Case Report

Primary malignant Giant cell tumour of femur

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Abstract:
Primary malignant giant cell tumors are distinctively separate from benign giant cell tumors which metastasize. It has a high mortality rate. Radiologically these tumors show total destruction of bone with soft tissue infiltration. With the breakdown of cortex, articular cartilage with infiltration to surrounding tissue or joint space indicates giant cell tumor is aggressive or malignant. The incidence is difficult to access as there are scattered reports without a large series published.

Key words : Primary Malignant Giant Cell Tumour, Femur

Introduction:
Primary malignant giant cell tumors are distinctively separate from benign giant cell tumours which metastasize (1) and secondary malignant giant cell tumors occurring in response to radiotherapy and repeated curettage. (2) It has a high mortality rate. Usually it affects lower end of femur and upper end of tibia. (3) Primary malignant giant cell tumors show histological evidence of conventional giant cell tumor along with areas of sarcomatous stroma. This should be correlated with radiological appearance. The incidence is difficult to access as there are scattered reports without a large series published. (4)

Case History:
A 21 year male presented with progressive swelling of at the lower end of left thigh. There was a history of trauma 8 weeks back, which appears to be coincidental. He narrated the history of manipulation by a local quack, after which the swelling started increasing at a faster pace. At the time of presentation, there was antalgic limp with rise in local temperature, skin was otherwise normal. Total duration of illness was eight weeks.

Conventional x-ray of left knee joint revealed an eccentric osteolytic lesion involving lower end of femur with predominance in lateral condyle with pathological fracture. Marrow involvement extending upto 6 cm from the condyles and soft tissue involvement was also seen (Fig. 1). Computerized Tomography (CT) Scan revealed osteolysis with bone destruction upto 12 cm and soft tissue involvement on the posterolateral aspect of lateral condyle (Fig. 2). With this presentation and radiological features the provisional clinical diagnosis was? Parosteal osteosarcoma ?? Giant cell tumor.

Fine needle aspiration cytology (FNAC) from tumor mass yielded cellular material consisting of plenty of
spindle shaped cells containing pleomorphic hyperchromatic nuclei. Plenty of mitotic figures and tumor giant cells were seen in hemorrhagic background. It was reported as pleomorphic sarcoma (Fig. 3). With preoperative investigations being within normal limits, above knee amputation was done and the specimen was sent for histopathological examination.

**Morphology:**
On gross received a specimen of above knee amputation of left leg. Portion below the knee appearance normal. The area above knee joint around it showed a swelling of 12x12 cm with normal skin above it. Cut surface had a variegated appearance with infiltration into muscles. Thorough dissection upto bone showed total destruction of lower end of femur along with fleshy grayish soft mass arising from lower end of femur, destroying periosteum and infiltrating into surrounding skeletal muscles and subcutaneous plane. Bone at the site of mass was soft and friable, could be cut with ease. Proximal end of amputed femur was unremarkable (Fig. 4).

**Microscopic Features**
Multiple serial sections taken after extensive sampling from various areas showed tumor mass consisting of highly cellular areas consisting of sheets and storiform arrangement of spindle shaped cells containing bizarre hyperchromatic nuclei with prominent nucleoli (Fig. 5). Plenty of mitotic figures were seen (more than 10 per ten hpf) with tumor giant cells. There was abundance of osteoclastic giant cells (Fig. 6). Large areas of haemorrhages and necrosis were seen. No osteoid could be detected. As there was no history of previous biopsy, surgery or radiation and absence of osteoid, the diagnosis of primary malignant giant cell tumor was conveyed.

**Discussion:**
Radiologically these tumors show total destruction of bone with soft tissue infiltration.\(^5\) A differential diagnosis of malignant giant cell tumor should include radiologically lytic sarcomas like osteogenic sarcoma, fibrosarcoma and malignant fibrous histiocytoma. However, expansile eccentric osteolytic lesions involving epiphysial region should point towards giant cell tumor. \(^6\) With the breakdown of cortex, articular cartilage with infiltration to surrounding tissue or joint space indicates giant cell tumor is aggressive or malignant. C.T. and M.R.I. scans are helpful to study nature, vascularity, necrosis, extent and infiltration.\(^7\) More cases of primary malignant giant cell tumor with proper treatment and follow up are essential to project definite trends on early diagnosis, treatment and to improve prognosis which is gloomy at present. \(^8\)

**Conclusion:**
Usually the Giant Cell tumor has been reported to be the benign or the locally malignant condition. Secondary malignancy is reported after inadequate curettage. But this lesion was very aggressive and the clinical & the radiological features were suggestive of osteogenic sarcoma. It was only on histopathology that revealed that lesion was Primary malignant giant cell tumor of femur.
Fig. 2 – C.T. scan showing destroyed lateral condyle of femur.

Fig. 3 – FNAC from tumor mass showing pleomorphic spindle cells with hyperchromatic nuclei (PAP 10x x 10x)

Fig. 4 – Above knee amputed left leg with variegated appearance on cut surface.

Fig. 5 – Shows fibro-sarcomatous pattern of tumor cells (H & E 10x X 10x)

Fig. 6 – Shows sarcomatous pattern with osteoclastic giant cells (H & E 10x X 10x)

Fig 4. Knee amputed leg
References:


