Journal of Bone Oncology 2 (2013) 89–91

Contents lists available at ScienceDirect

Journal of Bone Oncology

journal homepage: www.elsevier.com/locate/jbo

Case Report

The rare aggressive osteoblastoma in a two year old child in an unusual localization

Debdutta Chatterjee [a], Kiran Kumar Mukhopadhyay [b,*], Sanjay Kumar [b], Sudipta Chakraborty [c]

[a] Bankura Sammilani Medical College, Bankura, West Bengal, India
[b] N R S Medical College Hospital, Kolkata, India
[c] ESI PGI MSR, Maniktala, Kolkata, India

ARTICLE INFO

Article history:
Received 8 November 2012
Received in revised form
20 January 2013
Accepted 9 February 2013
Available online 7 March 2013

Keywords:
Aggressive osteoblastoma
Right 3rd metacarpal shaft
Clinico-radiological feature
Histolopathological feature
Surgical management

ABSTRACT

A rare bone tumor of 3rd metacarpal bone in a male child aged 2 years is being described. The patent presented with a gradually increasing, painful swelling over the dorsum of right hand which radiologically revealed an expansile, radioluscent mass, in 3rd metacarpal shaft with cortical destruction. The clinic-radiological differential diagnosis was aggressive cartilage tumor or an osteomyelitis. Histological examination of surgical biopsy material revealed randomly arranged woven bone lined by epithelioid osteoblast and after correlating the clinic-radiological features the diagnosis was an aggressive osteoblastoma. Appropriate diagnosis of such a rare tumor in an unusual location and age group facilitate adequate management by surgery alone without radiotherapy or chemotherapy.

1. Introduction

Osteoblastoma is a rare primary benign neoplasm comprising less than 1% of primary tumors of bone. The age of presentation ranges between 3 and 70 years with approximately 90% of patients diagnosed before the age of 30 years [1]. About 32% of patients develop in the posterior elements of the spine, 12% in the femur, 10% in the tibia, and 9% in the bones of the foot and ankle [2]. Unusual sites included ribs, small bones of the hands and feet, facial bones, calvarium, patella, scapula, and ilium. In some situation, the tumor may reveal aggressive clinical and radiological behavior. Aggressive osteoblastoma is a variant which histologically mimics an osteosarcoma [2]—hence appropriate clinico-radiological correlation is essential to avoid a misdiagnosis. One such case which occurred in an unusual site is being described.

2. Case report

A male child aged 2 years presented with gradually increasing swelling over the dorsum of right hand for 4 months. There was no history of trauma, fever or similar swelling over the body. The child was otherwise healthy, playful and afebrile. Local examination of right hand and wrist showed a diffuse swelling, about 4×3 cm^2 in dimension, over mid-dorsal region of hand, without any local rise of temperature (Fig. 1). However the child exhibited signs of local discomfort over the area. The overlying skin was healthy and there was no abnormality in palm. The third metacarpal shaft was poorly delineated. Fingers could be moved to the full extent. There was no local lymphadenopathy or neural deficit. Radiological examination revealed an expansile mass of 3×2.5 cm^2, in 3rd metacarpal shaft, the mass was radiolucent, divided into poorly defined small locule with focal mineralization (Fig. 2). In some areas, cortical destruction was noted. The clinico-radiologic features were indicative of an aggressive cartilage tumor or an osteomyelitis.

An incisional biopsy was undertaken. Peri-operative examination of the lesion revealed an expansile tumor mass brownish in color with focal whitish areas (Fig. 3). The lesion could be delineated from the surrounding cortical bone, and surrounding soft tissues. Histopathologically the biopsy material depicted a tumor composed of spicules of randomly arranged woven bones and are lined by a single layer of osteoblasts. The cells were two to three times larger than conventional osteoblasts, were epithelioid in morphology with large vesicular nuclei, prominent nucleoli and abundant eosinophilic cytoplasm (Fig. 4). The woven bones were focally mineralized. The stroma is rich in vascularity and scattered osteoclast-type multinucleated giant cells noted. A diagnosis of aggressive osteoblastoma was rendered after

* Correspondence to: Ruchira Residency, T-4/F-11/A4, 369 P K Main Road, Kolkata, West Bengal 700078, India. Tel.: +91 33 9433166270; fax: +91 33 23720540.
E-mail addresses: debduttachatortho@gmail.com (D. Chatterjee), orthokiran@gmail.com (K.K. Mukhopadhyay), sanjayortho@yahoo.co.in (S. Kumar), sudiptach@gmail.com (S. Chakraborty).

2212-1374/$ - see front matter © 2013 Elsevier GmbH. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).
http://dx.doi.org/10.1016/j.jbo.2013.02.001

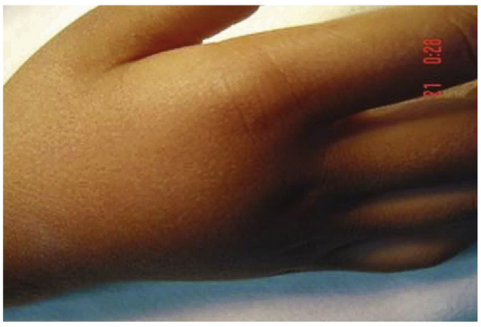

Fig. 1. Photograph of the swelling over mid-dorsum of right hand with intact skin.

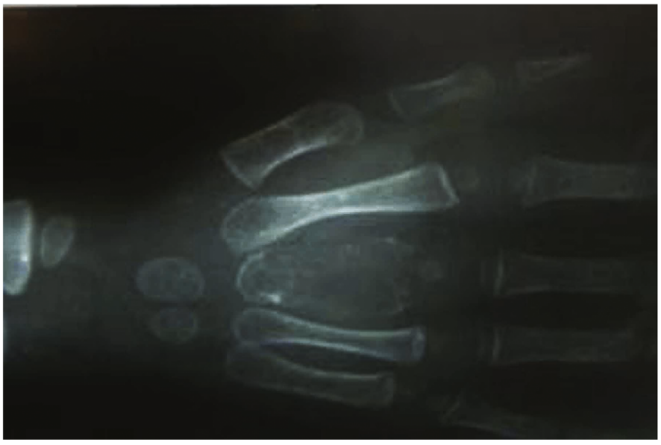

Fig. 2. X-ray of the lesion showing an expansile mass 3rd metacarpal shaft, the mass was radiolucent well defined margin, divided into smaller locule.

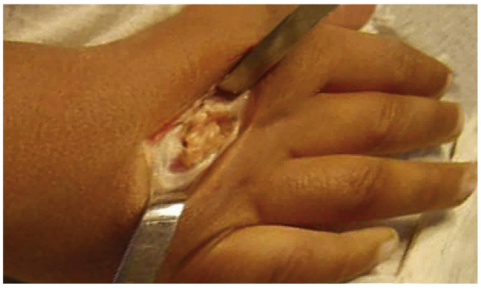

Fig. 3. Peri-operative expansile tumor mass brownish in color with focal whitish areas were noted.

considering clinical, radiological and histological features of the tumor.

Patient was treated with a ray amputation after counseling with the parents as the lesion could not be curated due to cortical involvement. Post-operative period was uneventful no recurrence was noted in a 2-year follow up.

3. Discussion

Osteoblastoma is a rare benign, bone-forming tumor, having similar clinical and histological features to those of osteoid osteoma [3]. The tumor is more commonly occurring in males (2.5:1), among the teenagers and young adults [3]. The present patient was a 2-year old male, which is not common age for occurrence of such tumor. Osteoblastoma is one of the rare neoplasms that predilects for the posterior elements of spine. In the appendicular long bones, the proximal femur, distal femur

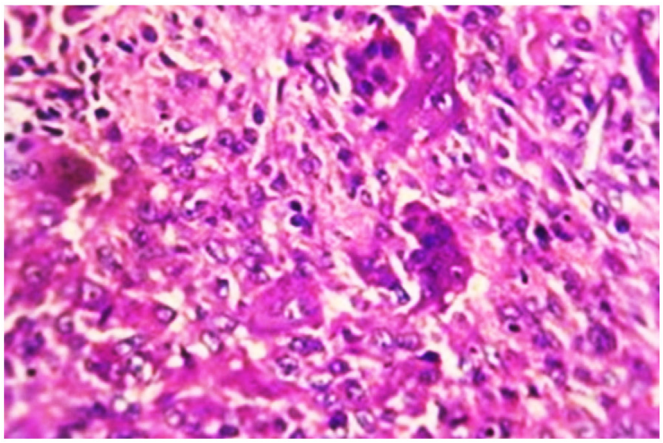

Fig. 4. Histopathological examination of the biopsy material showing a tumor with spicules of randomly arranged woven bones, large osteoblasts having large vesicular nuclei, prominent nucleoli and abundant eosinophilic cytoplasm. Scattered osteoclastic type of giant cell noted (H&E, $\times 100$).

and proximal tibia are most commonly involved. Still less commonly the tumor may involves the tarsal bones (talus and calcaneous) [3].

Patients with this tumor commonly presents with the insidious onset of a dull, aching pain which was noted in the present case. In contrast to the pain of osteoid osteoma, the pain of osteoblastoma may not always be nocturnal and is not relieved by salicylates [3,4]. Most osteoblastomas are in the range of 3–10 cm while osteoid osteomas rarely exceed 1 cm [3,4]. In the present tumor the mass was 3 cm in the greatest dimension.

Imaging studies reveals a characteristic oval to round, radiolucent lesion with scattered areas of mineralization having the central density of a trabecular bone and easily discernable margins. Mineralization varies from completely radiolucent to extensive. The present tumor had a well-defined margin and focal mineralization. Unlike osteoid osteoma, there is no surrounding dense cortical sclerosis [4]. The size and extent of the tumor in the cortical bones best evaluated by CT, hence helps in preoperative evaluation and planning for surgery. The extent of the lesion within the medulla, soft tissue, areas of cystic degeneration and hemorrhage (which may occur in a few cases) can be best perceived on MRI study [2,5]. The radiographic appearance of aggressive osteoblastoma and conventional osteoblastoma is similar. Osteoblastoma may have radiological features mimicking to those of malignant bone tumor, such as cortical destruction and extraosseous soft tissue expansion [6]. In the present case, cortical destruction was noted which radiologically mimicked an aggressive neoplasm. Khin et al. [5] reported one case of aggressive osteoblastoma in humerus where radiological features were mimicking osteosarcoma. Other radiological differential diagnoses are giant cell tumor and aneurysmal bone cyst [7].

Histologically osteoblastoma has identical features to osteoid osteoma, composed of woven bone spicules or trabeculae. These spicules are lined by a single layer of osteoblasts arranged haphazardly in a richly vascularized stroma. Osteoblasts may have mitoses but not atypical. Presence of diffusely scattered osteoclast-type of multinucleated giant cells may mimic a giant cell tumor and dilated vascular spaces may mimic an aneurysmal bone cyst. The tumor woven bone may be in aggregates or nodules and mimicking osteosarcoma.

In 1972 Dorfman incorporated osteoblastoma in an article describing malignant transformation of benign bone lesions and noted four of 23 cases of osteoblastomas displayed recurrent behavior and histologic features. These cases would merit the title "aggressive osteoblastoma," though characteristic histological

features were not specified [8]. Later, Dorfman and Weiss defined aggressive osteoblastoma as a borderline osteoblastic tumors in 1984 and proposed the presence of epithelioid osteoblasts was a characteristic histologic feature [9]. Incidence and skeletal distribution of aggressive osteoblastoma are currently not appraised as the tumor is significantly rare [10].

Aggressive osteoblastoma is characterized by epithelioid osteoblasts that are, two to three times larger than conventional osteoblasts, and have abundant eosinophilic cytoplasm, large vesicular eccentric round to oval nuclei with prominent nucleoli. Usually there are rare or absence of mitoses and if present, are normal in appearance [11]. The neoplastic bone may be trabecular or coarse and lace-like in appearance. Phenotypic markers that can help to discern an osteoblastoma over an osteosarcoma are sharp margin of the tumor and presence of osteoblastic rimming [2]. In the case of aggressive osteoblastoma, the tumor border shows peripheral maturation without sharp demarcation [12]. The border between pre-existing cortex or marrow trabeculae must be examined histologically as osteoblastomas do not infiltrate and isolate pre-existing lamellar bone structures as does osteosarcoma [3]. Moreover, significant nuclear atypia, altered nuclear-to-cytoplasmic ratios, and profuse mitoses together with atypical forms are characteristics of osteosarcoma and absent in osteoblastoma [2].

Osteoblastomas are surgically treated as the tumor may exhibit aggressive radiological appearance, despite its benign nature, as in the present case. Extended intralesional curettage is usually done in most locations while resection with wide margins is done in expendable bones like ribs and fibula [13]. Reconstruction may be required in cases where surgical excision or curettage creates a sizable defect. The present case treated with a ray amputation as cortical involvement prevented intralesional curette. Postoperative radiation or chemotherapy is not required in such tumor.

The various histologic subtypes of osteoblastoma have a similar clinical behavior, although there are some disagreements regarding the aggressive type. In most cases, osteoblastoma behaves as a benign tumor, having no metasizising capacity. However they can grow, become large, and produce bone destruction and pain [2]. Although prognosis of osteoblastoma is excellent, both relapse and malignant tumor transformation has been described, thus requiring thorough postoperative follow-up [14]. Recurrences are likely to happen in such a location which has difficult surgical access resulting in inadequate curette [3].

The present case represents an unusual aggressive variant of osteoblastoma located in 3rd metacarpal shaft—an unusual site. Both clinically and radiologically, the tumor mimicked a malignant neoplasm producing considerable diagnostic predicament. Hence diagnosis and subsequent appropriate management of the case required a harmonious contribution from orthopedic surgeon, radiologist and histopathologist. Appropriate diagnosis of such benign neoplasm of bone facilitates proper surgical management for a favorable outcome.

Conflict of Interest Statement

The authors declare that there are no conflicts of interest.

References

[1] McLeod RA, Dahlin DC, Beabout JW. The spectrum of osteoblastoma. AJR American Journal of Roentgenology 1976;126:321–5.
[2] Nielsen GP, Rosenberg AE. Bone-forming tumors. In: Folpe AL, Inwards CL, editors. Bone and soft tissue pathology (A volume in the series foundations in diagnostic pathology). 1st ed. Philadelphia, Saunders; 2010. p 309–29.
[3] Malcolm AJ, Schiller AL, Schneider-Stock R. Osteoblastoma. In: Fletcher CDM, Unni KK, Mertens F, editors. World Health Organization Classification of Tumours, International Agency for Research on Cancer (IARC), Pathology and Genetics of Tumours of Soft Tissue and Bone. Lyon: IARC Press; 2002 p. 262–263.
[4] Klein MJ, Parisien MV, Schneider-Stock R. Osteoid osteoma. In: Fletcher CDM, Unni KK, Mertens F, editors. World Health Organization Classification of Tumours, International Agency for Research on Cancer (IARC), Pathology and Genetics of Tumours of Soft Tissue and Bone. Lyon: IARC Press; 2002. p. 260–1.
[5] Khin YT, Peh WC, Lee VK, Teo HE, Ng SB, Nathan SS. Aggressive osteoblastoma of the proximal humerus. Singapore Medical Journal 2009;50:e1–4.
[6] Kroon HM, Schurmans J. Osteoblastoma: clinical and radiologic findings in 98 new cases. Radiology 1990;175:783–90.
[7] Resnick D. Tumors and tumor-like lesions of bone: imaging and pathology of specific lesions. In: Resnick D, editor. Diagnosis of bone and joint disorders. 3rd ed. Philadelphia: WB Saunders; 1995. p. 3786–96.
[8] Dorfman H. Malignant transformation of benign bone lesions. In: Proceedings—National Cancer Conference 1972;7:901–13.
[9] Dorfman H, Weiss S. Borderline osteoblastic tumors: problems in the differential diagnosis of aggressive osteoblastoma and low grade osteosarcoma. Seminars in Diagnostic Pathology 1984;1:215–34.
[10] Nishida K, Doita M, Kawahara N, Tomita K, Kurosaka M. Total en bloc spondylectomy in the treatment of aggressive osteoblastoma of the thoracic spine. Orthopedics 2008;31:403–6.
[11] Dorfman HD, Benign osteoblastic tumors. In: Bone Tumors. 1st ed. St Louis: Mosby; 1998. p. 85–127.
[12] Harrington C, Accurso BT, Kalmar JR, Iwenofu OH, Agrawal A, Allen CM, et al. Aggressive osteoblastoma of the maxilla: a case report and review of the literature. Head & Neck Pathology 2011;5:165–70.
[13] Golant A, Dormans JP. Osteoblastoma: a spectrum of presentation and treatment in pediatric population. University of Pennsylvania Orthopaedic Journal 2003;16:9–17.
[14] Cervoni L, Innocenzi G, Raguso M, Salvati M, Caruso R. Osteoblastoma of the calvaria. Neurosurgical Review 1997;20:51–4.