

Case report: Atypical presentation of giant cell tumour (GCT) of bone in the distal humerus of a child

M Tomlinson MBBCh

GJ Kubicek MD, MRCS aff. (London, UK)

N Ferreira BSc, MBChB, FC Orth(SA), MMed(Orth, PhD)

LC Marais MBChB, FCS Orth(SA), MMed(Ortho), PhD

Tumour Sepsis and Reconstruction Unit, Department of Orthopaedic Surgery, Grey's Hospital, Nelson R. Mandela School of Medicine, University of KwaZulu-Natal

Corresponding author:

Dr Nando Ferreira

Department of Orthopaedic Surgery

Grey's Hospital

3201 Pietermaritzburg

KwaZulu-Natal

South Africa

Tel: +27 33 897 3000

Email: Nando.Ferreira@kznhealth.gov.za

Abstract

Introduction: Giant cell tumour of bone is a rare entity in children and seldom involves the distal humerus, even in adult patients. We present a case of giant cell tumour in the distal humerus of a child.

Case report: A 12-year-old boy presented to our tertiary level tumour unit with progressive pain in his left elbow and was radiologically and histologically found to have a giant cell tumour of his left distal humerus. He was treated with extended curettage, cryotherapy and PMMA cement with the addition of a supportive posteromedial locking plate. There was no recurrence noted at one-year follow-up.

Discussion: The literature is reviewed to determine the epidemiology of this tumour, the histological and radiological findings unique to the lesion and the current best practice of treatment.

Conclusion: A rare case of giant cell tumour in the distal humerus of a child is presented. Giant cell tumour of bone should be considered in the differential diagnosis of a lytic metaphyseal lesion if the growth plates are open.

Key words: giant cell tumour of bone, skeletally immature, distal humerus

<http://dx.doi.org/10.17159/2309-8309/2016/v15n1a2>

Introduction

Giant cell tumour (GCT) is a primary benign bone tumour that is locally aggressive, rarely metastasises and seldom undergoes malignant transformation. It is diagnosed with combined radiological and histological assessment. GCT is infrequently seen before the closure of skeletal growth plates. When they occur in children, these lesions are generally localised in the metaphysis, as compared to adults where GCTs usually occur in the epiphysis. The ends of long, tubular bones are chiefly affected with the distal femur, proximal tibia and distal radius most commonly involved.

We present an especially unusual case of GCT that occurred in both a rare location (the distal humerus) and in a skeletally immature patient.

Case report

A 12-year-old male child was referred to our tertiary level tumour unit with a three-month history of progressive pain and swelling of the left elbow. The child was otherwise well, reporting no history of trauma to the elbow nor any constitutional symptoms.

The patient was admitted for local and systemic staging, which was followed by an incisional biopsy. Local staging included radiographs and a magnetic resonance imaging (MRI) scan. The radiographs showed a locally aggressive, geographic lytic lesion eccentrically located in the distal humeral metaphysis. The lesion had expanded the cortex, permeated through the bony confines, extended into the surrounding soft tissues, and no periosteal reaction was noted. According to

Campanacci radiographic staging system this lesion was classified as a grade III GCT¹ (Figure 1). MRI scans revealed a large heterogeneous T2 hyperintense, T1 hypointense lesion (Figures 2 and 3). Systemic staging confirmed that the patient was generally well with no co-morbidities and a normal nutritional status. Computerised tomography (CT) scan of the chest and abdomen did not identify any metastatic lesions and a skeletal survey confirmed a single lesion located in the left distal humerus.

An incisional biopsy was performed through a postero-medial approach. The histological evaluation described a GCT with no malignant features and confirmed the classic findings of mononuclear cells with interspersed fibro-collagenous strands and numerous multinucleated osteoclast-like giant cells (Figure 4).

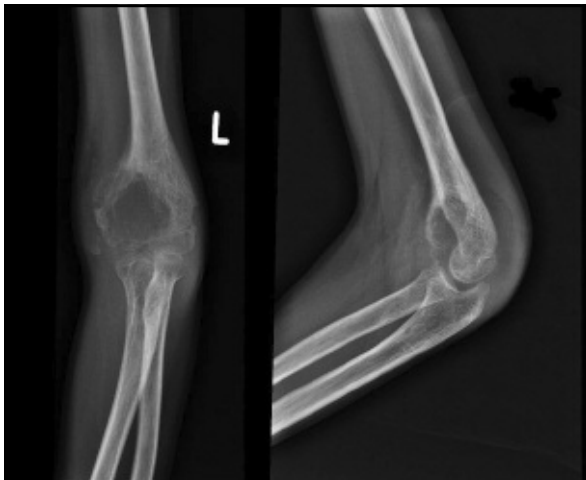


Figure 1. AP and lateral radiographs showing a large geographic lytic lesion of the left distal humerus with cortical expansion and destruction and extension into the physis and epiphysis

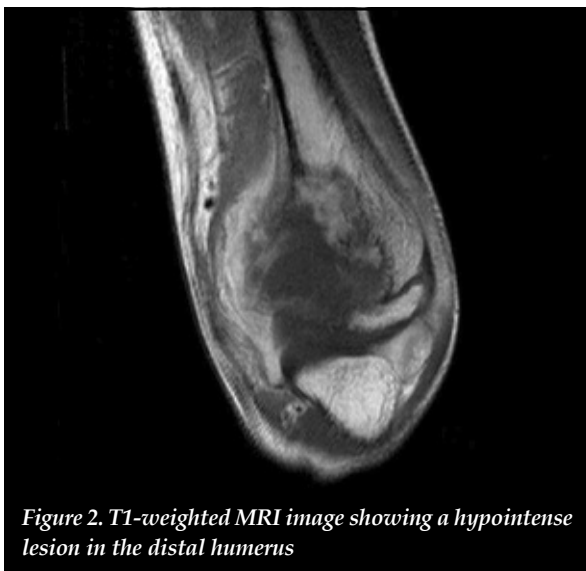


Figure 2. T1-weighted MRI image showing a hypointense lesion in the distal humerus

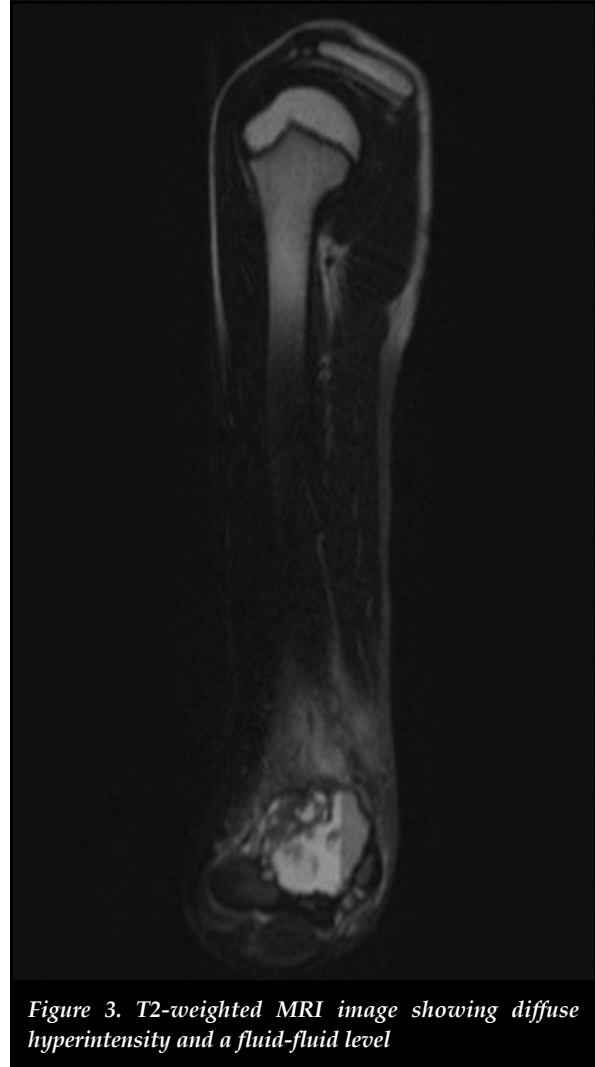


Figure 3. T2-weighted MRI image showing diffuse hyperintensity and a fluid-fluid level

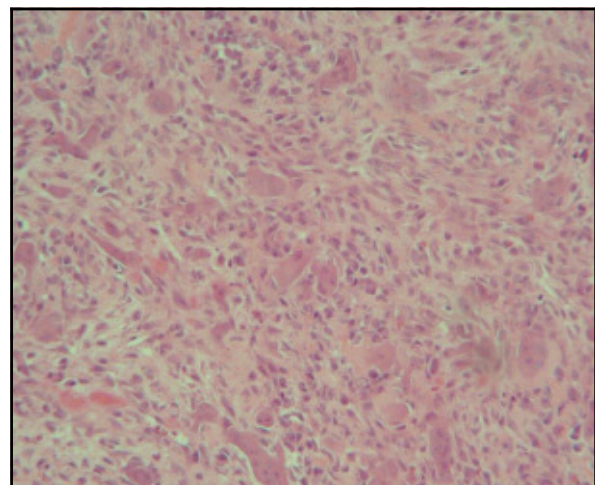


Figure 4. Histological specimen showing numerous multinucleated giant cells on a background of homogenous mononucleated cells. There is no evidence of atypia of the spindle cells



Figure 5. AP and lateral radiographs at a follow-up one year after surgery showing no signs of recurrence



Figure 6



Figure 7

Figure 6 and 7. Elbow range of motion of 0°–110°, one year after surgery

Definitive surgery was performed seven days after the histological confirmation was obtained. This was performed through a postero-medial approach based on the biopsy incision and consisted of curettage, extended with high-speed burring, and adjuvant cryotherapy with liquid nitrogen. The resultant cavity was filled with PMMA cement and supplemented with a postero-medial locking plate for stability (Figure 5). Confirmatory histology from the definitive procedure confirmed the incisional biopsy results of a benign GCT. At one-year follow-up the patient reported no pain, and a 0°–110° range of motion of the elbow. There were no clinical or radiological signs of recurrence (Figures 6 and 7).

Discussion

The World Health Organisation (WHO) classifies giant cell tumour of bone as a locally aggressive, potentially malignant lesion that can rarely produce metastases.² The tumours generally develop slowly, metastasise in 1–3% of cases and do so most commonly to the lungs.³ Recurrence rates of between 0% and 63% have been published and depend largely on the type of treatment.⁴ Malignant transformation occurs in 5–10% of cases and most commonly follows radiation therapy of the primary lesion.⁵ Primary malignant GCT is a rare entity where synchronous coexistence of a sarcoma and benign GCT is found in the same lesion.^{2,6,7} Multicentric tumours are rare (<1%) and are associated with a more aggressive course.⁸

These lesions are classically described in individuals between the ages of 20–40 years with a slight female predominance and are rare in children before the closure of growth plates.^{2,6,9} The incidence in children younger than 19 years has been reported in several studies and ranges from 1.7% to as high as 10.6% of all patients with GCTs.^{10–15} GCTs are typically located in the epiphyseal region of long bones with the proximal tibia, distal femur and distal radius being the most commonly affected.² In children there seems to be a relationship between age and proximity to the epiphysis. In young children the tumours are generally further from the epiphysis and more metaphyseally located.^{10,12,15}

Involvement of the humerus comprises approximately 6% of all GCTs, but almost all of these are located in the proximal humerus with distal humeral lesion being exceedingly rare.¹⁶ The current case demonstrates a rare occurrence of a GCT in the distal humerus of a child. At present only a single occurrence of distal humerus GCT has ever been reported in a child. This case, however, involved multicentric disease with foci also reported in the hand.¹⁷

GCT of bone is diagnosed histologically by the characteristic appearance of a heterogeneous group of cells containing most notably mononucleated plump spindle- or ovoid-shaped cells and osteoclast-like multinucleated giant cells. The histology found in this case represents a Jaffe grade I lesion.

Although Jaffe's grading system was developed in order to predict patient outcomes, by his own admission, he was unable to correlate the histological findings with the probability of the lesion undergoing malignant transformation, the incidence of metastases or the frequency of recurrence.^{2,18}

Typical radiographic findings include lytic lesions located eccentrically in the epiphysis that do not penetrate the joint. The rim of the lesion is generally not sclerotic and can extend into the surrounding tissue, which indicates a poorer prognosis. Differential diagnoses based on radiological findings include aneurysmal bone cyst, non-ossifying fibroma, primary hyperparathyroidism with brown tumours, chondromyxoid fibroma, chondroblastoma, clear cell chondrosarcoma and telangiectatic osteosarcoma. Researchers using MRI have found haemosiderin deposits in up to 60% of cases and although these findings are neither sensitive nor specific, it may be a useful adjunct in determining a diagnosis.¹⁹

Surgical treatment usually consists of extended curettage or en-bloc resection. Current evidence indicates that curettage should be supplemented with a combination of chemical cautery, cryotherapy or electrical cautery followed by methylmethacrylate cement to limit recurrence.^{20,21} In the instance of this case a combination of curettage, burring and cryotherapy was used, after which cement was inserted in the cavity. The distal humerus was then stabilised with a postero-medial plate. At the one-year follow-up the patient had no clinical or radiographic signs of recurrence. We continue to follow him up, as a minimum of three years' follow-up is suggested in GCT as most recurrence is seen within this period.²⁰

Conclusion

This case report describes a rare occurrence of a GCT in the distal humerus of a child, and includes the diagnostic and treatment algorithms. Giant cell tumours have a variable presentation and should form part of a differential diagnosis of lytic metaphyseal bone lesions in children.

Consent

Written consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Conflict of interest statement

The authors declare that they have no conflict of interests and no financial support was received for this study.

Authors' contributions

All authors made contributions toward the conception and design of the research, acquisition of data and drafting of the manuscript. The final manuscript was read and approved by all the authors.

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