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Primary Extraskeletal Osteosarcomata of Liver, Kidney, Breast and Thigh

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ABSTRACT

Extraskeletal or soft-tissue osteosarcoma is rare, occurring approximately as 1.2% of all soft-tissue sarcomas and 4% of all osteosarcomas. The peak occurs in patients, which are in the 6th decade of life. The males are slightly more frequently affected. Symptoms often include a slowly growing painful mass, with a history of trauma (12% of cases). Common sites of involvement are the deep soft tissues of the thigh (47% of cases), upper extremity (20%), and retroperitoneum (17%). The identification of neoplastic osteoid matrix formation and aggressive characteristics are necessary for histological diagnosis of osteosarcoma. We are going to discuss 06 cases of extraskeletal osteosarcomas including primary renal, primary hepatic osteosarcoma, one arising in a cystosarcoma phylloides and 03 cases involving the thigh regions.

Key words: Osteosarcoma, extraskeletal, renal (kidney), hepatic (liver), breast, thigh.

INTRODUCTION

Osteosarcoma is the most common primary malignant tumour of bone in adolescents and young adults. It accounts for approximately 15% of all primary bone tumors confirmed at biopsy. There are numerous types of primary osteosarcoma, including intramedullary osteosarcoma, juxtacortical (surface) osteosarcoma, and extraskeletal osteosarcoma. Osteosarcoma may also occur as a secondary lesion in association with underlying benign conditions. The identification of neoplastic osteoid matrix formation and aggressive characteristics are necessary for histological diagnosis of osteosarcoma. Extraskeletal or soft-tissue osteosarcoma is rare, occurring approximately as 1.2% of all soft-tissue sarcomas and 4% of all osteosarcomas[1-6]. The peak occurs in patients in the 6th decade of life and males are slightly more frequently affected. Symptoms often include a slowly growing painful mass, with a history of trauma (12% of cases)[6]. Common sites of involvement are the deep soft tissues of the thigh (47% of cases), upper extremity (20%), and retroperitoneum (17%)[4-6].

These are usually large (average, 9 cm in diameter) with variable amounts of neoplastic osteoid and bone[5]. Other components, include cartilage, fibrosarcoma, malignant fibrous histiocytoma, and malignant peripheral nerve sheath tumor, are also frequently seen[4].

Radiologically large soft-tissue masses with focal to massive areas of mineralization and a lack of osseous involvement are seen[7]. Non-mineralized areas have muscle attenuation values on CT scans, nonspecific intermediate signal intensity on T1-weighted images, and high signal intensity on T2-weighted MR images and show contrast enhancement[7]. A pseudo capsule may also be apparent. Scintigraphy often reveals increased radionuclide uptake in both primary and metastatic foci.

Treatment includes amputation or wide surgical resection with neoadjuvant chemotherapy or radiation therapy. Tumor size is the most important prognostic factor, patients with lesions larger than 5 cm have a worse outcome[4-6]. Even with aggressive therapy, overall prognosis is poor;

with death of patient occur in at least 60% of cases[4-6]. Frequent metastases are identified; involving the lung, regional lymph nodes, and bone.

MATERIAL AND METHODS

We had received biopsy specimens in the private clinical laboratory. Representative sections were taken and subjected to routine paraffin processing. Tissue sections cut from these paraffin blocks underwent time tested Hematoxylin and Eosin stain, were examined under the light microscope.

Case No. 1

A 40 years old female patient, presented with gradually increasing lump in her right breast, for the last 06 months. She was operated and later we received a simple mastectomy specimen measuring 13x10x11 cm. The nipple bearing ellipse of skin measures 13x8 cm. On serial sectioning a greyish white tumour is seen measuring 7x6.5x5 cm with central irregular area of necrosis (Fig. 1).

Multiple sections from this greyish white necrotic tumour reveals a malignant spindle cell neoplasm composed of spindly cells containing hyperchromatic vesicular nuclei. Frequent mitoses are seen. Scanty epithelial component is seen at places forming duct like structure. Extensive tumour necrosis is seen Focal areas reveal neoplastic osteoid and multinucleated tumour giant cells. A microscopic diagnosis of malignant phylloides tumour with focal osteosarcomatous change was made (Fig. 1).

Case No. 2

A 30 years old male, resident of rural area of Punjab presented with haematuria and dull pain in the left flank. There was no evidence of any primary tumour or bony lesion at any other site. We received a left nephrectomy specimen measuring 7x5x6cms with attached part of blood vessel measuring 2.5cms in length. The cut surface reveals grayish white tumour with areas of hemorrhage involving most of the renal parenchyma. Ureter was not identified (Fig. 2).

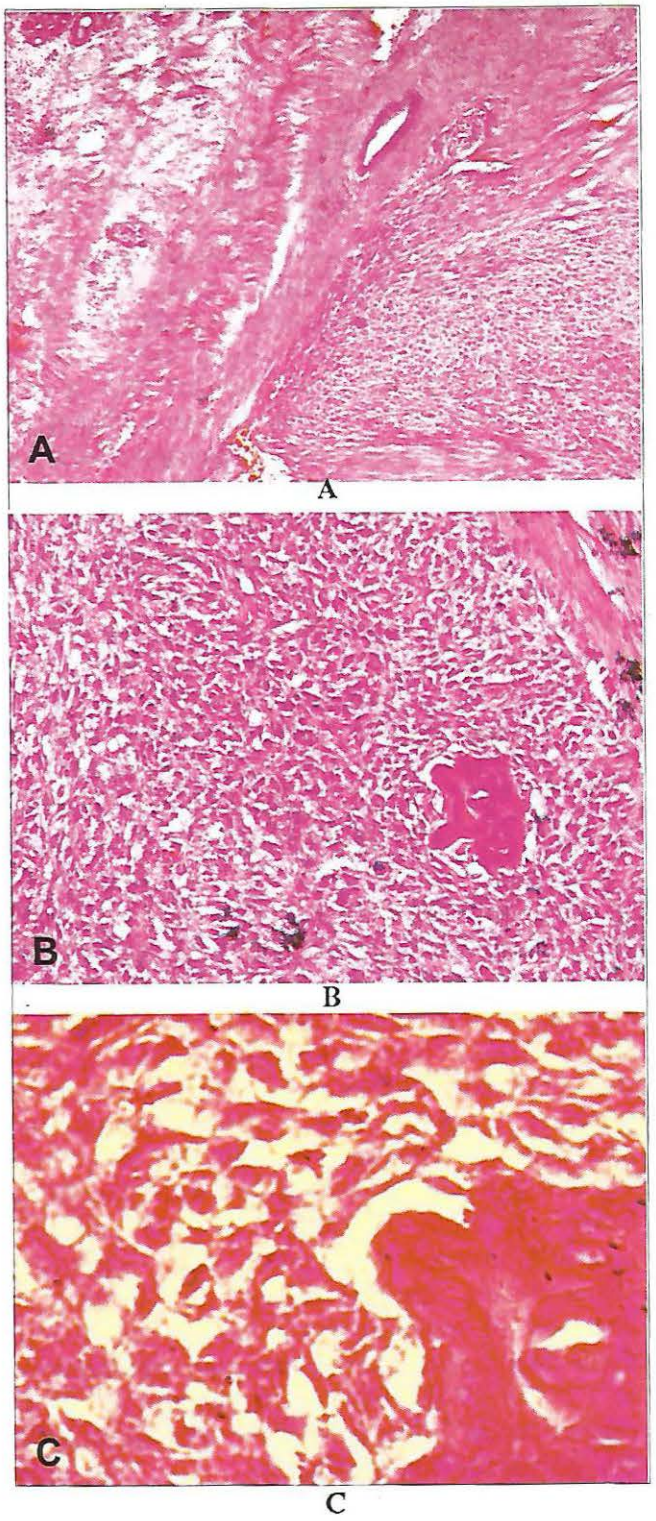

Fig. 1. Osteogenic sarcoma in cystosarcoma phylloides tumour of breast. A) scanner view (2x); B, low power view (10x); C, high power view (40x).

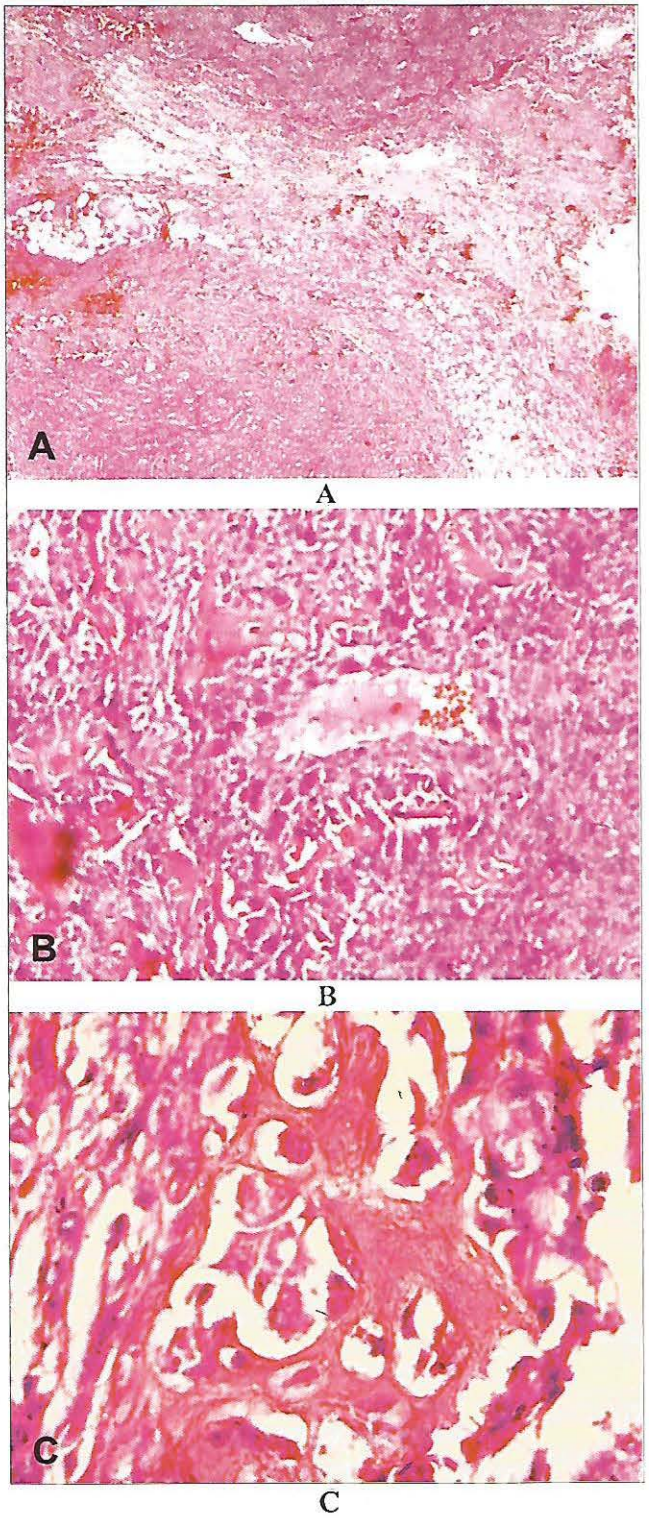

Fig. 2. Primary osteogenic sarcoma of kidney. A) scanner view (2x); B, low power view (10x); C, high power view (40x).

Sections from this nephrectomy specimen reveal renal tissue infiltrated by a malignant mesenchymal neoplasm composed of pleomorphic malignant round to spindly cells containing hyperchromatic nuclei. Mitoses are present. Neoplastic osteoid is seen surrounded by these malignant neoplastic cells. There is no evidence of capsular or vascular invasion by the neoplasm.

Case No. 3

A 70 year old man presented with a painful mass of liver. There was no evidence of any primary bone tumour or bony lesion at any other site. After surgery we received a part of liver measuring 10x6x7 cm mostly replaced by a firm grayish brown haemorrhagic tumour (Fig. 3).

Microscopically, the sections from this liver segment reveal hepatic tissue infiltrated by a malignant mesenchymal neoplasm. This neoplasm is composed of sheets of pleomorphic malignant cells containing hyperchromatic nuclei, abnormal mitoses and multinucleate giant cells. Extensive osteoid formation is seen.

Cases 4 to 6

Two male patients of 67 and 68 years respectively presented with painful masses in the lower part of their thighs and 01 patient of 72 year age complaint of painful lump in the middle part of his thigh. On radiographic examination these growths were not related to or involving the underlying femur and reveled soft tissue masses with focal calcification. There was no evidence of any primary tumour or bony lesion at any other site. Open bone biopsies were taken and diagnoses of extraskeletal osteosarcoma were made (Fig. 4).

DISCUSSION

Extraosseous osteosarcoma can occur anywhere in the body but it is exceedingly rare to find this malignancy in kidney and liver and also in the breast. A review of literature revealed only 20 cases which appeared to be clearly identifiable as malignant extra-osseous bone forming sarcoma of the kidney since 1936 and 26 cases of primary osteosarcoma of liver The study of these cases shows that these tumours can occur in both males

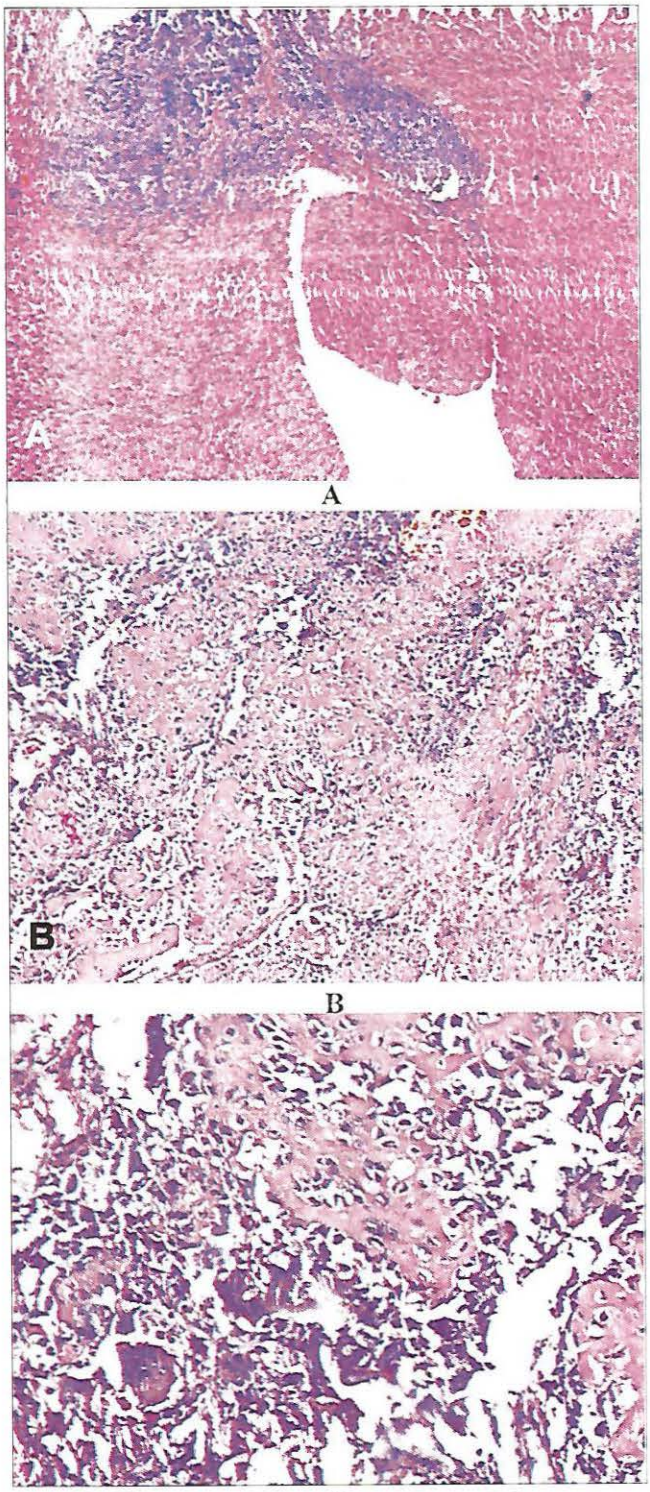

Fig. 3. Primary osteogenic sarcoma of liver. A) scanner view (2x); B, low power view (10x); C, high power view (40x).

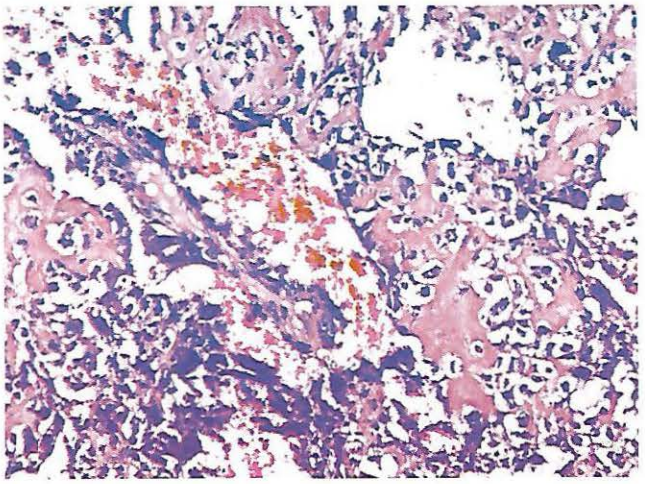

Fig. 4. Extra skeletal primary osteogenic sarcoma of thigh (40x).

and females. Sixty cases of primary osteosarcoma of breast were reported. Ages range between 30 – 72 years. The usual presenting complaints are abdominal mass and or gross haematuria. Roentgenographic evidence of calcification was seen within these tumours. The patients usually develop metastasis and die of their tumours. The origin of renal/hepatic or mammary osteo-sarcoma is not known. An origin from indifferent mesenchymal, as we postulate for renal liposarcoma and rhabdomyosarcoma, seems likely in view of the well documented potential of osteosarcoma to develop in extra-osseous tissues. Rarely, both nephroblastomas and sarcomatoid renal adenocarcinomas are accompanied by malignant bone formation; in nephroblastoma, as one line of malignant differentiation and in renal adenocarcinoma, most likely as a form of malignant metaplasia. There is no evidence to suggest that any of these osteogenic sarcomas reported in kidney represent either nephroblastoma or renal adenocarcinoma[8-13].

CONCLUSIONS

The radiographic imaging modalities, including sonography, bone scintigraphy, CT, and MR imaging, provide vital information for preoperative staging and in planning surgical management. The diagnosis is always based on

histopathological findings, for which adequate sampling of lesions is necessary, failing which one can miss the neoplasm. Radiological examination also allows evaluation of tumor response to chemotherapy, identification of metastatic disease, and postoperative evaluation of recurrent neoplasm, all of which have important prognostic implications. Recognition of these imaging features is an important guide to our clinical colleagues, throughout the often difficult and complex treatment of patients with osteosarcoma and results in improved clinical outcome.

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