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Dedifferentiated Chondrosarcoma of Femoral Shaft: A Case Report

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Abstract: Dedifferentiated chondrosarcoma is a highly malignant variant of chondrosarcoma. It makes upto 10-15% of all reported chondrosarcomas. Prognosis of dedifferentiated chondrosarcoma is poor and metastasis is common at the time of initial presentation. A 52-year- old man presented with numbness and dragging sensation of right lower limb and pain and wasting of right thigh of 4 month duration. Excision biopsy revealed dedifferentiated chondrosarcoma of femoral shaft. We report this case because of its rare nature and due to absence of metastasis at the time of presentation. **Keywords:** Dedifferentiated chondrosarcoma, High grade, Shaft of femur

INTRODUCTION

Dedifferentiated chondrosarcoma is a rare high grade sarcoma consisting of a nonchondroid spindle cell sarcoma component associated with a low grade cartilaginous component with an abrupt transition between the two components [1]. It makes upto 10-15% of all reported chondrosarcomas. Generally, it occurs in elderly patients and is more common in males. Prognosis is poor. We report a case of dedifferentiated chondrosarcoma of shaft of right femur.

CASE REPORT

52- year- old male presented with numbness, pain and dragging sensation of right lower limb of 4 months duration. History revealed that he is diabetic, hypertensive and dyslipidemic and is on regular treatment. Physical examination showed wasting of right thigh muscle. X-ray showed lytic lesion in the shaft of femur with cortical erosion (Fig. 1). MRI showed lytic lesion with cortical destruction and associated soft tissue extension in the proximal part of right femur. No major vessel or nerve encasement noted. In PET scan, intense metabolic activity in the lytic lesion with soft tissue component involving proximal shaft of femur is found. No other foci of elevated metabolic activity else where in the body was noted. A trucut biopsy from the lesion showed malignant spindle cell neoplasm. Following this report, wide tumour resection and reconstruction with mega module prosthesis was done. The ressected specimen of proximal femur with attached soft tissue whole measuring 19.5x7.5 cm. Attached soft tissue was dark brown in colour. Coronal section showed an ill defined

grey white glistening lesion measuring 8.5x3.5cm in the medulla of shaft of femur with focal cortical erosion (Fig. 2, 3). Microscopic examination of the sections revealed a malignant spindle cell neoplasm composed of spindle shaped cells arranged in bundles and fascicles. Large bizarre cells and multinucleated cells were seen. Medullary portion of neoplasm showed lobules of cartilage with chondrocytes in lacunae showing mild nuclear enlargement. Areas with destruction of bone and extension to soft tissue were also noted (Fig. 4, 5). Histological features suggest the diagnosis of dedifferentiated chondrosarcoma. Post operatively vitals of the patient were stable .He was taken up for chemotherapy. He has completed two cycles of chemotherapy and is on regular follow-up.

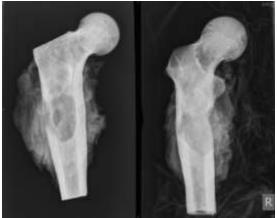


Fig. 1: X-ray showed lytic lesion in the shaft of femur with cortical erosion

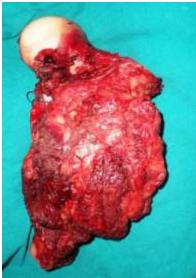


Fig. 2: The ressected specimen



Fig. 3: Cut section showing tumor

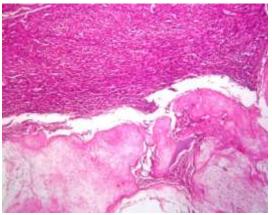


Fig. 4: Low power view showing both low and high grade areas

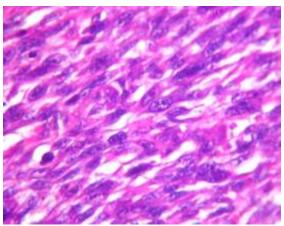


Fig. 5: High power view showing malignant spindle cells

DISCUSSION

Dedifferentiated chondrosarcoma is affecting the age group ranges from 15-69 years with a slight male predominance [2]. Most common sites of involvement are femur, pelvis and humerus. Peripheral dedifferentiated chondrosarcoma involves pelvis, scapula and ribs [3, 4]. Patients usually present with pain, numbness and mass.

dedifferentiated In about 50% of chondrosarcomas, heterozygous mutations of isocitrate dehydrogenese 1 &2 genes are found [5]. Usually the two components of the tumour can be separately identified grossly. Under microscopy the cartilaginous component ranges from enchondroma like appearance to grade I/II chondrosarcoma. The high grade component can show features of osteosarcoma, fibrosarcoma or pleomorphic sarcoma with malignat fibrous histiocytoma like features [6,7]. Rarely, it can show features of angiosarcoma, leimyosarcoma and rhabdomyo sarcoma. In our case, the malignant spindle cell area showed features of fibrosarcoma. Response to post-operative chemotherapy is poor in dedifferentiated chondrosarcoma and it has a poor prognosis [8].

Poor prognostic factors which are very commonly seen in this variant of chondrosarcoma are presence of pathological fracture, metastasis at time of diagnosis, pelvic location and increased age [2]. Most common site of distal metastasis is lung. In the present case, there was no metastasis at the time of diagnosis.

CONCLUSION

We present this case of dedifferentiated chondrosarcoma because of its rarity and absence of metastasis at the time of diagnosis.

REFERENCES

 Dahlin DC, Beabout JW; Dedifferentiation of lowgrade chondrosarcoma. Cancer, 1971; 28(2): 461-466.

- Grimer RJ, Gosheger G, Taminiau A, Biau D, Matejovsky Z, Kollender Y *et al.*; Dedifferentiated chondrosarcoma: prognostic factors and outcome from a European group. Eur J Cancer, 2007; 43(14): 2060-2065.
- 3. Rozeman LB, de Bruijn IH, Bacchini P, Staals EL, Bertoni F, Bovée JV *et al.*; Dedifferentiated peripheral chondrosarcomas: regulation of EXTdownstream molecules and differentiation-related genes. Mod Pathol., 2009; 22(11): 1489-1498.
- Staals EL, Bacchini P, Mercuri M, Bertoni F; Dedifferentiated chondrosarcomas arising in preexisting osteochondromas. J Bone Joint Surg Am., 2007; 89(5): 987-993.
- Amary MF, Bacsi K, Maggiani F, Damato S, Halai D, Berisha F *et al.*; IDH1 and IDH2 mutations are frequent events in central chondrosarcoma and central and periosteal chondromas but not in other mesenchymal tumours. J Pathol., 2011; 224(3): 334-343.
- Johnson S, Têtu B, Ayala AG, Chawla SP; Chondrosarcoma with additional mesenchymal component (dedifferentiated chondrosarcoma). I. A clinicopathologic study of 26 cases. Cancer, 1986; 58(2): 278-286.
- Reith JD, Bauer TW, Fischler DF, Joyce MJ, Marks KE; Dedifferentiated chondrosarcoma with rhabdomyosarcomatous differentiation. Am J SurgPathol., 1996; 20(3): 293-298.
- Meis JM; Dedifferentiation in bone and soft tissue tumours. A histological indicator of tumor progression. Pathology Annual, 1991; 26(Pt. 1): 37-62.