CASE REPORT

Extraskeletal Osteosarcoma of the Gluteal Region

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Abstract:

Extraskeletal osteosarcomas are rare malignant mesenchymal tumors. They mimic many bone and cartilage forming soft tissue tumors and tumor like conditions. Most cases pursue an aggressive clinical course with high rate of local recurrence and metastasis. We report a case of extraskeletal osteosarcoma of the left gluteal region in a 50 year old male with varied histology. It was diagnosed as malignant mesenchymoma but, immuno-histochemistry excluded its possibility.

Keywords: Extraskeletal, Osteosarcoma, Immuno histochemistry

Introduction:

Extraskeletal osteosarcoma (ESOS) is a rare malignant mesenchymal neoplasm that accounts for 1% to 2% of all soft tissue sarcomas [1-4]. It was first described by Wilson [5] in 1941. By definition, ESOS is unattached to the skeleton and composed of malignant cells of osteoblastic phenotype, which produce osseous matrix [2, 4]. The extraskeletal location is determined by the radiographic examination or inspection during the operative procedure [3]. It may pose diagnostic problems in differentiating from other soft tissue sarcomas with bone production [3, 4]. We report a case of ESOS in the gluteal region, which displayed varied histology.

Case History

A 50-year-old man was admitted to the hospital for evaluation of mass in the left buttock of one year duration. He had throbbing pain and purulent discharge from the mass for the last 15 days. There was no history of local trauma or radiation. Physical examination revealed a mass in the left gluteal region measuring 18x15x10cm with an area of ulceration and restricted mobility. There was no regional lymphadenopathy. The systemic examination was within normal limits. The chest X-ray was normal. A clinical diagnosis of lipoma with secondary changes was made and hence, X-ray of the left gluteal region was not taken. At operation, the mass was in the subcutaneous plane infiltrating the skin and free from adjacent bony attachments. Simple excision was done and sent for histopathological examination.

Grossly the mass was measuring 15x12x7cm, partly covered by skin with an area of ulceration. Cut surface showed a well circumscribed, solid, lobulated, pale white tumor with focal bony hard, mucoid and necrotic areas (Fig.1 a &1b).

Microscopy showed an unencapsulated, subcutaneous, moderately, cellular tumor with varied histology. The predominant pattern was

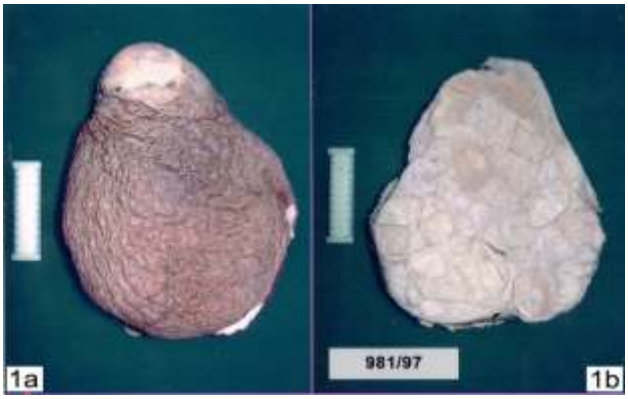

Fig.1. Gross Photograph of the Mass Covered By Ulcerated Skin (1a) with Pale White Lobulated Cut Surface (1b)

osteosarcomatous, composed of pleomorphic polygonal to spindle cells, arranged in sheets and lobules. The cells showed hyperchromatic nuclei, prominent nucleoli and eosinophilic cytoplasm. Mitoses were 1-2/10HPF. Amidst these cells homogeneous, eosinophilic osteoid was seen in lace-like and trabecular pattern (Fig.2a). Moderate areas of necrosis were noted (2b). At places the tumor cells were compressed by osteoid matrix (Fig.3). In other areas islands of cartilage with chondrosarcomatous differentiation were noted (Fig.4).

At places the tumour showed areas resembling synovial sarcoma, neurogenic sarcoma and

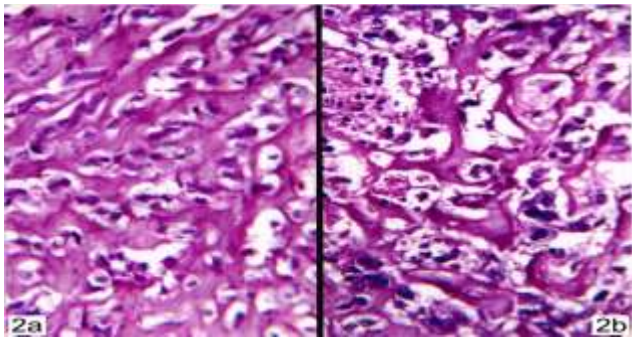

Fig 2: Microphotograph Showing Osteoid in Trabeculae Rimmed by Pleomorphic Tumor Cells (a) with Areas of Necrosis (b) (H&E, x 200).

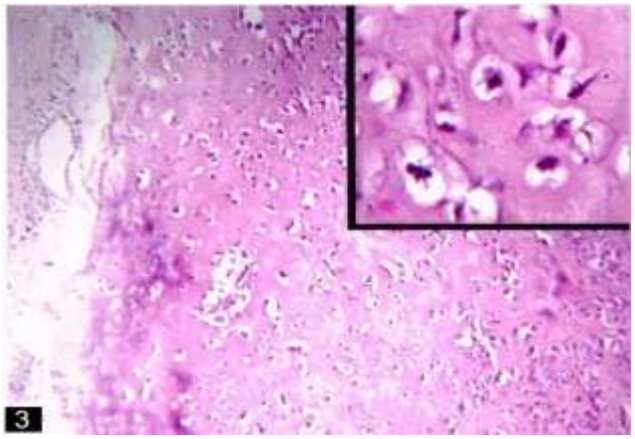

Fig 3: Microphotograph showing Tumor Cells (Inset: H&Ex400) Compressed by Osteoid Matrix (H&E, x 200).

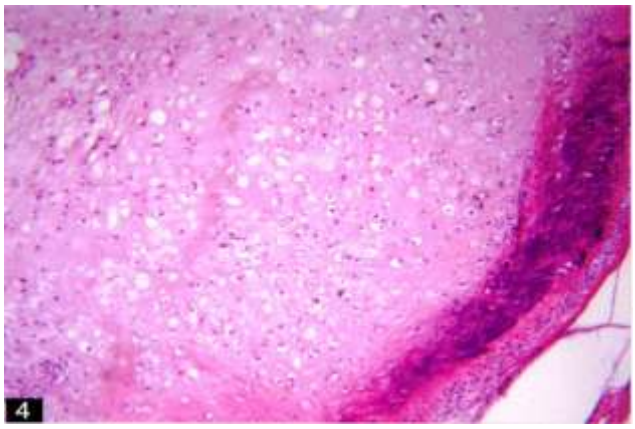

Fig 4: Microphotograph showing Chondrosa -rcomatous Areas with Calcification (H&E x100)

liposarcoma in myxoid background. There were areas of hyalinization merging with osteoid and bone. A diagnosis of malignant mesenchymoma was considered. On immunohistochemistry (IHC) the tumor cells were positive for vimentin and negative for desmin, S-100 protein, CK, EMA, SMA, GFAP, Ulex lectin and Factor VIII related antigen.

Based on these, a diagnosis of extraskeletal osteosarcoma of the left gluteal region was given. Subsequent test for serum alkaline phosphatase

level was found to be normal. The postoperative course was uneventful without local recurrence. The patient died 18 months after the diagnosis with pulmonary metastasis.

Discussion:

ESOS is a high grade malignant neoplasm [2, 5], commonly seen in the sixth decade [4-6]. The most common location is the lower extremity with thigh as the commonest site [2, 3, 6-9]. It may arise in unusual locations like larynx, tongue, mediastinum, spermatic cord, penis and central nervous system [3]. Cases have been reported with previous history of local trauma [5, 8, 9], radiation [1, 5, 6, 9], dermatomyositis [5] and myositis ossificans [5, 9]. ESOS shows histologic patterns comparable to those described in skeletal osteosarcoma [7]. The present case showed predominantly osteoblastic component. The patients with fibroblastic, chondroblastic and well differentiated variants are said to have better prognosis compared to osteoblastic variants, but the findings have not been confirmed by larger studies [6].

ESOS should be differentiated from other malignant bone forming soft tissue tumors like synovial sarcoma, malignant fibrous histiocytoma (MFH), liposarcoma, malignant mesenchymoma and myositis ossificans [6]. In most of the tumors, osteoid or bone is confined to a small portion of the tumor and is relatively well differentiated [3]. Malignant mesenchymoma is a rare, aggressive soft tissue tumor, which shows at least 3 different malignant soft tissue phenotypes other than any undifferentiated components. There are very few cases reported in the literature. According to the recent view, this term may be avoided and the tumor may instead be described as having various components as one would do for germ cell tumours [10]. ESOS is uniformly positive for vimentin and occasionally positive for desmin, actin, S-100 protein, EMA and CK [6]. The present case was diagnosed as malignant mesenchymoma but, IHC excluded its possibility as it was positive only for vimentin. Osteocalcin is the most specific antigen for identifying ESOS which is expressed in the malignant cells and matrix in 82% and 75% of cases respectively [6]. Immunostain for osteocalcin was not done in the present case. The prognosis of ESOS is poor with mortality rate of 70% to 80% [9]. Tumour size greater than 5cm is predictor of worst outcome [6, 9]. They show recurrence and metastasis in more than 50% of the patients [6]. [1]. Most of the patients succumb to metastatic growth within 2 to 3 years after diagnosis [3]. Metastasis to the lungs are the most common [1, 3, 5, 7] followed by lymph nodes, bone, brain, liver and skin [6]. The treatment of choice is surgery with adjuvant radiotherapy and chemotherapy [8].

To conclude ESOS should be considered in the differential diagnosis of any malignant mesenchymal mass in the thigh in view of its varied histology and aggressive behavior.

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