

Telangiectatic Osteosarcoma Arising in a Treated Unicameral Bone Cyst of the Proximal Humerus: A Case Report

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Learning Point of the Article:

Malignant transformation of a unicameral bone cyst is exceedingly rare, but new or recurrent symptoms after treatment should prompt immediate re-evaluation.

Abstract

Introduction: Unicameral bone cysts (UBCs) are benign, fluid-filled lesions most commonly affecting the proximal humerus in children. Malignant transformation of a histologically confirmed UBC is extraordinarily rare, with only isolated cases reported in the literature.

Case Report: A 12-year-old girl developed high-grade telangiectatic osteosarcoma of the left proximal humerus 18 months after curettage and injection of a bone substitute for a biopsy-proven UBC. She presented with new-onset shoulder pain, and further imaging and biopsy confirmed telangiectatic osteosarcoma. She underwent neoadjuvant chemotherapy followed by wide resection and alloprosthetic hemiarthroplasty reconstruction.

Conclusion: This rare case highlights that new or recurrent symptoms after treatment of benign bone tumors warrant immediate re-evaluation, as malignant transformation, though rare, can occur.

Keywords: Osteosarcoma, unicameral bone cyst, pediatric bone tumor, proximal humerus, malignant transformation.

Introduction

Unicameral bone cysts (UBCs), also known as simple or solitary bone cysts, are benign fluid-filled lesions that slowly expand, leading to progressive thinning of the surrounding bone [1]. They typically arise in the metaphyseal region of long bones with open physes, most often in the proximal humerus, an important growth center in children [1,2]. Over 85% of UBCs occur in children aged 3–14, with a twofold male predominance [1]. The pathophysiology of UBCs remains unclear, though proposed mechanisms include impaired venous drainage, intramedullary hemorrhage, and abnormal bone remodeling [1,3]. Most UBCs are asymptomatic and found incidentally; symptoms usually

appear after a pathologic fracture with localized pain and deformity [1,4]. On gross examination, UBCs present as a large intramedullary cavity filled with straw-colored fluid and lined by a thin, reddish-brown membranous capsule [1,5]. Histologically, they are composed of fibrovascular stroma with focal osteoclast-type multinucleated giant cells [5]. Radiographs typically reveal a central, radiolucent lesion, with the pathognomonic “rising bubble sign” aiding in diagnosis [2]. The differential diagnosis includes aneurysmal bone cyst (ABC), which characteristically demonstrates internal septations and fluid-fluid levels [6]. Management ranges from observation of asymptomatic lesions to surgery for symptomatic or fracture-prone lesions. The

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Author's Photo Gallery



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prognosis is generally favorable, with reported recurrence-free survival approaching 88% [7]. There are anecdotal reports of UBCs undergoing malignant transformation. Isolated case reports, describing osteosarcoma developing at a prior UBC site, have raised questions about the malignant potential of UBCs [8]. We present a rare case of a 12-year-old girl previously treated for a proximal humeral UBC who, 18 months later, developed new-onset shoulder pain, and subsequent imaging and biopsy revealed a high-grade telangiectatic osteosarcoma. The patient underwent neoadjuvant chemotherapy followed by wide resection and reconstruction with an allograft-prosthetic composite hemiarthroplasty.

Case Report

A 12-year-old girl with a known history of a UBC in the left proximal humerus presented with worsening shoulder pain. Eighteen months earlier, she had undergone open biopsy, curettage, and calcium phosphate bone grafting at an outside facility for a benign-appearing lesion (Fig. 1). The initial radiographs demonstrated a radiolucent diaphyseal lesion with cortical thinning and a pathologic fracture (Fig. 1a and b). Although certain features are atypical for a classic UBC, including diaphyseal location, asymmetry, indistinct margins, and periosteal reaction, these are attributable to the acute

pathologic fracture rather than an inherently aggressive process. Specifically, the apparent gravity-defying position of the fallen cortical fragment on the anteroposterior view reflects displacement into the surrounding fracture hematoma, a recognized phenomenon in acutely fractured UBCs and not a feature of malignancy. The periosteal reaction visible on the post-treatment radiographs (Fig. 1c and d) reflects the expected healing response following curettage and bone grafting. Histopathology at that time confirmed a UBC, with the outside surgical pathology report documenting scant lymphohistiocytic cells and eosinophils in a hemorrhagic background with no cytologic atypia, mitotic activity, or osteoid production. Although the original histologic slides are unavailable for independent review, as the procedure was performed at an outside institution, this report provides adequate documentation of a benign bone cyst diagnosis. Her post-operative recovery was uneventful. She was symptom-free for 12 months, and then developed progressive shoulder pain without antecedent trauma, constitutional symptoms, or weight loss, leading to re-evaluation 6 months later. There were no other palpable masses or musculoskeletal findings on physical examination. Given her history and new symptoms, repeat imaging was obtained. Radiographs demonstrated a new, aggressive-appearing lytic lesion involving the proximal left humerus, slightly proximal but in continuity with the site of the

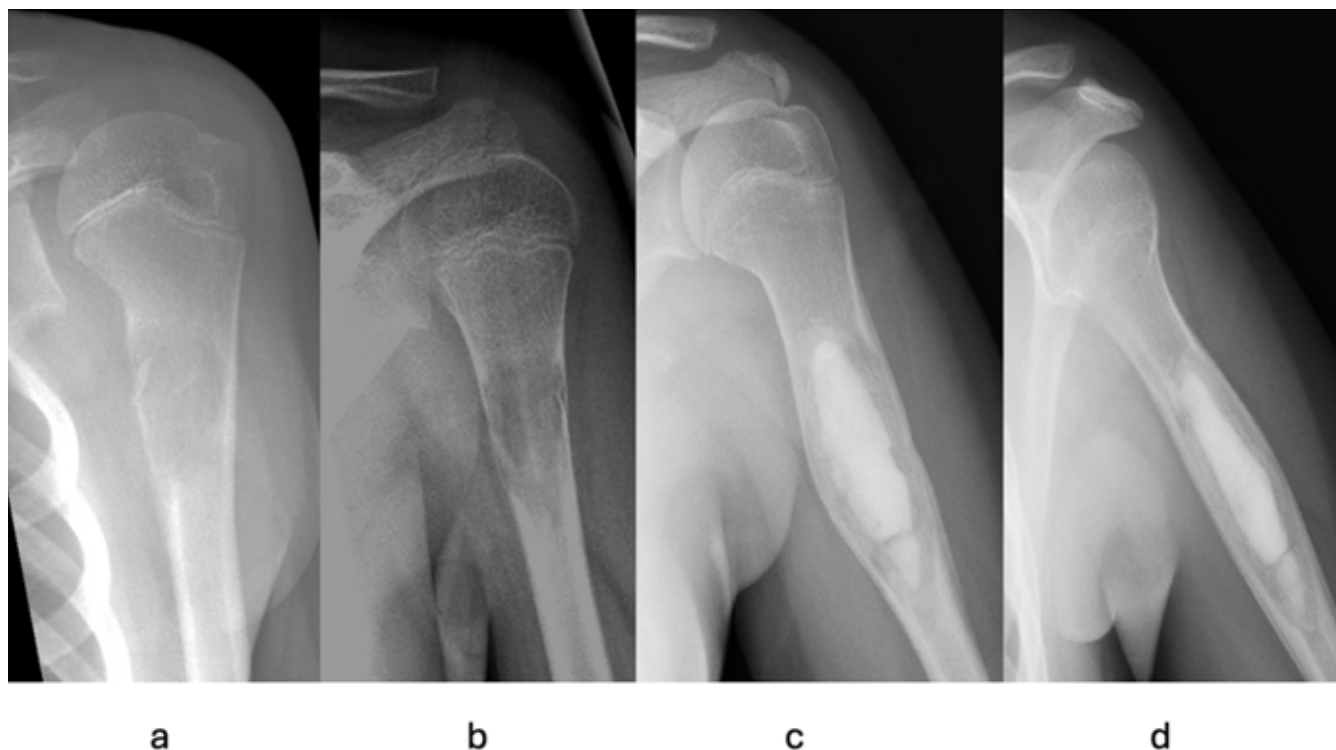


Figure 1: Initial unicameral bone cyst. Radiographic images of the left proximal humerus at initial presentation showing the unicameral bone cyst. Anteroposterior (AP) (a) and lateral (L) (b) radiographs demonstrating a radiolucent lesion in the left proximal humeral diaphysis with a pathologic fracture; AP (c) and L (d) post-treatment radiographs showing the lesion after curettage and calcium phosphate bone grafting, with radiopaque graft material filling the cavity.

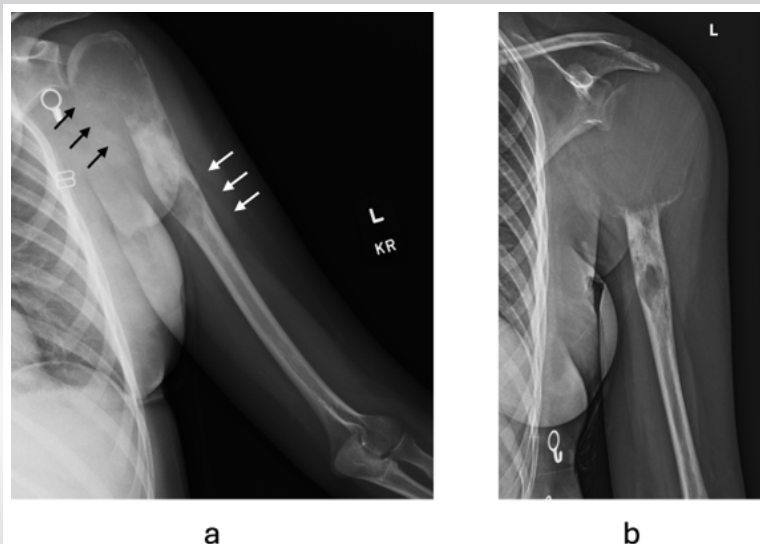


Figure 2: New aggressive lesion at 18 months. Radiographs obtained 18 months after initial treatment demonstrated malignant transformation. Anteroposterior (a) radiograph showing destructive cortical changes in the proximal medial humeral head and neck (black arrows) and ill-defined radiolucency in the distal aspect of the previously treated “unicameral bone cyst” (white arrows); (b) Lateral view demonstrating the expansile nature of the lesion with the associated soft-tissue component, located at the site of the previously treated uncameral bone cyst.

previously treated UBC (Fig. 2). A staging chest computed tomography (CT) showed no evidence of metastatic disease. Magnetic resonance imaging of the entire humerus revealed a heterogeneous, expansile lesion centered in the proximal humeral metaphysis, with internal fluid-fluid levels and demonstrated heterogeneous post-contrast enhancement (Fig. 3a-e). The patient subsequently underwent a CT-guided biopsy, which revealed a high-grade tumor with focal osteoid and hemorrhagic and cystic features interpreted as a telangiectatic osteosarcoma. Low-power microscopy of the biopsy demonstrated a hemorrhagic cystic mass with solid tumor components at the cyst wall (Fig. 4a). High-power examination revealed malignant pleomorphic cells with enlarged hyperchromatic nuclei, prominent nucleoli, and filigree neoplastic osteoid adjacent to pools of hemorrhage, diagnostic of telangiectatic osteosarcoma (Fig. 4b). The tumor was composed of cytologically malignant polyhedral cells with enlarged hyperchromatic nuclei and eosinophilic cytoplasm. Multiple mitotic figures were observed, along with areas of necrosis and hemorrhagic cystic change.

Immunohistochemistry showed that the tumor cells exhibited positivity for SATB-2 and the expression of INI-1 was retained. The p53 stain exhibited a mutant pattern with loss of expression. The patient was started on neoadjuvant chemotherapy consisting of adriamycin, ifosfamide, and methotrexate. After completion of the neoadjuvant regimen, she underwent wide resection of the proximal humerus through an extended deltopectoral approach. The defect was reconstructed with an allograft-prosthetic composite hemiarthroplasty with a Pyrocarbon humeral head (Fig. 5). She was discharged home the following day with no complications. The resected specimen measured 6.5 cm (Fig. 3b), and on microscopic review, 100% necrosis was observed with widely negative margins (Fig. 3c, d, e). At her 18-month follow-up, the wound was well healed. Although active shoulder range of motion remained limited, she was able to passively abduct and forward flex to 90° and could place her hand over her mouth and on top of her head. Her Musculoskeletal Tumor Society functional score was 25/30. There was no clinical evidence of disease recurrence, and the reconstruction remained stable.

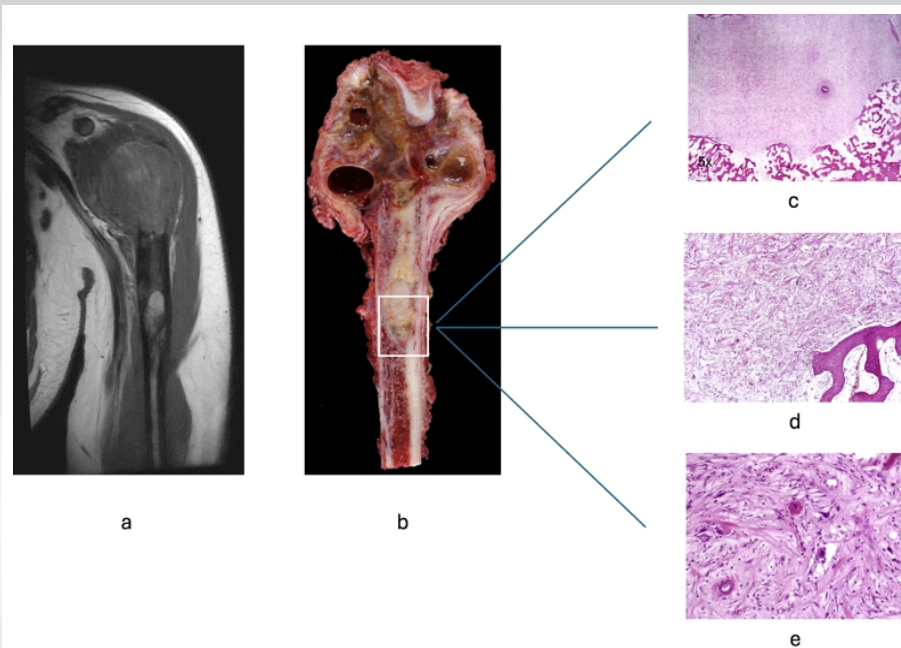


Figure 3: Magnetic resonance imaging (MRI), gross pathology, and histopathology. (a) Coronal T1-weighted post-contrast MRI showing a heterogeneous, expansile lesion in the proximal humeral metaphysis with multiple fluid-fluid levels and heterogeneous enhancement. (b) Gross specimen of the resected proximal humerus demonstrating the hemorrhagic and cystic nature of the tumor with multiple blood-filled cavities characteristic of telangiectatic osteosarcoma. The white box indicates the area of tissue for histologic interpretation; (c) Low-power photomicrograph $\times 5$ (Hematoxylin and Eosin [H&E] stain) of the post-treatment resection specimen demonstrating a nodule of tumor surrounded by cancellous bone; (d) Medium-power photomicrograph $\times 10$ (H&E stain) showing mostly collagen and a significant treatment effect; (e) High-power photomicrograph $\times 40$ (H&E stain) demonstrating 100% treatment effect with mostly collagen and rare residual tumor cells.

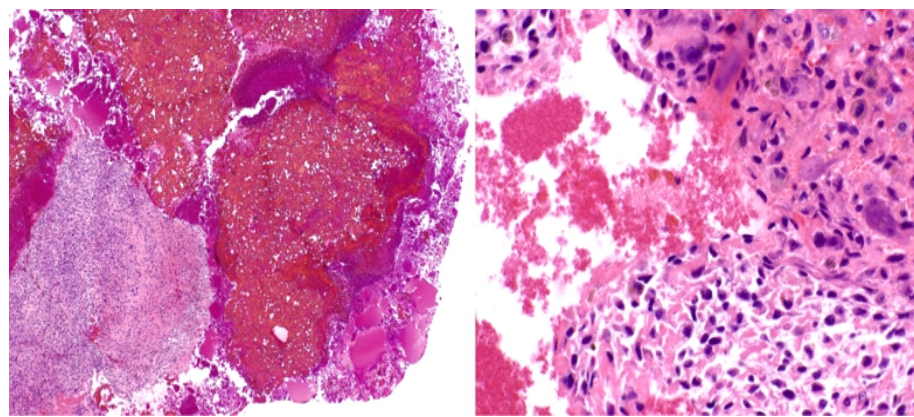


Figure 4: Pre-treatment biopsy histopathology. (a) Low-power photomicrograph (Hematoxylin and Eosin [H&E] stain) showing a hemorrhagic cystic mass with solid components of the cyst wall composed of tumor; (b) High-power photomicrograph (H&E stain) showing malignant cells associated with filigree neoplastic bone adjacent to a pool of blood, diagnostic of telangiectatic osteosarcoma.

Discussion

This case represents a rare instance of a high-grade telangiectatic osteosarcoma arising at the site of a previously treated UBC. Malignant transformation of a histologically confirmed UBC without prior exposure to radiation therapy is very uncommon, and only one similar case has been documented in the literature [8]. In that report, a patient developed osteosarcoma 22 months after undergoing curetting and grafting for a proximal femoral lesion initially diagnosed as a UBC [8]. Only one other case in the literature has described malignant transformation of a UBC, where an undifferentiated high-grade pleomorphic sarcoma developed at the same location [9]. While the exact mechanism behind this rare transformation remains unclear, several possibilities exist. One theoretical possibility is that a primary osteosarcoma may have developed independently at the same site, possibly triggered by a genetic predisposition or de novo mutation. Although this patient had no identifiable risk factors, such a scenario cannot be excluded. The possibility of initial misdiagnosis must also be considered. UBCs and ABCs can share overlapping radiographic and histologic features, and misdiagnosis is possible, particularly in limited biopsy samples [6,10]. It is conceivable that the original lesion was an ABC or early telangiectatic osteosarcoma presenting with UBC-like characteristics. However, pathology review confirmed a benign UBC without malignant or aggressive findings. Kyriakos and Hardy's comprehensive analysis of malignant transformation in ABCs highlighted the challenge of distinguishing true malignant transformation from initial misdiagnosis, particularly in cases of telangiectatic osteosarcoma [10].

In the present case, the original outside pathology report documented a benign bone cyst without cytologic atypia, mitotic activity, or osteoid production, confirming a benign diagnosis. The 12-month symptom-free interval following surgery, combined with the absence of malignant histologic features on initial biopsy, further supports true malignant transformation rather than misdiagnosis. Taken together, the most plausible explanation is true malignant transformation of a histologically confirmed UBC. While the biological mechanisms underlying such transformation are poorly understood, this case adds important evidence to the

limited literature on this phenomenon. Although rare, this occurrence highlights the importance of maintaining vigilance during follow-up of patients treated for presumed benign bone lesions.

Conclusion

This case highlights the importance of maintaining vigilance

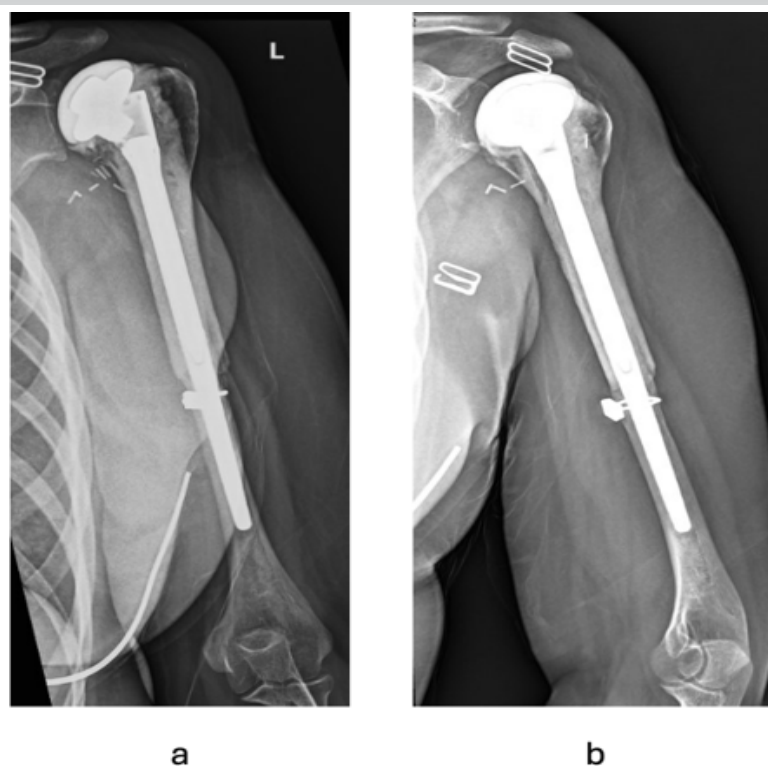


Figure 5: Post-reconstruction follow-up radiographs. Post-operative radiographs showing the allograft-prosthetic composite reconstruction. (a) Anteroposterior view demonstrating the pyrocarbon humeral head hemiarthroplasty with proximal humeral allograft, secured with cerclage wires and a long humeral stem. (b) Lateral view showing stable reconstruction with well-positioned components and maintained alignment.

following treatment of benign bone tumors in pediatric patients. Although malignant transformation of UBCs is extraordinarily rare, new or recurrent symptoms should prompt immediate imaging and tissue sampling to facilitate early detection and optimal treatment outcomes. Further research into the mechanisms of malignant transformation in benign bone lesions is warranted to better understand this rare but significant complication.

Clinical Message

Malignant transformation of a UBC, though extraordinarily rare, can occur without prior radiation exposure. New or recurrent symptoms following treatment of benign bone lesions in pediatric patients should prompt immediate imaging and tissue sampling, as early detection is critical for optimal outcomes.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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