CASE REPORT

Unicameral Bone Cyst With Atypical Location In Distal Femur: A Case Report

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ABSTRACT

Unicameral bone cysts (UBCs) are benign fluid-containing cavities commonly found in the proximal humerus and femur. However, their rare occurrence in atypical location like the distal femur requires further evaluation. We present the case of a six-year-old boy with pathological fracture of the distal femur following a trivial injury. Radiographic imaging revealed a centrally located lytic lesion with 'fallen fragment' sign, otherwise no mass seen within the cyst. Atypical location raised concerns about other possible differentials such as aneurysmal bone cyst, eosinophilic granuloma, fibrous dysplasia or telangiectatic osteosarcoma. We utilized intraoperative frozen section to rule out malignancy before proceeding with surgical curettage, bone grafting and plate stabilization. The final histopathological result confirmed the UBC. Imaging during follow-up showed evidence of cyst and fracture healing. At one-year, the patient had achieved full function with no cyst recurrence. Further evaluation would be necessary to confirm the diagnosis of UBCs if found in atypical locations. Intraoperative frozen section is valuable diagnostic and therapeutic tool.
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INTRODUCTION

Unicameral bone cysts (UBCs) are benign intramedullary lytic lesion characterized by fluid-containing cavities that can lead to cortical thinning and, in some cases, pathological fractures as they progress (1,2). They primarily affect metaphysis of long bones in children and adolescent aged between three and 14 years (1,2). Although the proximal humerus and proximal femur are typical locations of UBCs in 80-90% cases, they also can be found in less common sites such as proximal tibia, distal humerus, calcaneum, pelvic, and spine (1).

To the best of our knowledge, there is limited literature reporting on these rare locations of UBCs and their managements (3). The presence of UBCs at atypical locations poses diagnostic and management challenges, potentially leading to misdiagnosis, delayed treatment, or inappropriate interventions. Recognizing the possibility of UBCs in these locations is crucial for timely and effective management. Here, we report a case of UBC that showed typical radiological characteristics, including the 'fallen fragments' sign, but occurred at atypical site, specifically the distal femur. We emphasize on intraoperative frozen section as part of diagnostic approach, surgical management, and outcomes of this atypical presentation of UBCs.

Parental informed consent was obtained for the publication of the medical case details and any accompanying images.

CASE REPORTS

A previously healthy six-year-old Malay boy presented with painful swelling and deformity in left thigh following a trivial fall while playing at the playground. He was

initially presented at a referring hospital for treatment in July 2023. On further questioning, he denied previous history of left thigh pain or any constitutional symptoms. Also, there was no history of fever or infective symptoms. Physical examination revealed a swollen and deformed left thigh, with no open wounds or signs of infection. Neurovascular status was intact. Blood investigation showed raised level of alkaline phosphatase (ALP) and erythrocyte sedimentation rate (ESR), while other infective parameters were within normal limits. Plain radiographs revealed a comminuted distal third left femur fracture through a centric lytic lesion, which was located approximately 3 cm proximal to the distal femoral physis. There was a narrow zone of transition over its margins. Furthermore, the presence of fractured fragment within the medullary canal of distal femur was a typical characteristic of fallen fragment sign. Otherwise, there was no evident periosteal reaction (Fig. 1a,b).

Fig. 1: (a,b) Anteroposterior and lateral radiographs of the left femur showing a comminuted distal third femur fracture through a centrally located lytic lesion, approximately 3cm proximal to distal femoral physis. The lesion exhibits a narrow zone of transition with visible fallen fragments. (c) Sagittal CT scan highlighting the characteristic "fallen fragment" sign within the cyst. (d) MRI T2-weighted image (T2W) of coronal view demonstrating a relatively well-defined lesion with a hyperintense signal at the fracture site. (e) Gadolinium-enhanced MRI (Gado), shown in sagittal view, confirming peripheral rim enhancement.

Due to the trivial injury and location of lytic lesion, the patient underwent advanced imaging at the referring hospital within two weeks of admission. A contrast-enhanced computed tomography (CECT) scan and a Magnetic Resonance Imaging (MRI) of the left femur were performed to better evaluate the lesion. The CT scan confirmed the presence of a 'fallen fragment' sign within a cyst containing hematoma, and no evidence of a mass observed (Fig. 1c). Additionally, the Magnetic Resonance Imaging (MRI) revealed a relatively well-defined lesion with hyperintense signal on T2-weighted imaging, rim enhancement on post-Gadolinium imaging, and the pathognomonic 'fallen fragment' appearance. There was also minimal cortical thinning and patchy blooming artifact at the periphery, which was suggestive of hemosiderin deposition. High signal intensity on STIR sequences in the surrounding muscles was in keeping with oedematous changes. Otherwise, there was no mass or fluid-fluid level was reported (Fig. 1d,e). Following the imaging workup, the patient was transferred to our tertiary centre for further evaluation and management.

Although both clinical and radiological findings point towards the diagnosis of UBC as the likely diagnosis, however, given the atypical location of the suspected UBC, we decided for intraoperative frozen section for histological confirmation. Surgery was carried out within three weeks after the injury. The distal femur was approached surgically from a standard lateral approach till the fracture site was exposed (Fig. 2a). Intraoperatively, a blood-filled cyst was identified, with no evidence of pus collection or slough tissue observed. Fresh sample of the blood-filled cyst and its lining were obtained from the fracture site and submitted for frozen sections analysis. A total of ten section samples were reviewed. They predominantly showed osteoblastic ring with callus tissue, along with proliferation of blood vessels and fibroblast, indicating fracture healing. Otherwise, there was no definite cyst lining or malignancy cells seen in all sections. Following that, a definitive intralesional bone curettage was performed. The fracture was stabilized using a medial distal tibia locking plate, which was pre-bent to accommodate the contour of distal femur and avoid the distal femoral physis (Fig. 2b). The choice of implant was made based on the relatively small size of the distal femur. The void area was then packed with cancellous allograft. Additionally, tissue samples were sent for cultures to rule out infection.

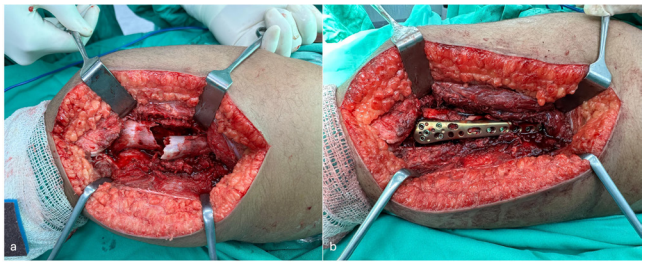

Fig. 2: (a) Intraoperative appearance of fracture over the distal femur. (b) Plate stabilization using a medial distal tibia locking plate.

The final histopathological analysis (Fig. 3a-d) demonstrated fibrocollagenous tissue having scattered multinucleated giant cells supporting a diagnosis of UBC. Tissue cultures showed no evidence of infection. Postoperatively, his recovery was uneventful. By eight weeks, he began partial weight-bearing, progressing to full weight-bearing with full knee motion by three months. At the nine-month follow-up, the fracture had completely united and good remodelling observed and showed no signs of local recurrence (Fig. 4a,b). By one-

year, there were no new lesions or evidence of cyst recurrence and he had regained full function.

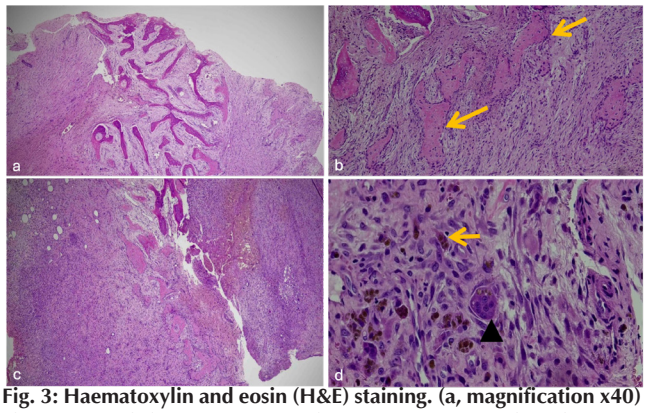

Fig. 3: Haematoxylin and eosin (H&E) staining. (a, magnification x40) The section exhibits fragments of fibrous tissue composed of admixture of mature bony fragments with new woven bone embedded within a fibroblastic stroma. (b, magnification x200) Evidence of fractured callus is indicated by the presence of osteoid rimmed by reactive osteoblasts (orange arrows). (c, magnification x40; d, magnification x400) Evidence of both new and old haemorrhages is observed, characterized by the presence of hemosiderin-laden macrophages. (d, magnification x400) Hemosiderin-laden macrophages (orange arrow) and scattered multinucleated giant cells (black arrowhead) are also identified within the fibrocollagenous tissue.

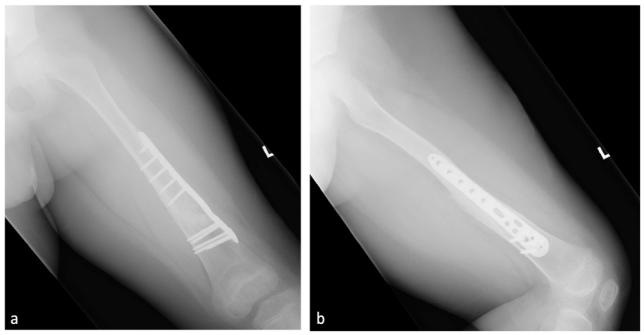

Fig. 4: (a,b) Plain radiographs taken nine months post-surgery show complete union and good remodelling with no signs of local recurrence.

DISCUSSION

Unicameral bone cysts are benign tumour-like lesion primarily affecting children and adolescent in about 85% of cases (2). While these cysts can occur in any bones, 80-90% are exclusively found in the proximal humerus and proximal femur in individual under the age 17 years (1,2). When they involve in less common or rare locations such as pelvis, ilium, calcaneum or talus, they tend to manifest in older individuals (2). To our knowledge, there are limited cases of UBCs in the distal femur (1-3). Noordin et al. described the typical characteristics of UBC in the distal femur on plain radiograph and MRI in eight-year-old child who had a pathological fracture (2).

Clinical behaviour of UBCs varies based on the distance to physis in the long bone. They are considered 'active growing' if located within 5-10mm from the epiphyseal growth plate and become 'latent' upon migrating to diaphysis with skeletal growth (2,3). While the majority of UBC cases are asymptomatic, some patients may present with significant symptoms due to pathological fractures (2). Typically, a plain radiograph is adequate for diagnosing UBCs with typical characteristics of centrally located lytic lesion with a thin sclerotic margin, and the pathognomonic 'fallen fragment' sign when fracture has occurred. The use of CT or MRI scans are rarely indicated unless in cases of atypical presentation or when plain radiographs reveal insufficient information (2). While the common sites of UBC are well established, diagnosing UBCs in atypical locations poses a diagnostic challenge. In our patient, clinical and radiological findings suggest UBC, specifically a pathognomonic "fallen fragment" sign. However, the atypical location prompted further investigation to exclude other differentials, primarily aneurysmal bone cyst, followed by monostotic fibrous dysplasia, eosinophilic granuloma, intraosseous ganglia (1,2) and, importantly, malignant tumour like telangiectatic osteosarcoma. MRI was performed in our patient to evaluate the presence of any mass within cyst. However, it only revealed fluid and haematoma from the fracture, without any differentiating features to exclude other differentials. While primary bone tumours are a major consideration in such cases, the possibility of infection must also be carefully evaluated. Therefore, intraoperative tissue samples were sent for microbiological analysis to rule out infection, and all cultures returned negative.

The goal of managing UBCs involve fracture prevention or management of pathological fracture, achieving the cyst healing and minimizing the recurrence or refracture (2). There are various treatment options in UBCs but the optimal one remains controversial. These include percutaneous aspiration, steroid injection, surgical decompression, curettage, bone grafting and fracture stabilization (1). Surgical options are considered for cyst in high-risk fracture areas like proximal femur, expanding cysts and those with pathological fractures (2). The surgical curettage for UBCs offers healing rates of up to 90%, with no significant difference between the use of autograft or allograft (3). Surgical treatment is necessary in our patient because of the fracture is adjacent to the knee joint and risk of malunion is high in non-operative treatment.

The dilemma in our management was on the decision to either perform local staging with a confirmatory biopsy before definitive surgery, which could prolong the immobilization period, or proceed directly to definitive surgery. Given the suspicions of other possible diagnoses in atypical location, we decided to use intraoperative frozen section to rule out malignancy. Once confirmed, we performed surgical curettage, bone grafting using cancellous allograft and fracture stabilization. Intraoperative frozen sections are useful tool for diagnosis and evaluation of clearance margin with an accuracy of 89 to 99% (4). However, freezing the hard cortical bones within a short interval can be

technically demanding (5). In our case, we obtained the fresh sample from the cyst wall lining and organized haematoma, thus making the frozen section procedure less demanding and easily performed. Ultimately, final histopathological result confirmed the UBC diagnosis. The patient is ambulating without pain and has a good range of left knee motion. The entire cyst has healed as evidence by the new bone formation and bridging callus observed at six-weeks post-operatively. At three-month follow-up, complete fracture union was achieved, and good fracture remodelling was seen at nine-month following the surgery. The patient regained full function with no evidence of cyst recurrence at one-year follow-up.

This case report has a few limitations. Firstly, the findings from this single case may not necessarily apply to a larger group of patients. Nevertheless, it may serve as a guide for clinicians when suspecting UBCs in atypical locations. Secondly, the follow-up was limited to a one-year period, which may not be adequate to completely assess the long-term risk of cyst recurrence. Longer follow-up for UBCs in atypical locations would provide a better understanding of their natural history.

CONCLUSION

Although UBC is primarily a clinic-radiological diagnosis, in cases with atypical presentations, using an intraoperative frozen section can be a valuable tool to confirm the diagnosis and guide the definitive treatment. This approach minimizes the risk of unexpected surgical complications from inappropriate diagnosis.

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