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MANAGEMENT OF DISTAL FEMUR OSTEOCHONDROMA WITH LARGE CARTILAGINOUS CAP AND LEFT UPPER LIMB HEMIMELIA AND ECTRODACTYLY – A RARE CASE REPORT

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ABSTRACT

Introduction: Osteochondroma are the most common benign primary bone tumour. They are bony growth away from joint line surrounded by a distinguishing cartilaginous cap, most commonly arising from the long bones. They are most often symptomless, usually revealed as incidental findings
before the third or fourth decade of life.

Case Report: A 36 male patient with left upper limb hemimelia and ectrodactyly presented to our outpatient department at our tertiary care hospital with swelling over right thigh since few years which aggravated over last 6 months and turned painful. He underwent extensive investigations and was posted for resection surgery. Diagnosis was confirmed by histopathological study and patient has been pain free ever since.

INTRODUCTION:

Osteochondromas around the knees are common developmental bony lesions. The majority are solitary and occur due to sporadic mutations. [1] They are developmental deformities rather than true tumours, originating in the outer layer of the bone [2]. Most of these are asymptomatic, are usually growing away from the joint. A significant number of them can be on the posterior aspect of the knee where occasional cases could either cause pressure effects on popliteal artery [3] or Deep vein thrombosis (DVT) [1]. Some have been linked with limb deformities [4], fractures, bursitis [5], neurologic symptoms and rarely there maybe malignant transformation [6].

Case report:

Our patient is a 36-year-old male who consulted us for bone swelling above the right thigh, which had persisted for few years. Initially, the swelling was hemispherical and painless. He was symptomless. However, over the past 6 months he developed pain accompanied by a gradual increase in the swelling to ovoid features. Patient experienced pain on knee flexion terminally. Patient also has hemimelia of left upper limb.

Fig-1

Fig-2

Clinical examination (Fig -1 and Fig-2) demonstrated an oval-shaped osseous mass from the anteromedial aspect right distal femur. The skin over the mass was close-fitting but intact. Upon palpation, the swelling was tender and had an irregular surface, hard consistency, and was immobile. Palpation along the medial line of the knee joint suggested a bony

swelling 15 cm by 10 cms originating from the metaphyseal-diaphyseal zone of the femur. There were no signs of neurovascular compression. Knee rom was 0-110 degrees and was terminally painful.

Fig-3 **Fig-4**

Pre operative anteroposterior and lateral radiographs (Fig-3 and Fig-4) demonstrated osseous growth on medial aspect of distal femur growing away from knee joint with stippled calcification of the apex.

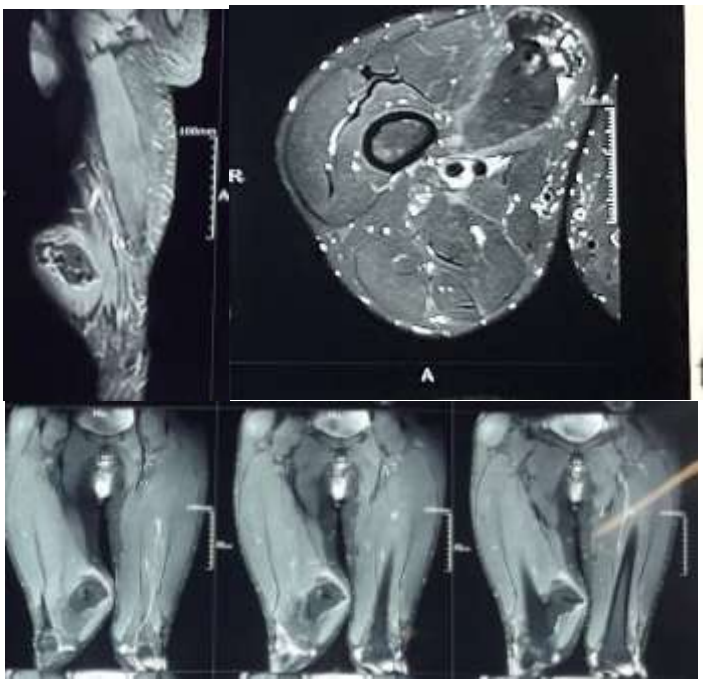

Fig-5

Magnetic resonance imaging demonstrated hypointense lesion in T-1 sequence and hyperintense in T-2 sequence with enhancement. The cartilage cap measured 5.4 mm on ultra sound scanning and cms. Stippled calcification of cartilaginous cap was also seen.

Procedure:

Under regional anaesthesia, we carried out a linear anteromedial incision and approached the mass with subvastus approach. Fig-6 shows capsule held by forceps and osteochondroma with cartilage cap.

Fig-6

Fig-7

En block resection with excision of capsule was done with burring of the base [Fig-7]. Vastus medialis muscle was otherwise normal.

Post operative period was uneventful, patient being able to carry out knee range of motion from 0-130 degrees without pain. He was advised 6 weeks of partial weight bearing.

Fig-8

FIG- 9

FIG-10

FIG-11

Patient also has hemimelia and Ectrodactyly in the left upper limb(FIG- 9, FIG-10 & FIG-11).

DISCUSSION

Osteochondromas are the most common benign bone tumour. They are bony outgrowths surrounded by a cartilaginous cap and arise on the external surface of bones [7]. Solitary osteochondromas most commonly affect the long bones such as the femur and tibia [7,8].

There have also been studies showing the possible role of genetic mutations such as those in the exostosin gene, causing accumulation of heparan sulfate proteoglycans within the cytoplasm, which prevent them from participating in the normal diffusion of Indian hedgehog ligands in the extracellular space. This results in loss of normal polarity, causing chondrocytes in the growth plate to grow in the wrong direction [7,9].

Quite often X-rays, CT scans and magnetic resonance imaging do provide significant information, thus enabling the anatomical description of the lesion [10]. Osteochondromas typically present as pedicles or projections similar to bone. Cancellous and cortical structures exhibit a close association with normal bone. The cartilage shadow is distinctly recognizable at the apex of the tumour, featuring irregular calcification and/or ossification at the centre

Malignant transformation occurs in 1% of solitary lesions and 3–5% in Hereditary Multiple Exostosis,

with the commonest malignant transformation to chondrosarcoma [11, 12]. However large osteochondromas causing substantial compression and symptoms need to

be excised to provide relief. The standard En block excision protocol involves excision of a large base of sessile osteochondromas, with good long-term outcomes [13].

Our presented case did not have compressive symptoms. He does have left upper limb hemimelia and Ectrodactyly. We searched through literature for any association of hemimelia, ectrodactyly and osteochondroma occur together, but did not find any association or co-occurrence of the same.

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