

Giant Cell Tumor of the Distal Ulna: Multimodal Radiological Investigation of a Very Rare Location

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Patient: Male, 28-year-old
Final Diagnosis: Giant cell bone tumor • osteoclastoma
Symptoms: Wrist swelling and pain
Medication: —
Clinical Procedure: Radiology imaging • orthopaedic surgery
Specialty: Orthopedics and Traumatology • Radiology

Objective: Rare disease

Background: Giant cell tumor is a rare tumor of mesenchymal origin. According to World Health Organization classification, it is considered a benign tumor with locally aggressive characteristics and the capacity to metastasize. The tumor typically occurs in the epiphyseal regions, most often of long bones after the completion of bone growth. The disease is characterized by severe pain and swelling of the affected area. Tumor growth is expansive but relatively slow. The tumor rarely metastasizes, but when it does, the lungs are primarily affected.

Case Report: A 28-year-old man, otherwise healthy, presented with pain in the right wrist joint, limited range of motion, and spindle-shaped thickening/swelling in the same area, which he had noticed several months earlier. After a comprehensive diagnostic evaluation (wrist X-ray, computed tomography, magnetic resonance imaging, ultrasound-guided biopsy, and histopathological analysis), he was diagnosed with giant cell tumor of the right ulna. The tumor was surgically removed with good recovery, and the patient continued to be seen thereafter in regular followup.

Conclusions: The wide range of benign and malignant differential diagnostic entities requires a detailed diagnostic approach and comprehensive assessment, using different radiological modalities, as was done in this case. The final diagnosis was confirmed by histopathological analysis of core biopsy material.

Keywords: Elasticity Imaging Techniques • Giant Cell Tumor of Bone • Magnetic Resonance Imaging • Multidetector Computed Tomography • Radiography • Ultrasonography

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Background

Giant cell tumor of bone (GCTB) or osteoclastoma is a rare neoplasm. Although cases of GCTB were described and treated in the 1800s, the term GCTB was coined by Bloodgood, while Jaffe differentiated it from other giant-cell skeletal lesions and proposed a histological grading [1-3]. The World Health Organization (WHO) classifies GCTB as a primary benign mesenchymal tumor. Because of its potential to metastasize, it is considered to be a borderline neoplasm. Due to its rare occurrence, accounting for approximately 4.9-9% of all primary osseous neoplasms, little is known about the epidemiology of malignant GCTB. The largest systematic research studies on

the occurrence and distribution of GCTB were conducted in 2 populations: American (estimated US incidence was 1.6 per 10 million persons per year) and Chinese (incidence ranging between 1.49 and 2.57 cases per million persons per year). Both of these studies confirm the rare occurrence of GCTB and suggest that age and stage at the time of diagnosis are strongly associated with long-term survival [4,5]. In most cases, GCTB occurs in the metaphyseal and epiphyseal regions of long tubular bones, most commonly in the distal femur, proximal tibia, and distal radius. Other rare locations are feet, hands, patella, and talus. The distal ulna is a very rare location for giant cell tumors, with reported incidence of 0.45-3.2% [6,7]. GCTB lesions include locally aggressive neoplasms characterized by

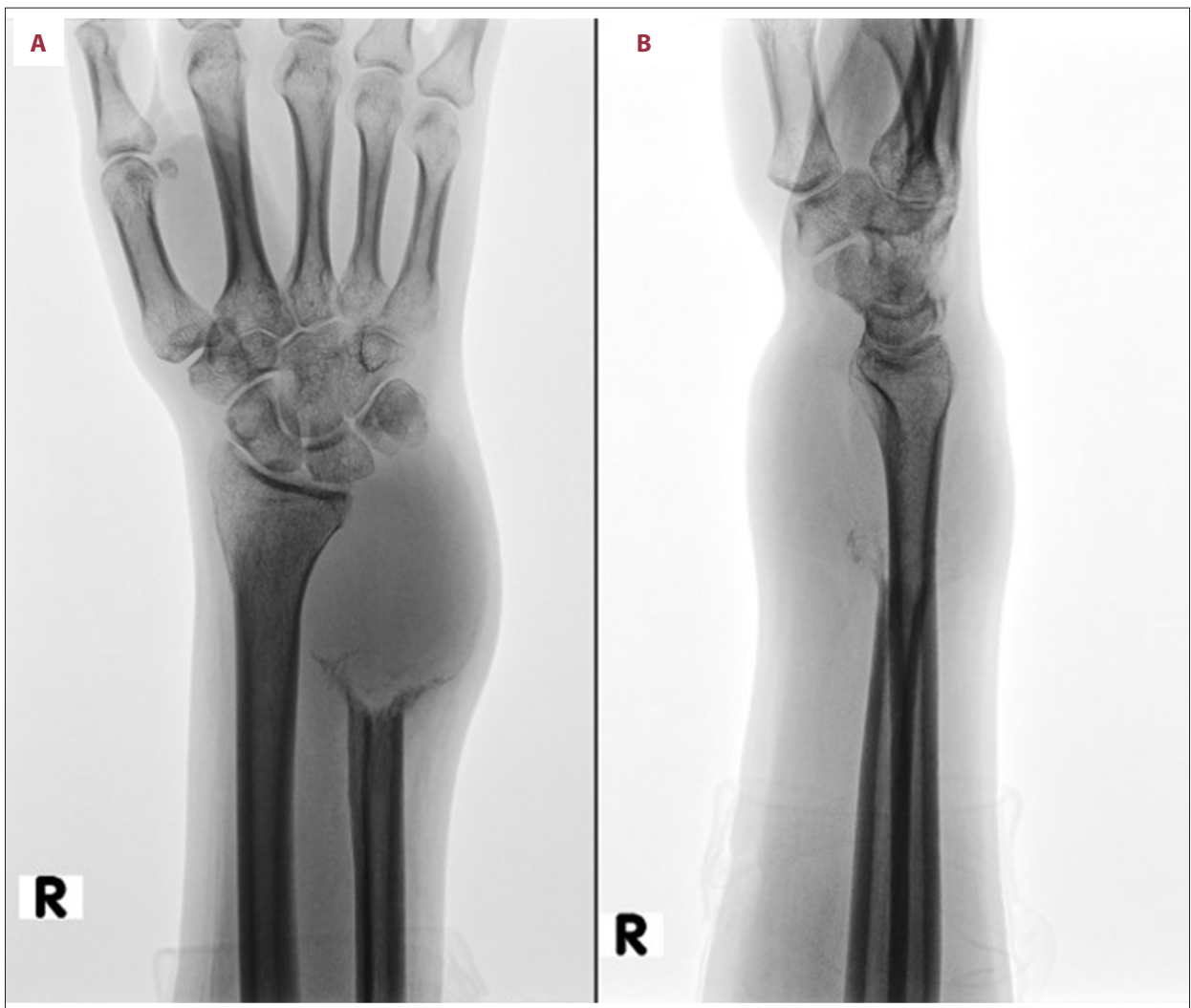


Figure 1. Antero-posterior (A) and latero-lateral (B) X-ray images of the right forearm and wrist demonstrate a lytic expansile lesion involving the distal epiphysis of the ulna, which is completely consumed. “The osteolytic features are so prevalent as to give the roentgen-ray picture of a *tabula rasa*” [17]. The lesion margins are irregular with an aggressive appearance and relatively broad zone of transition without sclerotic zones. There is no convincing periosteal reaction. No evidence of calcifications can be seen within the tumor stroma. The styloid of the radius, carpal bones, and bases of the metacarpal bones seem osteopenic; cortical bone is intact.

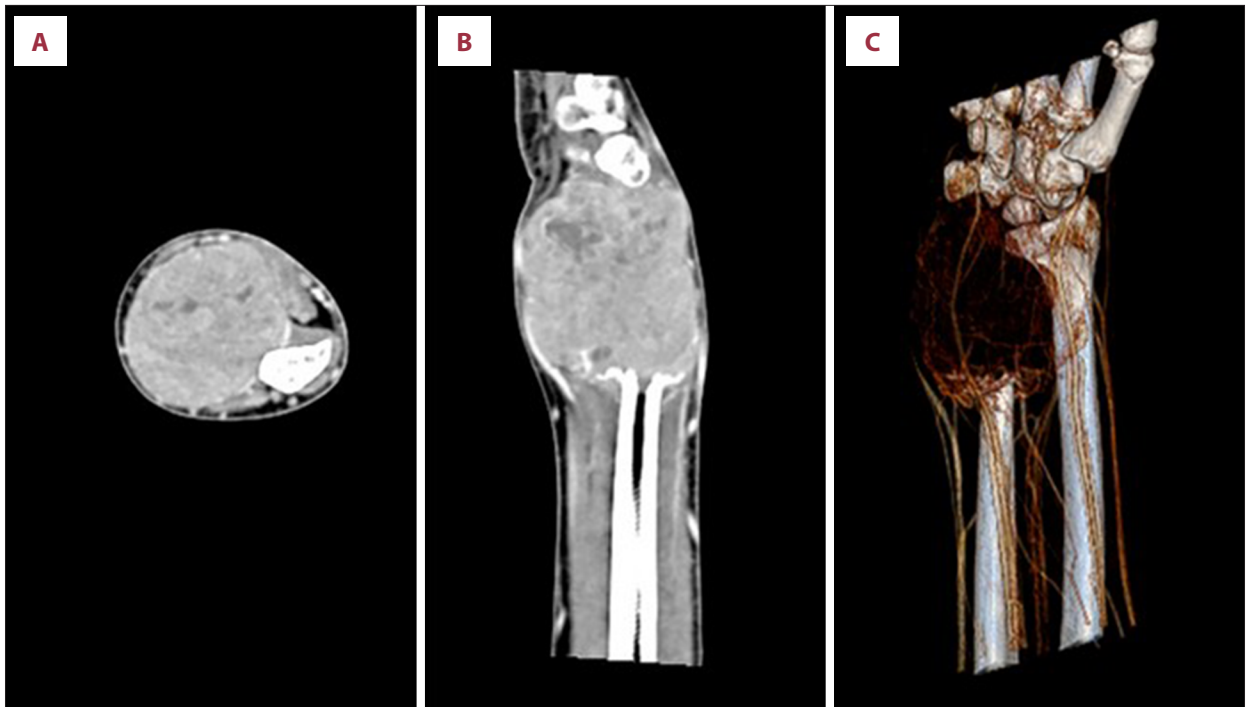


Figure 2. Postcontrast axial (A) and sagittal (B) CT slices confirmed an expansile lytic tumor of the distal ulna, with somewhat heterogeneous, partly cystic stroma, without calcifications. Significant contrast uptake is seen in the tumor stroma. No signs of infiltration of surrounding soft tissues. Volume Rendering Technique image (C) demonstrates lysis of the bone, abundant vascularization of the tumor, as well as major blood vessels diverging around the lesion.

mononuclear mesenchymal tumor cells that are mixed with multinucleated osteoclast-like giant cells as reactive infiltrate. The latter is present in a variety of bone lesions, including brown tumor of hyperparathyroidism (Recklinghausen disease), giant-cell reparative granuloma, aneurysmal bone cyst, chondroblastoma, giant-cell osteosarcoma, and malignant and benign GCTB [6,8-10]. Early diagnosis of GCTB in clinical practice is quite challenging because characteristic symptoms and physical examination findings overlap with several conditions of similar clinical presentation, as noted above. For the same reason, numerous synonyms have been used for GCTB, including myeloid sarcoma, tumor of myeloplexus, and osteoblastoclastoma. Typical clinical presentation includes soft tissue swelling and pain over the affected area with mechanical difficulties and pathological fractures due to osteolytic bone lesions. This variety of differential diagnoses is a reason for previously incorrectly used synonymous terms for GCTB. The final diagnosis is crucial because of the effective therapeutic approach and can be accurately accomplished by multimodal radiological imaging and histopathological biopsy analysis.

This case report focuses on “recognition patterns” of GCTB in various radiology imaging modalities. As such, in addition to presentation as a case report, it is presented as educational material to help clinicians in the identification of similar cases in their clinical practice.

Case Report

A 28-year-old male patient was referred to our department for radiological diagnostic proceedings due to distal right forearm discomfort and pain associated with wrist swelling lasting about 6 months. The wrist joint had limited range of motion. Skin in the swollen area was of normal color and temperature. The lesion was firm and tender on palpation. The patient’s past medical history, including family medical history, was not significant.

Standard wrist radiography in posteroanterior and lateral projection revealed a wide osteolytic lesion of the distal ulna involving the epiphysis, the metaphysis, and the distal third of the diaphysis. The radius and carpal bones were not affected (Figure 1).

Computed tomography (CT) exam revealed a well-circumscribed, lytic, expansile soft tissue mass (dimensions 49×67×48 mm) arising from the distal ulna and showing heterogeneous contrast enhancement (Figure 2). Necrotic parts of the lesion presented as irregular hypodense areas without contrast enhancement. The corresponding magnetic resonance imaging (MRI) confirmed heterogeneous enhancement of solid tumor components and amorphous, curvilinear necrotic areas (Figure 3). Fat components, hemorrhage, and calcifications were not present in the tumor stroma. On both CT and MRI, there were no

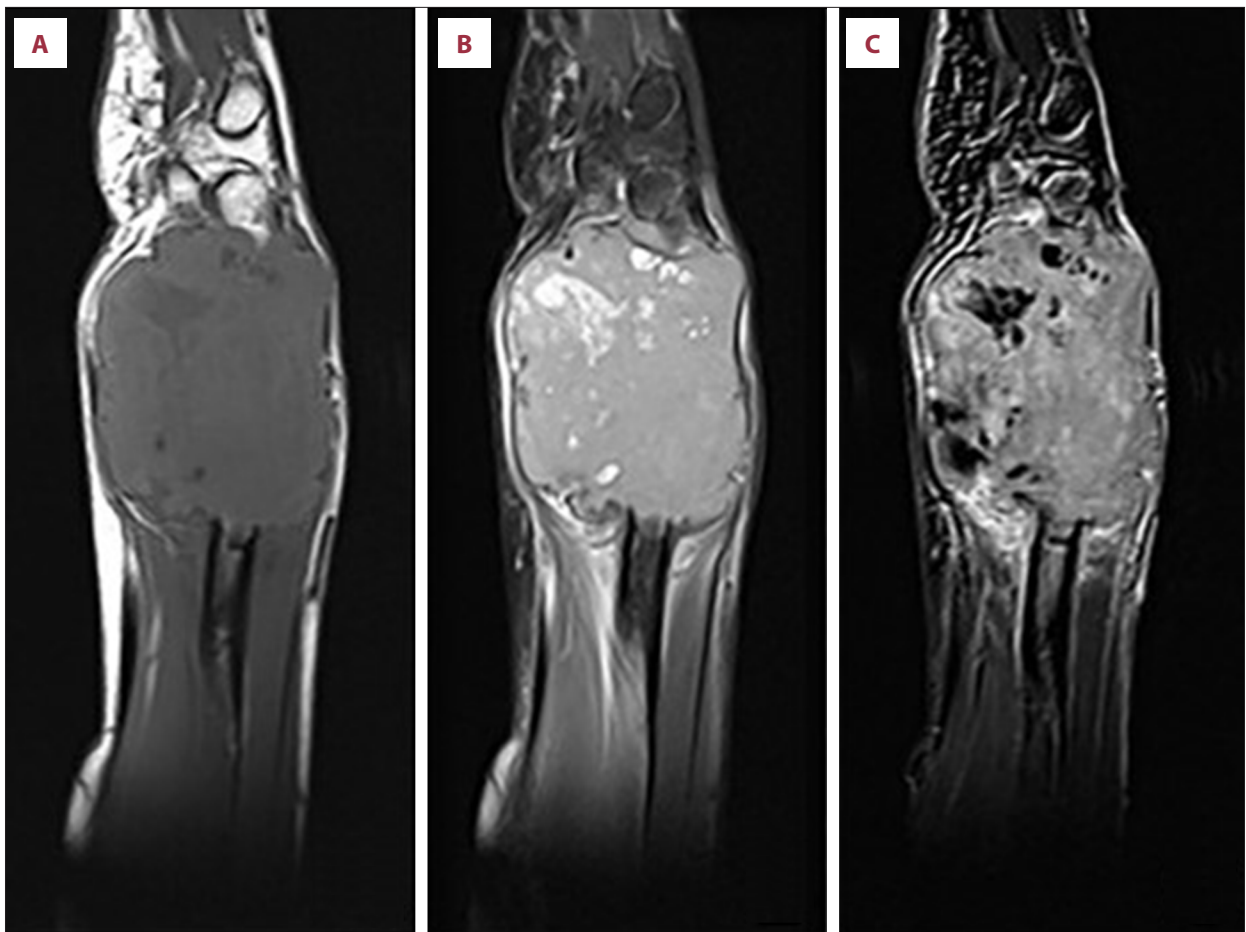


Figure 3. Sagittal MRI obtained using T1-weighted (A), proton-density-weighted, fat-saturated (B), and postcontrast subtracted T1-weighted (C) sequences demonstrates a large, irregular epiphyseal tumor of the distal ulna. The tumor stroma shows T1 hypointense, proton-density-fat-saturated hyperintense signal intensity, with internal cystic zones. The solid part shows avid contrast enhancement. Although signs of locally aggressive growth are evident, there are no signs of infiltration of surrounding tissues.

signs of extrasosseous tumor growth, with completely intact radius, carpal bones, blood vessels, nerves, and forearm muscles, the tendons of which were suppressed by tumor expansion.

Ultrasound examination and ultrasound-guided core needle biopsy were performed using a high-resolution (15 MHz) linear transducer. The ultrasound showed a relatively hypoechoic (compared with adjacent muscles) solid mass, with central cystic or necrotic portions (Figure 4). The color Doppler and spectral analysis findings included chaotically organized low-resistance arterial flow in the solid regions of the mass. Pre-biopsy shear wave elastography demonstrated areas of predominantly low to intermediate stiffness in the solid parts of the lesion (Figure 4). Thoracic CT findings were normal. Histopathological tissue analysis revealed numerous mononuclear cells and multinucleated osteoclast-like giant cells (Figure 5). Reactive woven bone spicules bordered by prominent osteoblasts were also seen.

Discussion

This paper presents a case of a young adult male patient with an extremely rare localization of GCTB in the distal ulna. Up to now, only a few cases of this localization have been recorded in the literature through case reports, and other papers on the subject do not mention this localization at all [6,7].

Until the diagnostic criteria were laid down in 1940 by Jaffe [3], the definition, classification, and prognosis of GCTB have been relatively vague. Previously, incorrect terms (sarcoma gigantocellulare, brown tumor) have often been used for this tumor due to its similarity in clinical, radiological, and histological presentation with other differential diagnostic entities of bone pathology. For this reason, exact diagnosis is crucial. In the first place, it includes different imaging modalities and pathohistological diagnosis, as demonstrated in the case of our patient.

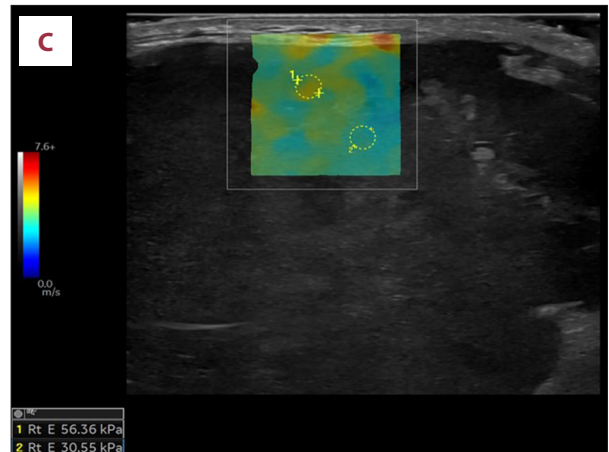
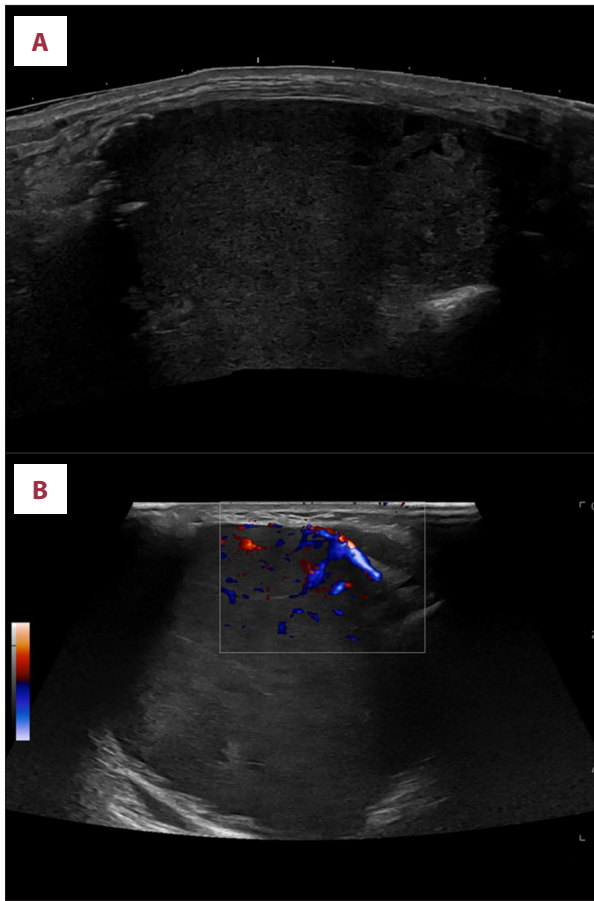


Figure 4. Images obtained during a core biopsy. Extended field-of-view B-mode image (A) shows an irregular hypoechoic mass in the distal ulna. Color Doppler flow imaging (B) demonstrates hypervascular tumor stroma with numerous sinusoidal vessels typical for giant cell tumor of bone. On shear-wave elastography (C), the color elastogram shows predominantly blue/cyan and cyan/green readings performed on 2 small areas of the superficial aspect of the tumor stroma (areas of low to intermediate stiffness, measuring 30-60 kPa).

Radiological imaging modalities, including radiography, ultrasound with Doppler and elastographic analysis, CT, and MRI often indicate the accurate diagnosis. They revealed an osteolytic, heterogeneous solid lesion centered in the metaphysis and extending to the diaphysis. Additionally, bone scintigraphy imaging is frequently used to evaluate these lesions for staging purposes and especially to evaluate lesions with unusual

locations. The hypervascular stroma of most GCTBs contains areas of necrosis and hemorrhage that may be associated with secondary aneurysmal bone cyst (ABC) [11].

The X-ray image of GCTB is very typical, regularly including osteolysis and the appearance of “inflated” bone with cortical thinning, which can be interrupted, but without a visible reaction of the periosteum, which differentiates it from the X-ray image of sarcoma. The tumor is usually sharply demarcated and without a sclerotic zone in the long bones; this is also typical of some radiologically similar processes, including osteoblastoma, chondroma, abscess, and non-ossifying

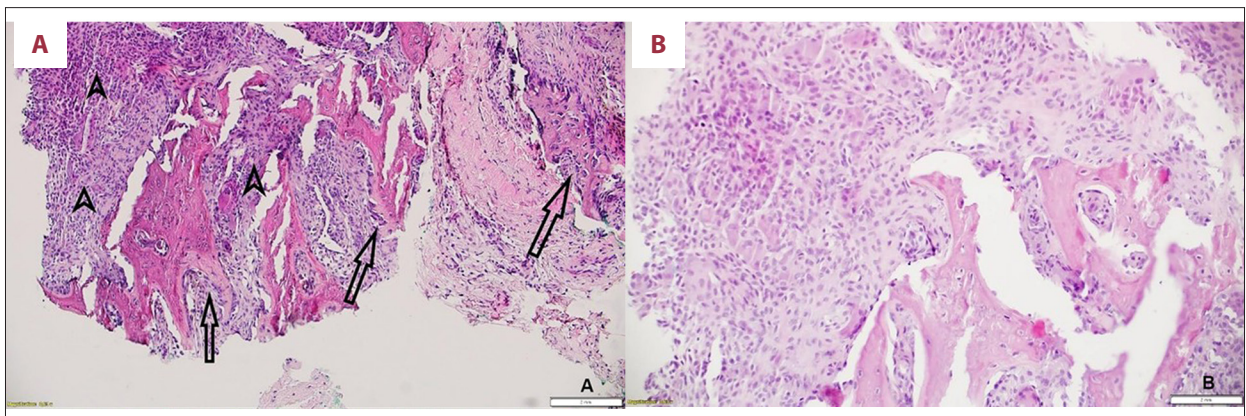


Figure 5. Hematoxylin-eosin-stained histological sections. (A) Magnification $\times 100$. (B) Magnification $\times 200$. Typical giant cell tumor of bone histology with multinuclear osteoclast-like giant cells (representing the neoplastic stromal cells; arrows) and mononuclear cells (arrowheads). Reactive woven bone spicules are apparent.

fibroma. GCTBs do not produce matrix-like osseous or cartilaginous neoplasms, and therefore mineralization within the lesion is absent. Tumor localization is usually asymmetric and eccentric compared with solitary bone cysts, which typically have central localization [11-13]. The tumor is almost always sharply demarcated and does not demonstrate infiltrative growth into the surrounding tissues (unlike sarcoma), which can be seen in our patient's example. These features, although not entirely pathognomonic, can be exactly and in detail differentiated by CT and MRI, which were indispensable in our case. The solid component of the tumor usually presents with low to intermediate signal intensity on T1-weighted sequence and increased signal intensity on images obtained with fluid-sensitive sequences (heterogeneously high signal intensity with areas of low signal intensity can be seen, due to fibrosis and hemosiderin deposits) [13,14]. Contrast enhancement of the solid part of the tumor is avid due to hypervascularity, but may not be uniform. Cystic zones enhance with a thin and delicate peripheral and septal pattern [11]. Due to inability of the ultrasound beam to penetrate the cortex, ultrasound is not commonly used in imaging of bone lesions [15]. In our case, however, lytic properties and superficial localization of the tumor allowed for excellent visualization of the tumor stroma. It was also feasible to perform a core biopsy under ultrasound guidance to avoid blood vessels displaced from their regular anatomical position by tumor expansion, and to collect a sample from a solid (rather than cystic or necrotic) region of the tumor. Elastography and Doppler analysis were also performed, demonstrating a hypervascular stroma with sinusoidal blood vessels and intermediate stiffness. While sinusoidal stromal vessels are a feature of GCTB [16], ultrasound findings are not sufficiently specific for a standard diagnostic procedure. Histopathological analysis enables an unequivocal diagnosis based on the pathognomonic finding of a typical cellular substrate in the form of numerous mononuclear cells and multinucleated osteoclast-like giant cells. The malignant potential of GCTB is currently assessed according to recurrence after surgical removal of the tumor. So far, neither radiological nor pathohistological criteria have been established that can unequivocally indicate malignant potential of GCTB. Our patient's thoracic CT findings did not show the spread of the disease in terms of lung metastases, which are rare, according to the literature. The patient is now included in the chemotherapy protocol (denosumab), with calcium and vitamin D supplementation, and will be regularly monitored for the timely detection of possible recurrence or the appearance of a new tumor at another location.

Despite modern diagnostic modalities, we want to highlight that same basic imaging principles have not changed for a long time: "In the majority of cases, an accurate radiographic diagnosis can be made by an experienced roentgenologist by the interpretation of adequate radiograms", as noted by Buschke and Cantril in 1949 [17].

Conclusions

In conclusion, we want to emphasize the key role of imaging methods in the diagnostic work-up of a lytic bone lesion. Because they are often encountered in clinical practice and include a lengthy list of differential diagnoses, a lytic bone lesion can be challenging even for experienced radiologists. Therefore, it is important to recognize distinguishing imaging characteristics of the lesion, even in the case of an unusual localization. In this report, the appearance of a single, sharply defined osteolytic lesion, the absence of sclerotic margins, and eccentric localization at the long bone epiphysis abutting the articular surface in an adult patient with closed epiphyses were the key features suggesting GCTB, although the ulna was affected. In addition, the localization of the tumor allowed for an ultrasound-guided core needle biopsy and histopathological verification. Timely diagnosis and adequate surgical treatment are important for long-term survival and minimizing postoperative patient disability.

Department and Institution Where Work Was Done

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Conflict of Interest

None.

Declaration of Figures Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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