Dedifferentiated Chondrosarcoma of Humerus- A case report

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Abstract:

Dedifferentiated chondrosarcoma is a rare and most malignant form of chondrosarcoma, accounting for approximately 11% of all chondrosarcomas. It generally occurs in older patients having slight male predilection. However, patient in the present case was comparatively younger female with tumour mass in Humerus. The prognosis of dedifferentiated chondrosarcoma is poor. Distant metastasis especially in lungs is often seen. This case is reported here because of its rarity, its association with worse prognosis and absence of pulmonary metastasis, however, subcutaneous nodules were seen at local site.

Key Words: Dedifferentiated chondrosarcoma, Humerus.

Introduction:

Dedifferentiated chondrosarcoma is defined as a high grade, nonchondroid spindle cell sarcoma associated with a low grade cartilaginous tumour (Dahlin & Beabout, 1971). It is approximately 11% of all chondrosarcomas & is among the most aggressive cancers described (Steven & Martha, 1996). These tumours generally occur in old patients and presenting with localized pain, swelling & parasthesia. Most common site is pelvis followed by proximal part of femur. Although there is no common cytogenetic abnormality, many cases show loss of a portion of 1p, trisomy 7 and breaks in q11 region of chromosome 22 (Steven & Martha, 1996). Wide spread metastasis is common with most patients dying within two years regardless of treatment modality. We report a case of dedifferentiated chondrosarcoma of upper end of right humerus in a 45 year old female. Absence of pulmonary metastasis at the time of diagnosis as well as during follow up, has prompted us to report this case.

Case Report:

A 45 year old female presented with gradually increasing swelling of right upper arm. She also complained of pain and restriction of movements at shoulder. Hematological and biochemical investigations were within normal limits. Radiological diagnosis of chondrosarcoma was made. A small biopsy tissue was taken from deeper areas, which revealed well differentiated chondrosarcoma on histopathological examination. Patient was operated upon and upper 1/3 of humerus along with attached tumour tissue was sent for histopathological examination. It revealed well differentiated chondrosarcoma in central portion of tumour while peripheral portion had become dedifferentiated. Patient developed nodules on right forearm one month after surgery for which she refused any surgery or further investigations. Patient also received a shot of chemotherapy. There was no radiological evidence of pulmonary metastasis.

Pathological findings:

Gross examination: Upper 1/3 of humerus (12cm long) with large bosselated mass measuring approximately 12x8x7cms was received for histopathological examination. Tumour mass involved metaphysis & part of diaphysis.

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Epiphyseal end of bone appeared intact (Fig. I). Cut surface showed grey white translucent lobulated tumour tissue towards madullary cavity and soft grey brown necrotic areas at the periphery.

**Microscopic findings:** Sections from tumour mass showed well differentiated chondrosarcoma in central portion of tumour while periphery of tumour had become dedifferentiated and showed areas resembling malignant fibrous histiocytoma (Fig. II), osteosarcoma & fibrosarcoma (Fig. III). Tumour had destroyed the medullary and cortical portions of bone reaching into adjacent soft tissue. Distal bony resection margin was free from tumour infiltration.

**Discussion:**

Dedifferentiated chondrosarcoma is a rare and most malignant form of chondrosarcoma. Dedifferentiated chondrosarcoma generally occurs in older patients typically over the age of 50 years having slight male predilection (Steven & Martha, 1996), however, patient in present case was comparatively younger female. Bones commonly involved are pelvis, ribs and long bones and metaphysis being the most common site of origin (Steven & Martha, 1996). Metaphysis as well as part of diaphysis of humerus was involved in the present case. Radiologically, there was lytic lesion with presence of heavily calcified central area surrounded by less denser periphery with splotchy calcification. There was extension of lesion to the soft tissue as seen in other studies (Rosai, 2004; Bansal et al, 1996). The abrupt transition betweenlytic lesion and cartilaginous tumour that usually is evident on X-ray in classical dedifferentiated chondrosarcoma (Steven & Martha, 1996) was not seen in the present case. However, microscopically, the junction between well differentiated areas and areas of dedifferentiated tumour was abrupt and well recognized.

Dedifferentiated areas may resemble malignant fibrous histiocytoma (MFH), osteosarcoma & fibrosarcoma. In this case, areas of osteosarcoma, fibrosarcoma and MFH were present with predominance of MFH like areas as reported in earlier studies (Johnson et al, 1986; Reith et al, 1996; Rosai, 2004). The prognosis of dedifferentiated chondrosarcoma is poor (Meis, 1991). Distant metastasis especially in lung is often seen (Steven & Martha, 1996; Bansal et al, 1996), contrary to this, in the present case no pulmonary metastasis was seen neither at the time of diagnosis nor during the period of one year follow up. However subcutaneous nodules appeared at local site for which patient refused to undergo further treatment, investigation and surgery.

**Conclusion:**

This case is reported here because of its rarity, absence of pulmonary metastasis and its association with worse prognosis.

**Bibliography:**


