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A huge abdominal lump with multiple bony bumps

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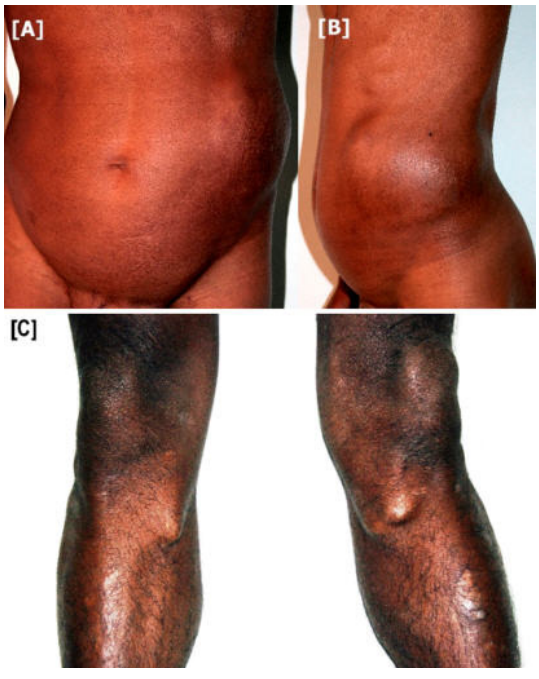
Hereditary multiple exostoses (HME) or osteochondromatosis is a familial autosomal dominant disorder associated with multiple bony protrusions, with cartilage caps usually arising from the lower femur, upper tibia, upper humerus, or the pelvis. Osteochondromas are usually asymptomatic, but may be associated with cosmetic deformities, impairment of neurovascular or musculo-tendinous functions, and malignant transformation into chondrosarcoma.

We report on a patient with multiple exostoses: well-differentiated secondary chondrosarcoma arising from an iliac osteochondroma.

Case report:

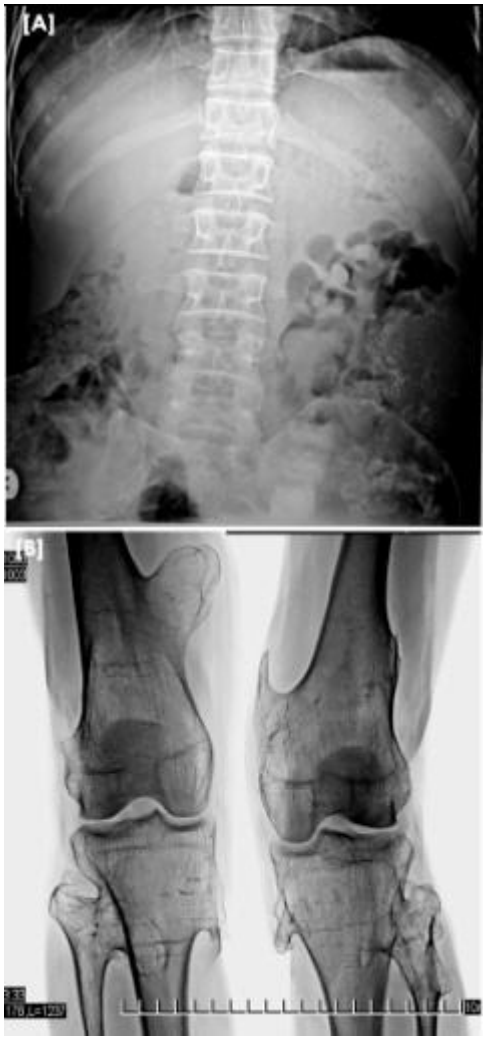
A 45-year Asian Indian male presented with a huge lump on the left side of abdomen of 5 months duration with dull aching continuous pain over it for 1 month. Examination revealed bony hard projections on the medial aspect of both the lower end of femur and upper end of tibia (Figure 1C). There was a lobulated, hard, immobile retroperitoneal lump (25×20 cm) occupying the left hypochondrium, lumbar, iliac, hypogastric, and umbilical regions (Figure 1A,B) whose lower border merged with the ilium.

Figures 1A: Anterior abdomen showing the lump; 1B: Left lateral abdomen showing the lump; 1C: Anterior aspect of both knee joints showing exostosis arising from the tibia



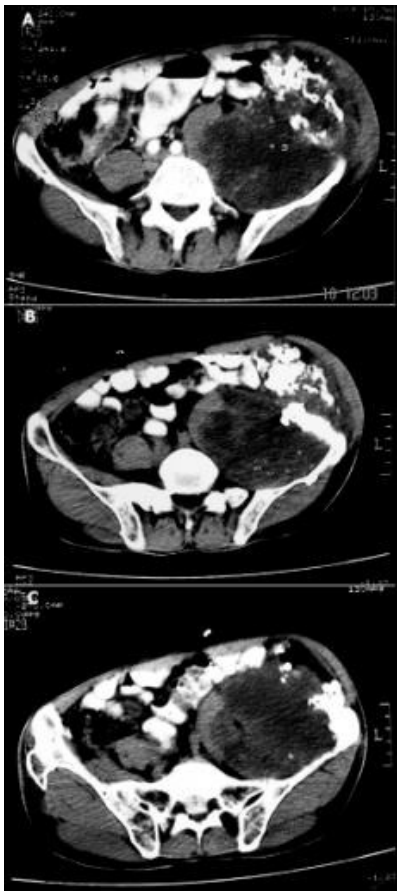
Ultrasonography demonstrated a retroperitoneal mass with mixed echogenicity extending into the pelvis. Radiography revealed a large soft tissue shadow with multiple calcific foci in left abdomen (Figure 2A) as well as pedunculated exostoses of tibia and left femur, and sessile exostoses of the right femur and the left fibula (Figure 2B).

Figures 2A: Radiograph of abdomen showing calcification with soft tissue shadow in the left lower abdomen; 2B: Radiograph both knees showing multiple exostoses

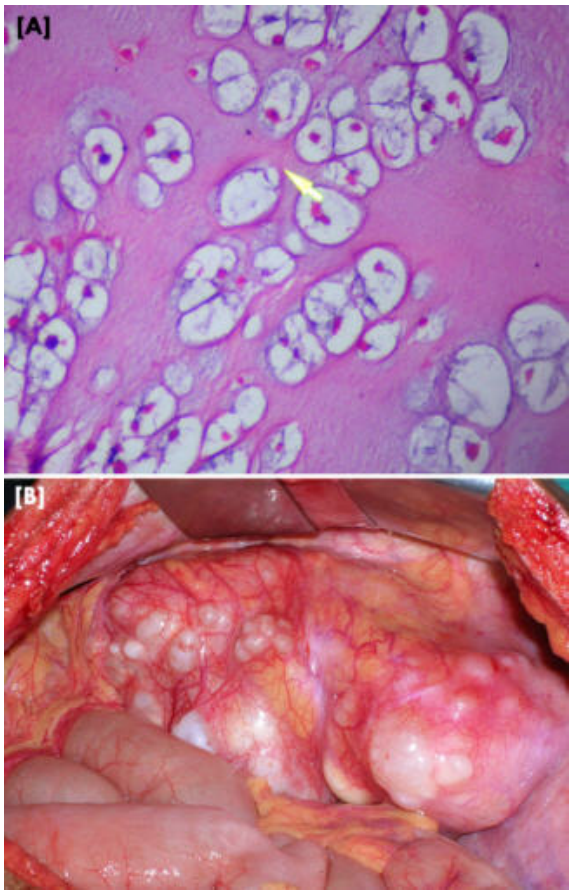


Abdominal CT (Figure3 A,B,C) showed a large heterogeneously enhancing soft tissue mass lesion in the left flank. There was evidence of splotchy calcification and central non-enhancing areas in the lesion. It was displacing the ilio-psoas muscle anteriorly, bowel medially, and left external iliac artery to right. There was pressure erosion of the left iliac wing with exostoses involving both the iliac blades.

Figures 3A,B,C: Axial CT scan abdomen through different levels showing heterogeneously enhancing soft tissue mass with calcification, pressure erosion, and bony exostoses



Figures 4A: Microphotograph 45 × (haematoxylin-eosin) showing nuclear atypia consistent with well differentiated chondrosarcoma. 3B: Intraoperative view showing a nodular white neoplastic mass



Histopathology after a trucut needle biopsy revealed multiple clustered chondrocytes with vacuolated cytoplasm, a few multinucleated, in lacunae within a chondroid matrix, consistent with a well-differentiated chondrosarcoma (Figure 4A). The patient underwent an exploratory laparotomy with wide excision of the tumour (Figure 4B).

Discussion

Osteochondroma is the most frequent benign bone tumour.[1] Multiple such tumours occur in an autosomal dominant disorder known as osteochondromatosis, hereditary multiple exostoses (HME) or diaphyseal aclasis. They are commonly seen at the metaphyses of the lower femur, upper tibia, upper humerus, scapula, and the pelvis; 1% to 5% of multiple osteochondromas may undergo malignant transformation into chondrosarcomas.[1–3]

Chondrosarcoma is the second most common primary malignant tumor of bone of cartilaginous origin, representing approximately 25% of all primary osseous

neoplasms.[4–7] They usually involve the pelvic bones, femur, humerus, ribs, scapula, sternum, or spine and are classified according their location into central, peripheral and juxtacortical.[1,4,8]

Primary chondrosarcomas arise *de novo*, whereas secondary chondrosarcoma originate in a preexisting lesion such as an enchondroma or osteochondroma.[4,5,8,9] The risk of chondrosarcoma is greater in people with enchondromatosis syndromes (Ollier disease, Maffucci syndrome, metachondromatosis) and in those with HME.[1,4] They are graded into well, moderately and poorly differentiated types based on their histological features such as cellularity, matrix content, character of cells, and replicative activity.[1,3–5,8,10]

Transformation of osteochondromas into chondrosarcomas occurs between the ages of 20 to 40 years.[1,4] Radiographs classically show lucent lesions due to cortical destruction, soft tissue mass, periosteal reaction, destruction or pressure erosion of adjacent bone, and endosteal scalloping with matrix calcification and well-organized calcific rings in well-differentiated tumors.

Poorly differentiated tumors contain scattered, irregular, punctate calcifications and sometimes tumour matrix with no calcification at all.[4,8,9] CT scan provides important knowledge regarding the intraosseous and soft tissue extent of the tumour. Tumours appear as lucent areas containing chondroid matrix calcification.

Differential diagnosis of retroperitoneal tumours with calcification include ganglioneuroma, schwannoma, paraganglioma, hemangioma, mature and malignant teratoma, undifferentiated pleomorphic sarcoma, dedifferentiated liposarcoma, malignant mesenchymoma, and extraskeletal osteosarcoma.[4,11]

MRI is useful in defining the full extent of the tumour in anatomically complex regions and shows lobulated homogeneous or inhomogeneous lesions of high signal intensity in T2-weighted spin echo images.[2,4,8] There is greater focal or diffuse enhancement following intravenous contrast administration in high grade chondrosarcomas.[8]

Surgery is the main treatment modality. Surgical options—which include radical excision; wide local excision; and even marginal, partial, or intralesional excision—depends on the tumour size, tumour grade, and the local infiltration. High-grade tumours require complete surgical excision while low grade chondrosarcomas may be treated with contaminated margins to reduce operative morbidity, without compromising survival rates.[5,6]

Chondrosarcomas respond poorly to radiotherapy and chemotherapy.[4–7,9,10] The prognosis with respect to local control and/or survival depends on the histological grade, the surgical stage, the subtype of chondrosarcoma, adequate margins of resection and, the location of tumour.[7,9,10]

Due to a high incidence of late local recurrence, chondrosarcomas should be histologically graded accurately, treated effectively via surgery, and carefully followed up for at least 5 years.

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