CASE REPORT

Primary Malignant Giant Cell Tumour of the Bone: A Case Report

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Abstract

GCT is a primary locally aggressive bone neoplasm affecting the epiphyseal or meta-epiphyseal regions of bone. GCT can be rarely multifocal which may be synchronous or metachronous. At the same time, it is pathologically difficult to differentiate benign GCT from a malignant one. Malignant GCT can be a primary malignant GCT or a secondary malignant GCT. The prognosis of GCT is based on clinical progression rather than the histological picture. Here we report a case of a 20 year old male who presented to us with a swelling of right ilium which was treated by curettage and bone cement fixation, but later went on to progress as a primary malignant metachronous lesion and eventually leading to his death within 2.5 months after the index surgery.

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Introduction

Giant cell tumour (GCT) of the bone is a primary locally aggressive bone neoplasm characterized by stromal mononucleate cells associated with uniformly distributed osteoclast-like multinucleated giant cells. Skeletally mature patients ranging from 20 to 45 years of age, especially women are affected. GCT usually involves the epiphyseal or meta-epiphyseal region of long bones, particularly the femur, tibia and distal radius. It appears radiographically as a purely osteolytic eccentric lesion.[1]

A GCT can be rarely multifocal which may be synchronous or metachronous. A synchronous GCT is one which has occurred at two sites within a span of 6 months. A metachronous GCT is one which occurred at two sites at more than 6 months interval.[1]

Though classically described as a benign lesion, GCT is an aggressive lesion and at times produces metastasis in the lung. Aggressiveness of GCT is unpredictable clinically, radiologically and histologically. This is the reason why WHO had shifted the position of GCT of the bone from benign to intermediate group with some other bone tumours like osteoblastoma. GCT is now considered as more aggressive than a Grade1 chondrosarcoma.[1]

Malignant GCT is a totally different entity and is an extremely rare condition than previously thought[2–4,6]. Presence of metastasis in GCT does not make it a malignant one. Metastatic lung lesions of GCT will have the same histological picture and are very slow growing. Malignant GCT is of two types. (1) Primary malignant GCT is a lesion in which there are areas of synchronous high grade sarcomatous growth next to areas of benign giant cell tumour. So it is also known as dedifferentiated giant cell tumour. (2) Secondary

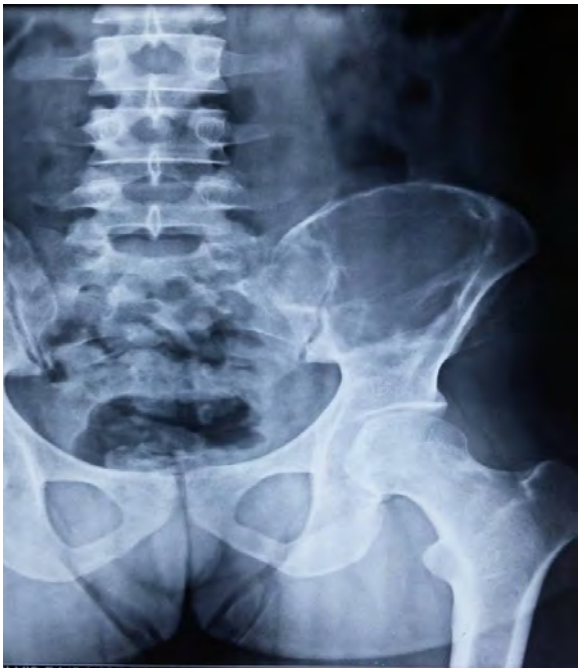


Figure 1. X-ray showing lytic lesion of Ilium.

malignant GCT is a benign GCT which has turned malignant following radiotherapy or surgery.[1–4] Primary malignant GCT is much more rare than Secondary.

In our case report we deal with a case of GCT which is primary malignant and metachronous.

Case Report

A 20 year old male presented in February 2016 with pain and swelling of right ilium of 2 months duration. He had a past history of right petro-mastoid extradural lesion (benign GCT with secondary ABC) for which he underwent lateral sub occipital craniotomy and gross total excision in 2009 followed by radiotherapy. *This was 7 years before the present episode.*

On examination there was a large 20 × 10 cm size lesion in the posterior portion of right ilium which was tender with local rise in temperature (Figure 1). He was unable to bear weight on that extremity. CT scan of pelvis showed a large lytic lesion in the right iliac bone, probably GCT. CT guided core needle biopsy report came as giant cell rich lesion possibly GCT. Patient underwent extended curettage followed by bone cement under GA (Figures 2–4). Immediate post op period was uneventful. HPR report came as GCT with extreme necrosis and focal areas of marked pleomorphism suggestive of malignant change (Figure 5). Further radiotherapy was planned, but since the flap was unhealthy, it was delayed.

One month later patient presented to our casualty with fever, nausea, vomiting and oozing from the wound. Local examination revealed swelling around the operated site with wound gaping and blood stained

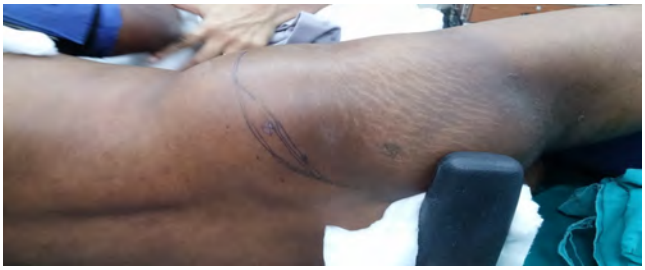


Figure 2. Pre op marking for skin incision.

Figure 3. Intra operative picture of curetted cavity.

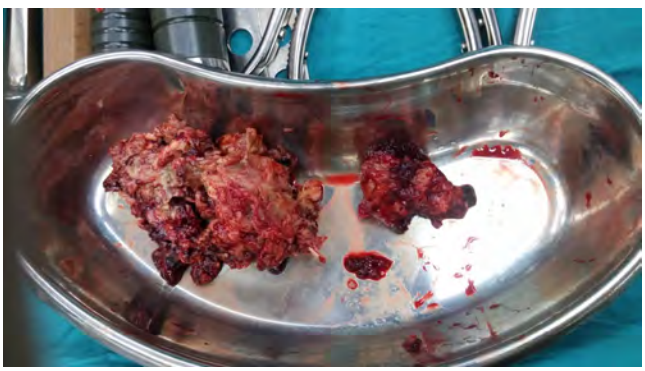


Figure 4. Curetted specimen of giant cell tumour.

serous fluid oozing from the surgical site. CT scan of the abdomen and lungs was suggestive of recurrent lesion with wide spread metastases (Figure 6(a)–(b)).

Chemotherapy, denosumab and radiotherapy after tumour debulking were considered but were deferred due to poor financial condition of patient and unpredictable results. Due to worsening general condition, relatives opted for palliative care at home. It was later known that patient succumbed to the illness 2 weeks after discharge.

Discussion

GCT has higher incidence rates in India as compared to the west, comprising 20% of all primary bone tumours compared to 4% to 8% in the U.S. and Europe.[5,7].

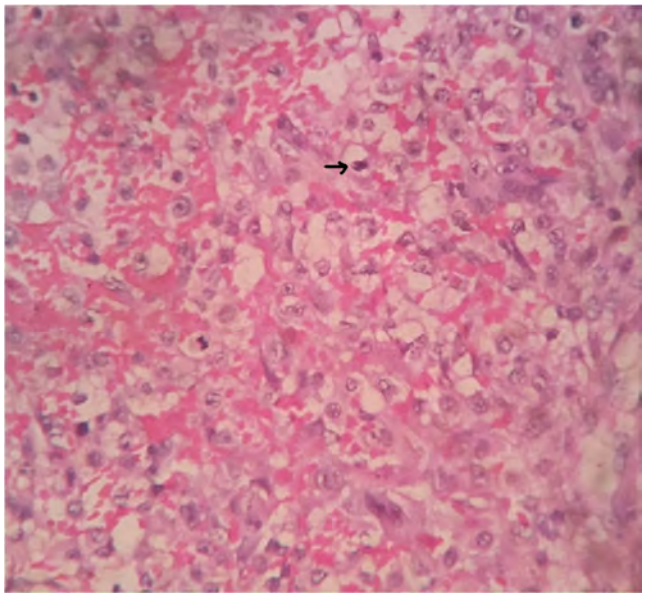
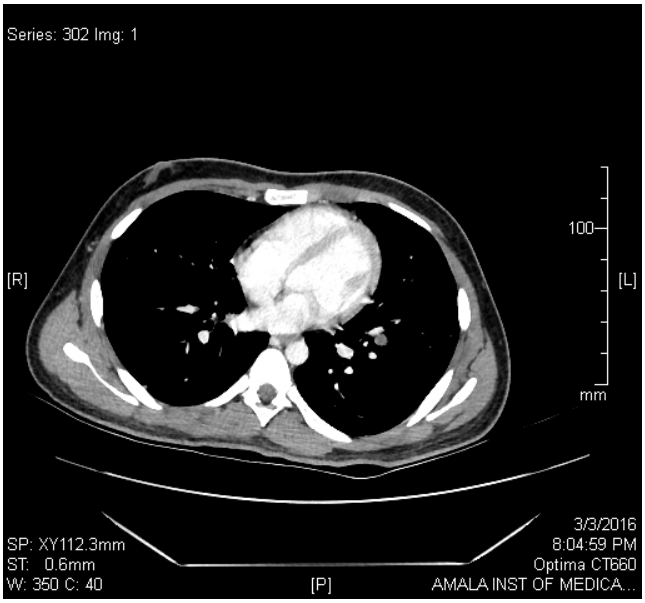
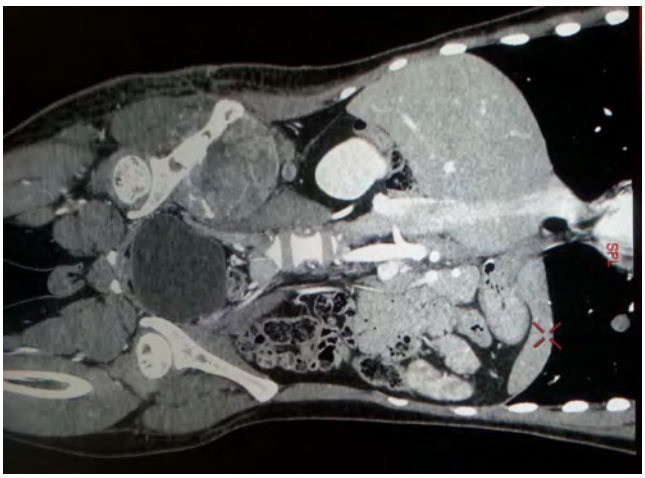


Figure 5. Pleomorphic areas with increased mitosis (arrow) 40×.



(a)



(b)

Figure 6. (a) & (b) CT image showing tumour and lung metastasis.

On analysis of a Tumour Registry maintained by the first author, GCT of the bone comes first in number among primary bone tumours including benign and malignant. (There were 347 cases of GCT of the bone treated from our institute from 1988 to 2016[8,9]) 6 of our cases had lung metastases. 2 cases were multifocal and synchronous excluding this case. It is for the first time that we have encountered a malignant GCT which is metachronous, which makes this case worth reporting.

The features of both benign and malignant GCT are very similar. In malignant GCT, sarcomatous features are present within the stromal components including cells with hyper chromatic nuclei with variably prominent nucleoli, mitotic activity and atypical mitosis. Malignant spindle cells may also be found infiltrating the bony trabeculae.[2,4] In our case report, most of the area showed conventional GCT but focal areas of necrosis and cellular pleomorphism were also found. Some foci showed eosinophilic material resembling osteoid.

Immunohistochemistry can be helpful in narrowing down some of the differential diagnoses, but the stains are highly non specific to GCT with malignant transformation .Consequently, the histological evaluation with basic haematoxylin and eosin is the most indicative diagnostic tool in determining malignant transformation of GCT of bone.[10]

Due to the extremely short life span between initial occurrence and when the tumour recurred this was most likely a primary malignancy of GCT of bone that was conceivably missed on initial biopsy. It is probable that the tumour was malignant from the start, and therefore not a recurrence. Looking back we should have treated this case more aggressively. But there was nothing to suggest malignancy in the initial biopsy and even if an open biopsy was done, the pathologist might still not be able to predict the malignancy as most of the areas showed conventional GCT.

Conclusion

Malignant GCT is an extremely rare condition. Malignant transformation can be difficult to diagnose clinically, radiologically and grossly due to the fact that the malignant components are interspaced within benign components which can be easily missed on biopsy or when sections are taken at the grossing bench. By generously sampling these cases, especially areas that appear to have a necrotic appearance, the

likelihood of missing the malignant components is minimised.

References

1. Christopher DM Fletcher, Julia A Bridge International agency for research on cancerWHO classification of tumours of soft tissue and bone, 2013.
2. Megan Greene, Mayo Clinic, Rochester MN, Malignant transformation of GCT: a casestudy; www.pathassist.org/resource/resmgr/2014_Handouts/GreeneMegan.pdf.
3. Franco Bertoni. Patrizia Bachini Malignancy in giant cell tumour of bone. Cancer 2003 May 15(2520–2529).
4. Rosalvo Zósimo Bispo Júnior I, Olavo Pires de CamargoII Primary malignancy in giant cell tumour: a case report. Sao polo Medical journal 2009.
5. Rao PT. Management of giant cell tumor of bone. Kini memorial oration. Indian J Orthop 1993;27:96-100.
6. Julia Rockberg, Bruce A. Bach, Incidence Trends in the Diagnosis of Giant Cell Tumor of Bone in Sweden Since 1958; J Bone Joint Surg Am, 2015 Nov 04; 97 (21): 1756–1766.
7. Huvos AG. Bone tumors: Diagnosis, treatment and prognosis. 2 nd ed. WB Saunders Co: Philadelphia; 1991.
8. Puthoor DK, Puthezhath K. Management of giant cell tumor of bone: computerized tomography based selection strategy and approaching the lesion through the site of cortical break. Orthop Surg. 2012 May;4(2):76-82. doi:10.1111/j.1757-861.2012.00172.x. PMID:22615151.
9. Dr. Dominic Puthur and Dr. Wilson Iype Management of GCT by curettage and bone grafting Indian Journal of Orthopaedics, vol 41 issue 2 April-June 2007 (121-123).
10. Gering L, Liu W, Sun X Histological and chemical characteristics of malignant giant cell tumour of bone Virchows archiv March 2012.