Case Report

Chondroblastic variant of extraskeletal osteosarcoma of thigh: A rare case report

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ABSTRACT

Extraskeletal osteosarcoma (ESOS) is a rare, malignant neoplasm comprising 1% of mesenchymal tumors. We hereby report a case of ESOS of the thigh which was diagnosed on histopathology. Magnetic resonance imaging showed two lobulated markedly heterogeneous masses in the subcutaneous fat of the lateral aspect of thigh measuring 5 cm × 4.2 cm × 4.2 cm and 7.1 cm × 5.7 cm × 6.0 cm. Femur and major neurovascular bundles were unremarkable. Excision biopsy revealed a circumscribed tumor comprising sheets and fascicles of spindle cells and abundant osteoid produced directly by malignant cells. Malignant cartilage and extensive areas of calcification were also seen. Hence, a diagnosis of ESOS was given. Prompt diagnosis and surgical resection are mandatory, because unlike conventional osteosarcoma, ESOSs are more aggressive and do not respond to chemotherapy.

Key words: Aggressive, extraskeletal osteosarcoma, malignant osteoid

INTRODUCTION

Extraskeletal osteosarcoma (ESOS) is a rare, malignant mesenchymal neoplasm in soft tissues with no attachment to the skeletal system. It accounts for <4% of all osteosarcomas (OSs) and approximately 1.2% of all soft tissue sarcomas. ESOS was first reported by Wilson and <300 cases have been reported.[1] Only a few reports of ESOS have been described in literature. Here, we describe a case of ESOS in the thigh.

CASE REPORT

A 35-year-old male patient presented with gradually progressing swelling over the outer aspect of right thigh for 2 months. The swelling was excised 1 month back but rapidly progressed to the present size. Overlying skin revealed ulcer covered with foul-smelling blood mixed purulent discharge. Magnetic resonance imaging (MRI) showed two lobulated markedly heterogeneous masses in the subcutaneous fat of lateral aspect of thigh measuring 5 cm × 4.2 cm × 4.2 cm and 7.1 cm × 5.7 cm × 6.0 cm. These were lying in opposition with the margins of the underlying muscle with loss of intervening fat plane but no obvious infiltration into it. Femur and major neurovascular bundles were unremarkable. Possibilities of malignant fibrous histiocytoma and fibromatosis were suggested. We received a soft tissue mass measuring 11 cm × 7 cm × 5 cm with a large overlying skin ulcer measuring 6 cm × 4 cm [Figure 1]. Base of ulcer was covered with necrotic slough. Cut section of the tumor was gray-white, glistening, firm to hard to cut. Microscopic examination of sections from growth revealed lining by keratinized stratified squamous epithelium with the area of ulceration covered with exudate. Deep dermis revealed a circumscribed tumor comprised sheets and fascicles of spindle cells and abundant osteoid produced directly by malignant tumor cells [Figure 2]. Tumor cells were highly pleomorphic, lobulated/bizarre/spindle-shaped nuclei, having hyperchromatic/vesicular nuclei, prominent one to

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multiple nucleoli, and moderate to abundant eosinophilic cytoplasm [Figure 3]. Numerous atypical mitotic figures were seen. Extensive areas of cartilaginous differentiation and calcification were also seen [Figure 4]. Circumferential and deep resection margins were free. A diagnosis of chondroblastic variant of ESOS was given.

DISCUSSION

ESOS is defined as malignant mesenchymal neoplasm composed of cells producing osteoid, bone, and/or chondroid material, with no attachment to bone or periosteum.[2] It is extremely rare accounting for 1% of soft tissue sarcomas.

It usually affects people after 50 years of age, unlike conventional OS which affects young individuals. Male/female ratio is 1.9:1. It presents as a progressively enlarging soft tissue mass (as was seen in our case) which is painful in about one-third of the patients. The duration of symptoms varies from weeks to several months.[3] It typically arises in the deep soft tissue of the thigh (47%). Other less frequent sites include the buttock, shoulder, trunk, and retroperitoneum.[2] Unusual locations include larynx, tongue, mediastinum, spermatic cord, penis, pleura, lung, heart, colon, and central nervous system. On computed tomography scan and MRI, ESOS manifests as a soft tissue mass with spotty to massive calcifications and no evidence of bone involvement was seen in our case.

Large and late examples of the tumor may ulcerate through the skin as was seen in our case. It most likely arises from the sarcomatous transformation of multipotent mesenchymal cells contained in soft tissues.[4]

Etiology is unknown. 13% cases are associated with previous trauma, irradiation, areas of myositis ossificans or heterotopic ossification.[5]

Grossly, they can be a well-circumscribed mass with a distinct pseudocapsule to an infiltrating tumor without discernible

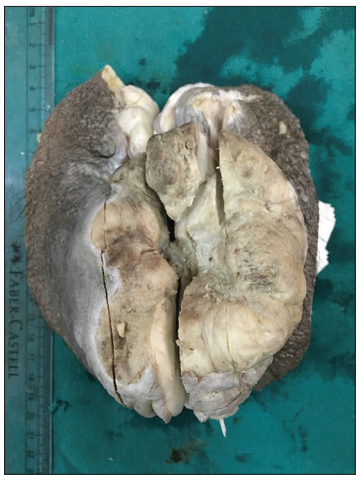

Figure 1: Soft tissue mass with large skin ulcer covered with slough

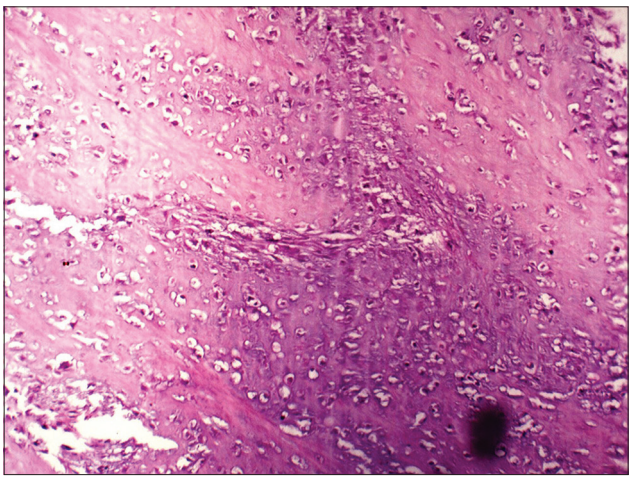

Figure 2: Abundant osteoid and cartilage (H and E, ×10)

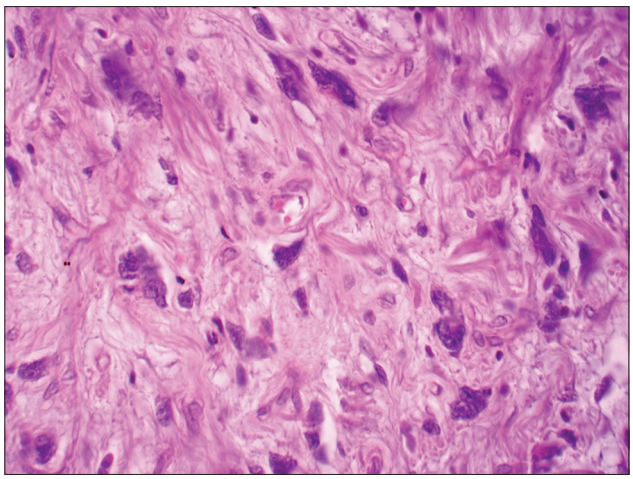

Figure 3: Malignant osteoid being laid directly by neoplastic cells and mineralization of osteoid (H and E, ×40)

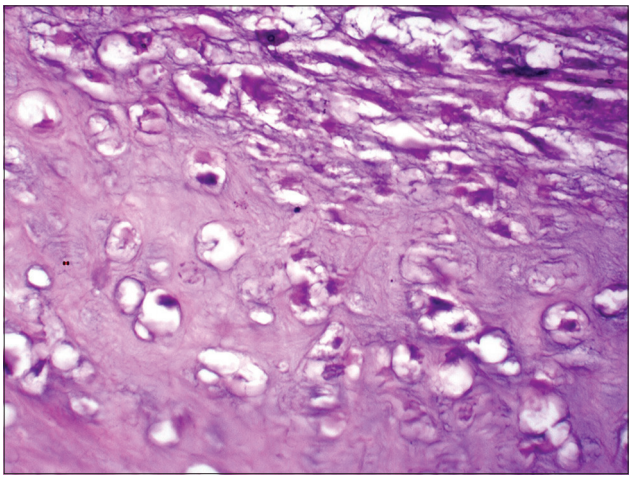

Figure 4: Malignant chondrocytes (H and E, ×40)

borders, firm to stony hard on palpation as was seen in our case. Cut section is granular white surface with yellow flecks and multiple foci of necrosis and hemorrhage. ESOS like OSs of bone can be fibroblastic, osteoblastic, chondroblastic, osteoclastic or giant cell, telangiectatic, and small cell OS.

Differential diagnosis includes benign lesions such as myositis ossificans circumscripta, ossifying lipoma, soft tissue osteoma or chondroma, and ossifying fibromyxoid tumor, and malignant lesions such as parosteal OS, mesenchymal chondrosarcoma, malignant melanoma, and synovial sarcoma.[4] In most of these neoplasms, osteoid or bone is confined to a small portion of the tumor and is relatively well-differentiated without the disorderly pattern and cellular pleomorphism of OS.

Myositis ossificans shows mature bone at periphery (zoning phenomenon) while ESOS shows a "reverse zoning phenomenon," i.e., central deposition of osteoid material and atypical spindle cell proliferation at the periphery. Amplification of genes in 12q13-15 region, such as *SAS*, *CDK4*, and *MDM2*, is relatively frequent in OS, especially low-grade parosteal OS, making them suitable markers for distinguishing them from benign ossifying lesions.[4]

ESOS is a relatively chemoresistant unlike OS of bone which is generally chemosensitive. Both cisplatin-based chemotherapy and doxorubicin-based chemotherapy are not active against ESOS. Therefore, it is asserted that ESOS should be treated as therapeutically distinct from conventional osseous OS.

ESOS carries an exceptionally poor prognosis and is generally involved with invasion and metastasis. The lungs constitute the most common metastasis, followed by the liver, bones, regional lymph nodes, and soft tissue. The recurrence, transfer, and 5-year survival rates are 45%, 65%, and 25%–37%, respectively.[1]

Tumor size, histologic subtype, and proliferation index have been proposed as prognostic variables. Tumor size of 5 cm or more was an unfavorable prognostic indicator. Patients with the fibroblastic and chondroblastic type of ESOS have slightly better prognosis than those with other histologic subtypes. Despite the dismal prognosis, combination therapy with radical surgery (possibly limb-sparing segmental resection as an alternative to amputation), radiotherapy, and sequential preoperative or postoperative chemotherapy should be carried out in the hope of improving survival.

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Conflicts of interest
There are no conflicts of interest.

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