Case Report

Diaphyseal osteosarcoma with varying histomorphologic patterns

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Abstract

Osteosarcoma is the most common primary malignant tumor of the skeleton involving predominantly metaphysis of the long bones. Diaphyseal osteosarcoma is a rare form, which accounts for approximately 10% of all cases of osteosarcomas. Osteosarcoma contains a family of lesions with considerable diversity in histologic features and grade. We present a case of a 29-year-old male with diaphyseal osteosarcoma of the left tibia, who presented with pain and swelling over middle one-third of the left leg. The biopsy of the lesion revealed varied histomorphologic features in a small tissue studied. This prompted us to report the various histologic patterns of osteosarcoma.

Key Words: Bone tumors, diaphysis, histological pattern, osteosarcoma

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INTRODUCTION

Osteosarcoma is a high-grade malignant mesenchymal tumor characterized by formation of immature bone or osteoid by the tumor cells.[1,2] It accounts for approximately 15% of all primary bone tumors.[3] It occurs predominantly in the metaphysis of the long bones of adolescents and young adults, however, a purely diaphyseal (10%) or epiphyseal (<1%) location may occasionally be encountered.[3,4] Osteosarcomas are grouped according to site of origin (intramedullary, intracortical, or surface), their degree of differentiation, multicentricity (synchronous or metachronous), and whether they are primary or secondary to pre-existing conditions. They may also be grouped according to their histologic features as osteoblastic, chondroblastic, fibroblastic, telangiectatic, small cell, or giant cell type. We came across a case of osteosarcoma located at the diaphyseal region. Histopathological analysis of the tumor revealed very varied histological patterns in the small biopsy tissue that was sampled.

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CASE REPORT

A 29-year-old male presented with painful non-progressive swelling in the left lower leg since 3 years. He gave history of pain while walking. There was no history of trauma. Past and family history was not significant. On examination, 10 × 6 cm firm to hard swelling was noted at the middle one-third of left lower leg. The skin over the mass was unremarkable. Chest X-ray was normal [Figure 1a]. X-ray of the affected lower limb revealed large lucent defect in the medullary cavity in the mid shaft, with extensive calcification/ossification in the soft tissue

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mass [Figure 1b]. The possibility of adult Ewings sarcoma or osteosarcoma was considered. Magnetic resonance imaging (MRI) scan revealed a well defined lobulated mass with infiltration in the medullary cavity suggestive of a malignant neoplastic lesion, possibly osteosarcoma [Figure 1c and d]. Three phase bone scan revealed pathology involving left tibial shaft without distant skeletal involvement. Fine needle aspiration cytology (FNAC) showed scattered small malignant cells suspicious for Osteosarcoma or Ewings sarcoma. Biopsy was advised. Tissue sections showed sheets of malignant cells. These cells were round to oval to spindle, they had a high N:C ratio, scanty cytoplasm with hyperchromatic nuclei. Foci of malignant spindle cell proliferations were seen, at places forming hemangiopericytomatous pattern. Osteoid formation with calcification was also observed. Other areas of the tumor showed telangiectatic features and atypical chondroid formation. Active mitosis was observed. Areas with islands of malignant round cells were also seen [Figures 2 and 3]. Based on these histopathological features, a diagnosis of high grade Osteogenic Sarcoma was offered. Patient was advised neoadjuvant chemotherapy followed by surgery of amputation of left lower limb. The patient went to oncology centre for treatment and has been lost for follow up.

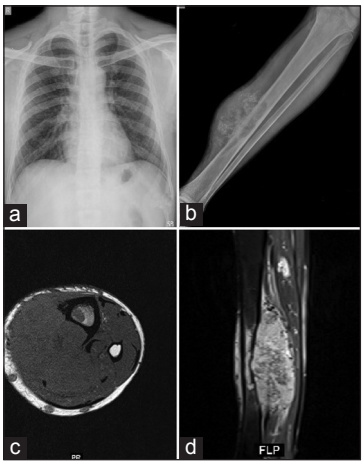

Figure 1: (a) Normal X-ray chest (b) X-ray left lower limb showing radiolucent mass in the mid shaft with extensive calcification (c and d) MRI scan showing well defined lobulated mass with infiltration in the medullary cavity

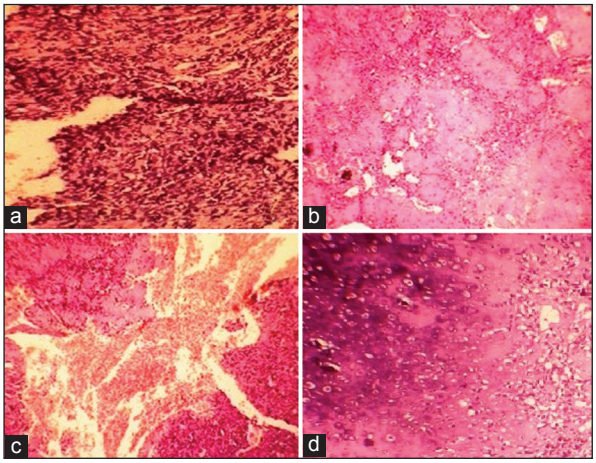

Figure 2: Photomicrograph showing (a) Malignant spindle shaped cells (b) Osteoid with pleomorphic cells (c) Large areas of hemorrhage with endothelial lined vascular areas and neoplastic cells (d) Chondroblastic areas (H&E, ×100)

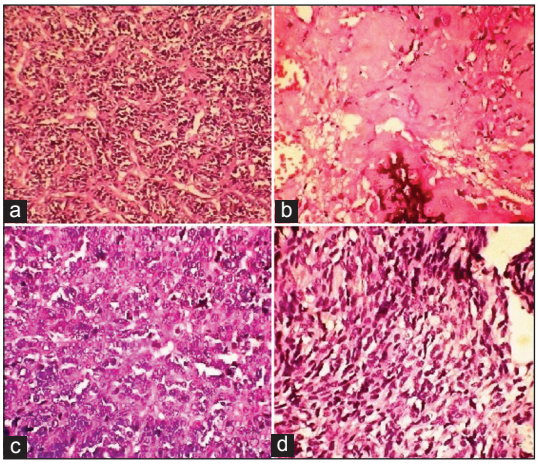

Figure 3: Photomicrograph showing (a) Vascular spaces arranged in stag horn pattern and spindle shaped cell arranged around it in hemangiopericytomatous pattern (H&E, ×100) (b) Osteoid with large pleomorphic cells (H&E, ×400) (c) Malignant small round cell pattern (H&E, ×400) (d) Fibroblastic areas (H&E, ×400)

DISCUSSION

Most of the bone tumors demonstrate a predictable "location" in the skeleton. This location is explained by "laws of field behavior and developmental anatomy" of the affected bone, a concept first popularized by L C Johnson.[3] The process of skeletal remodeling with its associated osteoblastic activity predominate in metaphysis. Hence most osteosarcomas tend to occur in this location. This remodeling is principally seen in adolescents, and interestingly this age group has the peak incidence of osteogenic sarcoma.

Conventional osteosarcoma accounts for 75-85% of all osteosarcomas.[3] It is composed of malignant pleomorphic cells that produces osteoid. These can be subclassified based on dominant tissue identified: Osteoblastic, chondroblastic, or fibroblastic.[4] Approximately 50% of all conventional osteosarcomas are osteoblastic, 25% are fibroblastic, and 25% chondroblastic. Fibroblastic variants show purely spindle cell growth with only small foci of matrix. This needs to be differentiated from fibrosarcoma. Few variants may show fibrohistiocytic differentiation, which may be mistaken for malignant fibrous histiocytoma. Sometimes, the tumor cells may grow in diffuse, nesting pseudocapillary arrangements. Vessels may be numerous, dilated and cells proliferating in

a hemangiopericytomatous appearance (staghorn type appearance). Osteosarcomas with chondroid differentiation show malignant appearing cartilage with hypercellularity and hyperchromasia. Even if clear cut osteoid production is sparse, a high grade chondroid neoplasm with sheets of spindle cells in a young patient should be considered as chondroblastic osteosarcoma.[4] Telangectatic variant, radiologically and pathologically resembles aneurysmal bone cyst.[5] Histologically, the lesion consists of vascular spaces separated by septa lined by the malignant cells. These tumor cells are spindle shaped and they also produce collagen and osteoid.[5] Multinucleated giant cells may be numerous in telangectatic osteosarcoma, but here the giant cells are not uniformly distributed and are present around the blood filled spaces. In addition, there usually is a great deal of nuclear pleomorphism evident in telangectatic osteosarcomas, which will help to differentiate it from aneurismal bone cyst.[6] In Small cell osteosarcoma, radiologic features are not consistently typical for osteosarcoma because there often is very little mineralized matrix produced. Histologically it may be mistaken for Ewing sarcoma/primitive neuroectodermal tumor because its cells are small and have round, hyperchromatic nuclei with very little nuclear pleomorphism characteristic of conventional high grade osteosarcoma. It constitutes between 1% and 2% of all osteosarcomas.[7,8] Tumors with telangectatic and small cell variant are high grade lesions, hence these varied morphologic patterns should be mentioned in the report. Osteosarcoma tends to metastasize to the lungs although, rarely, lymph nodes and other bones also may be involved.

Generally, one type of histological predominant pattern is observed in osteosarcomas. Our case revealed varied histologic patterns like osteoblastic, chondroblastic, telangiectatic, hemangiopericytomatous, small cell, and spindle cell pattern in the same slide of a small biopsy. Despite the varied histopathological patterns observed, the presence of osteoid produced directly by the tumor cells clinched the diagnosis. Furthermore, the tumor was located in a relatively rare site for an osteosarcoma.

The 5-year event-free survival rate of patients with localized high-grade osteosarcoma is up to 70%, but the prognosis for patients who present with metastatic disease is still poor, with a 5-year survival rate of approximately 30%.[9] Most osteosarcomas are resected after preoperative chemotherapy. The incidence of amputation for osteosarcoma has diminished dramatically. Amount of necrosis observed after chemotherapy correlates with prognosis.

Hence, it is important for surgical pathologists to evaluate the extent of necrosis in resected osteosarcoma.[4]

The case has presented due to its unusual location and to highlight its varied histomorphological patterns observed.

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