 3). The patient remained well on follow-up to date. He was followed-up closely for two years and sequential films showed a settling of the periosteal reaction, gradual resorption of the bone filler and no evidence of local recurrence (Fig. 4).

DISCUSSION

Osteoblastoma is a rare, benign, bone-forming tumour. This tumour has clinical and histological manifestations that are similar to those of osteoid osteoma. Therefore, it is also known as a giant osteoid osteoma. Males are affected approximately twice as often as females. It is a tumour of the younger population, with approximately 90% of patients diagnosed before the age of 30 years.[1,2] The most common presenting symptom of osteoblastoma is the insidious onset of a dull, aching pain. Unlike the pain of osteoid osteoma, the pain of osteoblastoma may not necessarily be nocturnal and is not readily relieved by salicylates.[3] Although osteoblastoma has been reported in a variety of skeletal locations, there is a strong predilection for the axial skeleton, particularly the posterior elements of the vertebral column (40%–55% of cases). In the appendicular sites, the proximal femur, distal femur and proximal tibia are the most frequent sites. The humerus is a very rare site. There is a slight predominance of metaphyseal over diaphyseal lesions, with very few lesions reported in an epiphyseal location.[4]

Radiographs are the most helpful single imaging technique for diagnosing osteoblastoma. Within the long bones, osteoblastoma is seen as a round or oval well-circumscribed osteolytic lesion arising in the diaphysis or metaphysis, with a thin shell of peripheral new bone. The lesion may or may not have calcification of varying degrees. The lesion is larger than 2.0 cm in diameter, and unlike osteoid osteoma, there is usually no surrounding large reactive zone of bone. Approximately two-thirds of osteoblastomas in tubular bones occur within the cortex and the remaining one-third appear within the medullary canal. Osteoblastoma may have

features similar to those of malignancy, such as cortical destruction and extraosseous soft tissue expansion.[5-7] The other imaging techniques are less specific compared to radiographs. CT provides information about the size and extent of the lesion in the cortical bone, and aids preoperative evaluation and planning for surgery. MR imaging is a helpful imaging tool in depicting the extent of the lesion within the medulla and also any soft tissue involvement. Bone scintigraphy is sensitive but not specific. It shows intense focal activity at the tumour site.

An aggressive type of osteoblastoma has been described that has characteristics similar to those of osteogenic sarcoma.[8,9] Our patient presented with clinical and radiographical features that were suggestive of an aggressive tumour, mimicking an osteogenic sarcoma. In our case, the osteolytic lesion was medullary-based, and showed cortical destruction and a florid aggressive-appearing periosteal reaction. The location in the proximal humeral shaft was typical for an osteogenic sarcoma and very unusual for an osteoblastoma. Our patient did not have radiographical features of the other differential diagnosis of osteoblastoma, including giant cell tumour and aneurysmal bone cyst.[6]

Osteoblastoma has been characterised histologically as a cellular osteoblastic tissue with active intercellular production of osteoid material and primitive woven bone. Immature bony trabeculae are lined with osteoblasts. Some of these trabeculae may have extensive ossification, whereas others may be without mineralisation. The stroma contains widely dilated capillaries and areas of large dilated blood sinusoids. Mitotic activity is nil to minimal in 89% of cases.[8] Osteoblastoma has a very low rate of mitosis, minimal cytologic atypia, has a tendency towards peripheral maturation and does not permeate surrounding bone. It rarely has the cartilaginous matrix that can be present with osteogenic sarcoma. These histological features are useful in differentiating osteoblastoma and osteoid osteoma from osteogenic sarcoma. Osteoblastoma should be differentiated from conventional osteogenic sarcoma not only because of its peculiar histological pattern, but also because of its different clinical and radiological features and much better prognosis.

Osteoblastomas should be treated surgically when detected, as the tumour may exhibit aggressive behaviour despite its benign nature, as in our case. Resection with wide margins is preferred for the lesions in expendable bones such as the rib and fibula, but an extended intralesional curettage is sufficient for lesions in most other locations.[9] Reconstruction may be required in cases where surgical excision or curettage creates a sizable defect.[9] Close follow-up is warranted for early detection of recurrence, as well as to assure appropriate rehabilitation and recovery. The prognosis of this tumour is very good, regardless of the type of treatment performed, although both relapse and, more rarely, malignant tumour evolution is possible.[10] Thorough postoperative follow-up is necessary to ensure an optimal outcome as the osteoblastoma may recur after incomplete removal. Conventional osteoblastoma has a reported recurrence of approximately 10%–20%.[6,11,12] Neither chemotherapy nor postoperative radiation therapy is indicated.[1]

In summary, we have presented a case of osteoblastoma with unusually aggressive radiographical appearances that is located at a very unusual site in the proximal humerus. This tumour mimicked an osteosarcoma clinically, radiographically and on frozen section histology. This case report reinforces the dictum that the diagnosis and treatment of aggressive osteoblastoma and bone tumours in general require the coordinated participation of dedicated musculoskeletal radiologists, bone pathologists and oncologic orthopaedic surgeons. Once the diagnosis of this tumour is made, the management and outcome are generally good.

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