Original Research Paper PATHOLOGY

Dediffrentiated Chondrosarcoma- A rare case report

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ABSTRACT

INTRODUCTION:-

Dedifferentiated chondrosarcoma was first proposed in 1971 by Dahlinand Beabout.[1] It occurs in patients older than 50 years and most commonly involves the femur, pelvis and humerus.[2] The male to female ratio is 1.3:1. They account for 10-15% of all chondrosarcomas and show an increased growth rate and rapid metastatic spread.[2] Dedifferentiated chondrosarcoma refers to the presence of a poorly differentiated sarcomatous component (osteosarcoma, fibrosarcoma, rhabdomyosarcoma and pleomorphic sarcoma with MFH like features) at the periphery of a typical low grade chondrosarcoma.

KEYWORDS : Chondrosarcoma, Neurosurgery, Dedifferentiated, Dura and Chondrocytes

CASE:-

40 years old female patient presented at neurosurgery outpatient department in RIMS, Ranchi, (Jharkhand) with swelling on the right parietal region since last 5 months. Her chief complaints were pain at the site of the mass with a gradual increase in its size, headache, weakness. On examination, the size of the swelling 6 x 6.5 x 3.5cm^3 with tenderness and soft to firm consistency. The swelling caused discomfort particularly during sleep. The swelling was attached to the skin and underlying structures. MRI revealed a large soft tissue lesion arising from the scalp in the right parietal region causing bony destruction and involving the underlying dura **(Figure-1).**

After the radiological report, surgery was done in the neurosurgery department and the specimen of the mass was sent in the pathology department for histopathological examination.

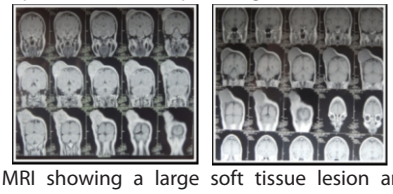

Figure-1: MRI showing a large soft tissue lesion arising from the scalp in the right parietal region causing bony destruction and involving the underlying dura.

PATHOLOGICAL FINDINGS:-

GROSS:

The mass measured 6 X 6.5 X 3.5cm3 with soft to firm in consistency. A few parts of the mass were nodular and there were also a few black patchy areas with greyish white areas on the outer surface. On cut section, shows greyish white with firm glistening tissue admixed with areas of fleshy tissue with areas of haemorrhage and necrosis.

Figure-2: Outer surface shows nodular appearance.

Figure-3: Cut section shows greyish white, fleshy tissue with area of haemorrhage and necrosis.

MICROSCOPIC FINDINGS:-

The section shows islands of chondrocytes juxtaposed to sarcomatous component containing multinucleated giant cells. The hypercellular section shows chondrocytes having plump vesicular nuclei with small nucleoli. The section also shows pleomorphism with bizarre tumour giant cells and mitoses (Figure-4 and 5).

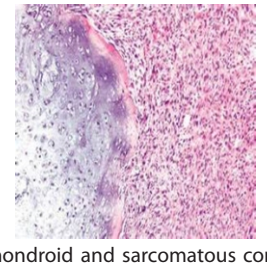

Figure-4: Shows chondroid and sarcomatous component. Multinucleated giant cell

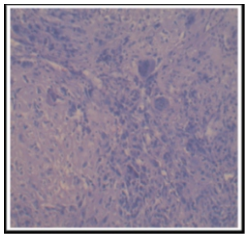

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Figure-5: Shows multinucleated giant cell.

DISCUSSION:-

Dedifferentiated Chondrosarcoma generally affects the age group ranging from 15-69 years with a slight male predominance.[3] Most common sites of involvement arefemur, pelvis and humerus.[4, 5] Peripheral dediffenetiated chondrosarcoma involves pelvis, scapula and ribs. In about 50% of dediffentiated chondrosarcomas, heterozygous mutations of isocitrate dehydrogenese 1&2 genes are found.[6] Usually the two components of the tumour can be seperately indentified grossly. Under microscopy the cartilaginous component ranges from enchondroma like appearence to grade I chondrosarcoma and occasionally has the features of a grade II chondrosarcoma.

Dedifferentiated component is sharply demarcated from the chondroid component, with no transition or intermidiate zone. The high grade component can show features of osteosarcoma, fibrosarcoma or Pleomorphic sarcoma with malignant fibrous histiocytoma like features.[7,8]Immunohistochemistry study showing S-100 chondroid component is positive and SMA, DESMIN, MYOGLOBIN, CD68 and CD34 is positive with depending on dedifferentiated component. Rarely, it can show features of angiosarcoma, leiomyosarcoma and rhabdomyosarcoma. Resp onse to post operative chemotherapy is poor in dediffere ntiated chondrosarcoma and it has a very poor prognosis.[9] Poor prognostic factors which are very commonly seen in this variant of chondro sarcoma are presence of pathological fracture, metastasis at time of diagnosis, pelvic location and increased age.[3] Most common site for distal metastasis is lung.

Patients with dedifferentiated chondrosarcoma have a poor prognosis. Reported survival rates at 5 years range from 7% to 24%. Radiation and chemotherapy have not been shown to improve prognosis. In this case, the site was quite unusual and there was no metastasis at the time of diagnosis.

CONCLUSION:-

From the above discussion we conclude that the site of the tumour is very rare. The metastasis of chondrosarcomas is also very common but in our case there was no metastasis present. With the help of radiotherapy and chemotherapy, the survival rates of the chondrosarcomas still remain poor.

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