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Case Report

Secondary malignant giant-cell tumor of bone

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ABSTRACT

Malignancy in giant cell tumor of bone (MGCTB) is extremely rare condition. We reported a case of 45 year female, with gradually increasing swelling over left wrist of 6 months. She was known case of giant cell tumor (GCT) of left radius operated 20 months back. Now on Magnetic Resonance Imaging (MRI) scan showed well defined mass lesion at distal end of forearm suggestive of recurrence of GCT. The excision of mass was done. On histopatholgical examination reported as secondary malignant GCT. We are presenting this case for its clinical, radiological and histopathological findings.

1. Introduction

Malignant giant cell tumor of bone (MGCTB) was first described by Stewart et al in 1938.[1] Malignant giant cell tumor of bone is extremely rare condition and occurring in less than 1% percent of cases.[2] Usually its diagnosis is difficult as there a lack of clear diagnostic criteria and terminology has been inconsistently used in the literature.[3] Secondary malignancy in GCTB is considered when the malignant changes occur at the site of previously treated GCTB. It typically follows the previous surgery or radiotherapy. Secondary malignant GCTB consist of sarcomatoid growth at the site of previously reported benign GCTB. While the primary malignant GCTB are considered when first diagnosis of GCTB with an area / nodules of highly pleomorphic mononuclear/ spindle cell within tumor.[4–6] Malignant GCTB is considered as high grade sarcoma.[7] We have a similar findings in our case.

2. Case Report

A 45 year female operated 20 months back for GCT of left radius. Now presented with 5x5x 4.3cm mass lesion at site of previous operation. It increased gradually in size within last 6 months. The MRI left wrist joint Plain and Contrast showed a well defined lobulated lesion (3.6x3.0x3.7cm) noted in dorso-lateral aspect of distal end of forearm. The lesion appears isointense on T l, heterogenously hyperintense with hypointense rim on T2. The lesion shows near homogenous post contrast enhancement. The lesion is seen abutting and displacing adjacent abductor pollicis longus, extensor carpi radialis longus and brevis tendons, and radial vessels medially. Magnetic susceptibilty artifact noted in proximal carpal row and distal end of forearm-post operative. Proximal carpal bones show osteopenic and degenerative changes. Fibular graft shows sclerotic changes and bone destruction at its distal end with thickened and irregular cortical margins-chronic osteomyelitis. Flexor retinaculum appears grossly normal. Impression was given as , in a post operated case of giant cell tumor of distal end of radius, well defined lobulated lesion in dorso-lateral aspect of distal end of forearm showing near homogenous post

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contrast enhancement suggestive of soft tissue recurrence of giant cell tumor.

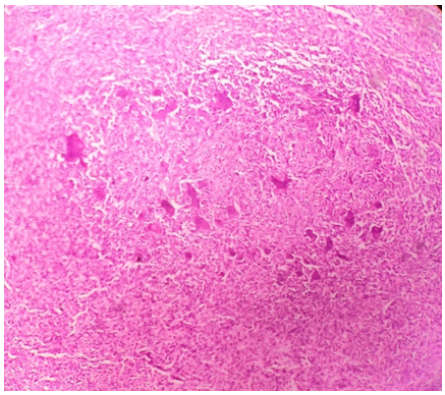

Fig. 1: Tumorcomposed of bimodal population of proliferating mononuclear cells with numerous osteoclast like multinucleated giant cell (H& E Stain,40x).

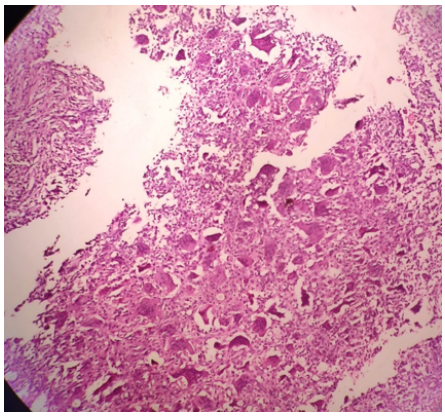

Fig. 2: Tumor composed of bimodal population with numerous osteoclast like multinucleated giant cell (H& E Stain,40x).

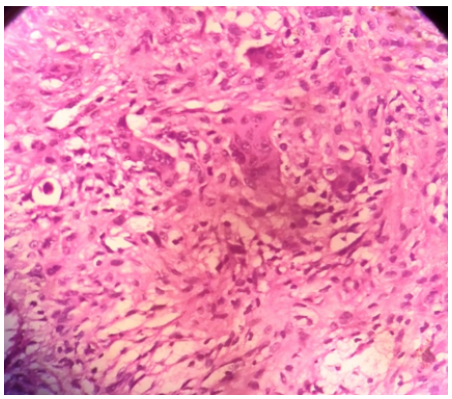

Fig. 3: Areas showing spindle cell, severe nuclear pleomorphism (H& E Stain,100x).

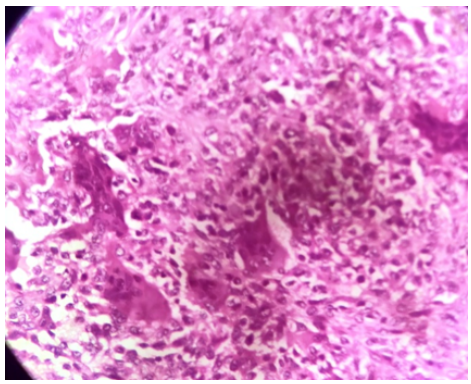

Fig. 4: Areas showing spindle cell, severe nuclear pleomorphism, multinucleated giant cells were uniformly distributed (H& E Stain,100x).

Patients other systemic examination, there was no any significant disorders noted. We received for histopathological examination an irregular gray, white, friable soft to firm tissue measuring 4.1x3.5x3 cm .On cut section showed fleshy nodular tumor with areas of hemorrhage and cystic changes.

On microscopic examination of multiple section showed a tumor composed of bimodal population of proliferating mononuclear cells with numerous osteoclast like multinucleated giant cells ($). The mononuclear cells were elongated spindle cells arranged in sheets, irregular contour and in areas storiform pattern. In areas spindle cell showed severe nuclear pleomorphism. The multinucleated giant cells were uniformly distributed and cells are round to oval having granular cytoplasm nucleus having one to two prominent nucleoli. (Figures 3 and 4). The giant cells having abundant eosinophilic cytoplasm with 20-40 nuclei. In areas vascular proliferation and fibrosis was noted. At places atypical mitotic figures >20/10 hpf were noted. We reported on histopathology in view of recurrence,pleomorphism, atypical mitotic figures > 20 /10 hpf, lesion is of secondary malignant giant cell tumor of excised mass from the left wrist.

3. Discussion

The most GCT of bone are benign tumors but have locally aggressive behavior. It is observed that GCT have high rate of recurrence upto 25%. In our case the secondary malignant giant cell tumor from left wrist was noted in recurrence. On histopathologically GCTB is classified as a) grade I as benign which shows no appreciable stromal cell abnormality, b) grade II as intermediate tumor which shows mild to marked stromal cell abnormality, c) grade III as malignant tumor which shows features of high grade stromal malignancy.[7] Due to limited clinical value this classification system is no longer used. The benign GCT in 2% of cases may show evidence of metastasis in lungs which is gradually

and may regress spontaneously.

Malignancy in GCTB is also called as differentiated GCT. It indicates high grade sarcoma developing at the site of previously documented benign GCTB. Previously excised or irradiated GCT called as evolutionary and radiation induced forms respectively. The secondary GCT incidence is very low and accounts for 1 to 2.4%.[8] It is usually seen at older age. In our case it is reported in relatively early age. The most common primary symptoms of the malignant form are pain and swelling.

The malignant transformation of GCT is proliferative changes occur at the site of curretage and bone grafting or it can follow surgery without adjuvant radiotherapy.[9]

As per the histopathological criteria given by Hutter et al.[5] and Dahlin et al.[4] MGCTB shows plump spindle or ovoid shaped stromal cells. The multinucleated giant cells are seen intermingling. The neoplastic spindle nuclei were hyperchromatic, elongated and vesicular, most of cells having prominent nucleoli. The cell having eosinophilc cytoplasm. Increased in mitotic activity with atypical mitosis are noted. The intercellular substance usually absent .The comprehensive histologic sampling is essential to ensure accurate diagnoses of malignant GCT. The secondary MGCTB showed strong expression of p53.

The MGCTB should be considered as a high-grade sarcoma and it must be distinguished from GCTB and other malignant tumors containing giant cells. The differential diagnosis with giant cells are osteosarcoma, fibrosarcoma, high-grade myxofibrosarcoma, leiomyosarcoma and undifferentiated high-grade pleomorphic sarcoma.[10,11]

Secondary malignancy in giant cell tumor of bone has a poor prognosis .In various studies it was observed that the prognosis was better in patients with primary malignancy in giant cell tumor of bone than secondary malignancy. As there is lack of long-term follow-up data on the prognosis of malignant GCTB is not fully understood. The 5-year disease-free survival in patients with secondary malignancy was 32% as observed in study at the Mayo Clinic.[6]

4. Conclusion

Secondary malignant giant cell tumor of bone is extremely rare condition. As it has a poor prognosis, it is important to have proper diagnosis and appropriate management. We are presenting this case for its clinical, radiological and histopathological findings.

5. Conflict of Interest

The authors declare that there is no conflict of interest.

6. Source of Funding

None.

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