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Pediatric Distal Femoral Surface Tumor With Extraosseous Lesion Mimicking Parosteal Osteosarcoma: A Diagnostic Challenge

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Abstract

Background/Aim: Parosteal osteosarcoma is a low-grade surface osteosarcoma that most commonly arises from the posterior aspect of the distal femur. In pediatric patients, a surface-based sclerotic lesion accompanied by an extraosseous component may closely resemble parosteal osteosarcoma, creating a diagnostic dilemma.

Case Report: A six-year-old boy was referred with right knee discomfort and progressive difficulty in deep flexion. Imaging revealed a broad-based sclerotic surface lesion on the posterior distal femur with an extraosseous component, closely resembling previously reported pediatric parosteal osteosarcoma, although an open biopsy revealed no malignant features. A hemicortical excision without internal fixation was performed, resulting in a cortical defect involving <25% of the femoral circumference. Histopathological examination confirmed a sessile-type osteochondroma. Postoperatively, the patient achieved full weight-bearing and unrestricted knee motion. Radiological remodeling was observed at six months without fracture or recurrence.

Conclusion: Sessile-type osteochondroma of the pediatric distal femur can radiologically and clinically simulate low-grade parosteal osteosarcoma by presenting as a heavily ossified exophytic surface mass with an extraosseous component. Histopathological validation coupled with the absence of MDM2 amplification provides definitive differentiation between these entities, guiding appropriate bone-conserving hemicortical excision without the need for internal fixation.

Keywords: Parosteal osteosarcoma, sessile-type osteochondroma, distal femur, pediatric, extraosseous.

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Introduction

Parosteal osteosarcoma, a low-grade bone sarcoma that arises on the cortical surface of the bone, is the most common and slow-growing type of surface osteosarcoma with peak incidence in young adults. Accounting for approximately four percent of all osteosarcomas globally, this malignancy has a predilection for the posterior aspect of the distal femoral metadiaphysis in about 70% of cases, followed by the proximal tibia and proximal humerus as the next most frequent sites (1, 2). On imaging, it typically presents as a densely ossified surface mass and may mimic a benign lesion (2-4). Although uncommon in children, parosteal osteosarcoma should be considered in the differential diagnosis of pediatric surface-based sclerotic lesions around the distal femur. Thus, an accurate diagnosis is critical because mistaking parosteal osteosarcoma with benign tumor leads to unplanned excision, which usually results in local recurrence and increases the risk for dedifferentiation into high-grade malignancy. Such progression may change the therapeutic strategy from limb-salvage surgery to more radical procedures, including amputation, to achieve adequate oncologic margins (5).

This diagnostic pitfall is frequently caused by similarities in the clinical and radiological features shared between parosteal osteosarcoma and benign bone tumors, including overlapping location, indolent growth behavior, and a densely ossified cortical surface mass (6). The risk of recurrence following inadequate initial surgery is very high; for instance, large-scale clinicopathological data (2) demonstrates that patients undergoing initial intralesional or marginal excisions – often mistakenly performed for a presumed benign mass – consistently require subsequent operations for local recurrence. Another consequence is that the tumor mostly transforms into an aggressive high-grade malignancy, which commonly ends with amputation (5). To anticipate these risks, a multi-modal yet accurate diagnostic approach is mandatory, combining imaging with meticulous histopathological review and, where necessary, advanced immunohistochemistry.

When evaluating pediatric distal femoral surface lesions, osteochondroma – particularly the sessile type arising from the posterior aspect of the distal femoral metadiaphysis – should not automatically be assumed to be benign. When located at the same predilection site as parosteal osteosarcoma, a broad-based lesion may closely mimic malignancy on both clinical and radiological grounds, especially in patients with long-standing symptoms and progressive limitation of range of motion (4, 5, 7, 8). The diagnostic challenge is further amplified when imaging demonstrates a heavily ossified exophytic mass, a feature shared by both entities. As a result, a dependency on a single modality may lead to misdiagnosis and inappropriate treatment (4, 7). Therefore, a comprehensive yet stepwise diagnostic approach should be performed to ensure the favorable long-term survival typically associated with this well-differentiated tumor.

In this report, we present a pediatric case of a surface bone tumor in the posterior distal femoral metadiaphysis with an extraosseous lesion mimicking parosteal osteosarcoma, focusing on the diagnostic strategy based on imaging and pathological findings to establish the diagnosis. Accurate recognition of this diagnostic dilemma is necessary to differentiate malignant from benign lesions and to ensure appropriate surgical decision-making in skeletally immature patients.

Case Report

A six-year-old boy complained of discomfort in the right knee for one year, which was otherwise unnoticeable. The complaint worsened in two months after he experienced difficulty bending his right knee while doing Seiza – a traditional, formal Japanese sitting posture requiring deep knee flexion and ankle plantarflexion, with the buttocks resting on the heels. Physical examination was positive for bone mass in the popliteal region. No other abnormal findings were noted on physical examination. Laboratory findings (Table I) showed an increase in alkaline phosphatase (ALP) 181 IU/l and Lactate Dehydrogenase

Table I. *Laboratory parameters of the patient.*

Parameter	Value	
Hemoglobin (g/dl)	13.0	
Platelets ($\times 10^3$)	440	
WBC ($\times 10^3$)	5.64	
RBC ($\times 10^6$)	4.82	
CRP (mg/dl)	0.05	
Na (mEq/l)	139	
K (mEq/l)	4.4	
Cl (mEq/l)	106	
Calcium (mg/dl)	9.5	
Blood Urea Nitrogen (mg/dl)	13	
Creatinine (mg/dl)	0.27	
Albumin (g/dl)	4.5	
γ-GTP (IU/l)	11	
AST (IU/l)	28	
ALT (IU/l)	10	
Creatine kinase (IU/l)	78	
ALP (IU/l)	181	High
LDH (IU/l)	245	High

WBC: White blood cell; RBC: red blood cell; CRP: C-reactive protein; γ -GTP: gamma-glutamyl transpeptidase; AST: aspartate aminotransferase; ALT: alanine aminotransferase; ALP: alkaline phosphatase; LDH: lactate dehydrogenase.

(LDH) 245 IU/l. A series of imaging evaluations, including plain radiograph, computed tomography (CT) scan, bone scintigraphy, thallium scintigraphy, and magnetic resonance imaging (MRI) were performed and depicted in Figure 1.

Radiographs and CT demonstrated a broad-based, highly ossified surface lesion arising from the posterior distal femoral metadiaphysis with an associated extraosseous lesion, which closely resembled previously reported cases in terms of location, broad-based morphology, dense ossification, and corticomedullary continuity (4, 7). No pulmonary metastases were detected on chest CT. MRI demonstrated intraosseous and extraosseous components showing iso-intensity on T1-weighted images and slightly high intensity on T2-weighted images without any visible cartilage cap. Thallium scintigraphy showed mild uptake, and bone scintigraphy showed mild-to-moderate uptake in the lesion. Given these data, malignancy could not be excluded; therefore, an open biopsy was needed.

Open biopsy performed through a lateral approach revealed no malignant features. Because low-grade parosteal osteosarcoma was suspected, Murine Double Minute 2 (MDM2) amplification was evaluated using fluorescence in situ hybridization (FISH), which showed no evidence of MDM2 amplification (Figure 2). Due to limited deep flexion of the right knee, and at the request of the patient and the patient's family, surgical resection of the lesion was planned (Figure 3). The patient was placed in the left lateral decubitus position. The tumor, including the extraosseous component, was excised using a micro sagittal saw and chisel.

Histopathological examination of the resected specimen showed thickened bone trabeculae with focal transition to cartilage, without cytologic atypia or malignant features, consistent with osteochondroma. The extraosseous lesion consisted of a bony nodule with partial cartilaginous transition and likewise showed no evidence of malignancy (Figure 4). Postoperatively, the patient underwent rehabilitation program, beginning with early range-of-motion exercises and progressing to full weight-bearing in one week.

During postoperative follow-up, the patient was clinically satisfied because the range of motion of the right knee was improved and able to sit in Seiza position without any limitation or pain. Gradual bone remodeling was noted during the six-month postoperative period, with no evidence of fracture or recurrence (Figure 5).

Discussion

In the present case, differentiation between parosteal osteosarcoma and sessile-type osteochondroma was challenging because of the patient's age, lesion location, and surface-based morphology. Parosteal osteosarcoma is a low-grade surface osteosarcoma that most commonly arises from the posterior side of the distal femoral metadiaphysis and typically appears as a highly sclerotic lesion. This characteristic location and imaging appearance contributed to the diagnostic challenge in the present case. The tumor may closely

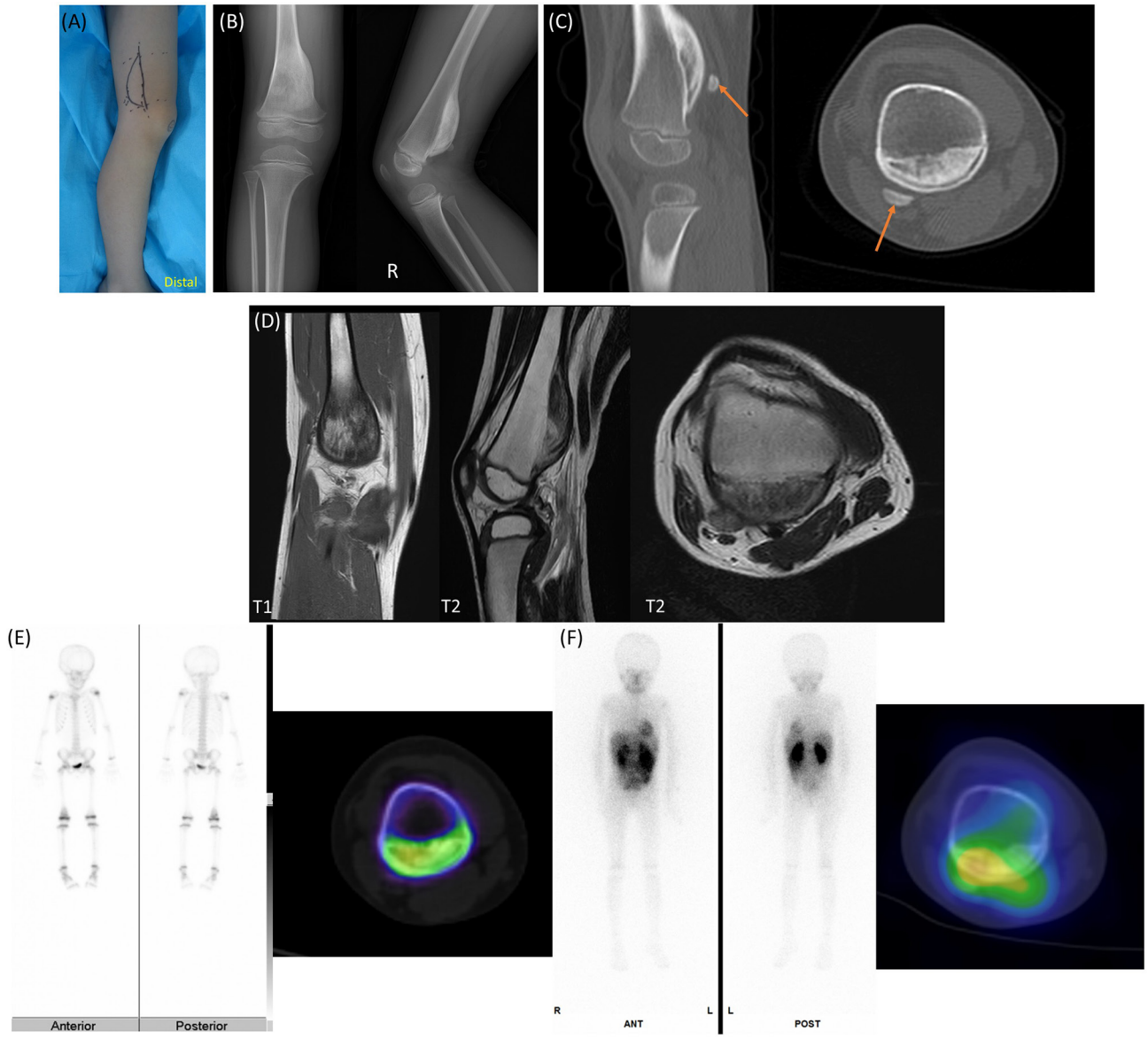

Figure 1. *Initial clinical and radiological presentation of the patient. (A) Clinical photograph showing a palpable mass at the posterior aspect of the distal femur. (B) Plain radiograph showed a protruding sclerotic, highly ossificated bone tumor located in the posterior side of distal femoral metadiaphysis. (C) Computed tomography (CT) scan of the affected area showed that the mass protruded outside from the surface of the bone with a pasted-on appearance, a clear border in the bone-tumor margin to some extent, and a clear border in the soft tissue accompanied by an extraosseous lesion marked with arrow. (D) Both the surface lesion and extraosseous lesions show iso-intensity on T1-weighted imaging, slightly high intensity on T2-weighted imaging without clear evidence of cartilage cap. (E) Bone scintigraphy showed mild to moderate uptake, whereas (F) thallium scintigraphy showed a mild uptake in the mass, with some uptake in the normal area of the distal femur.*

mimic a benign surface lesion in both appearance and clinical behavior. Although parosteal osteosarcoma is uncommon in pediatric patients, age alone does not reliably exclude malignancy in surface bone tumors. Sessile-type osteochondroma is usually asymptomatic and incidentally detected but may become symptomatic

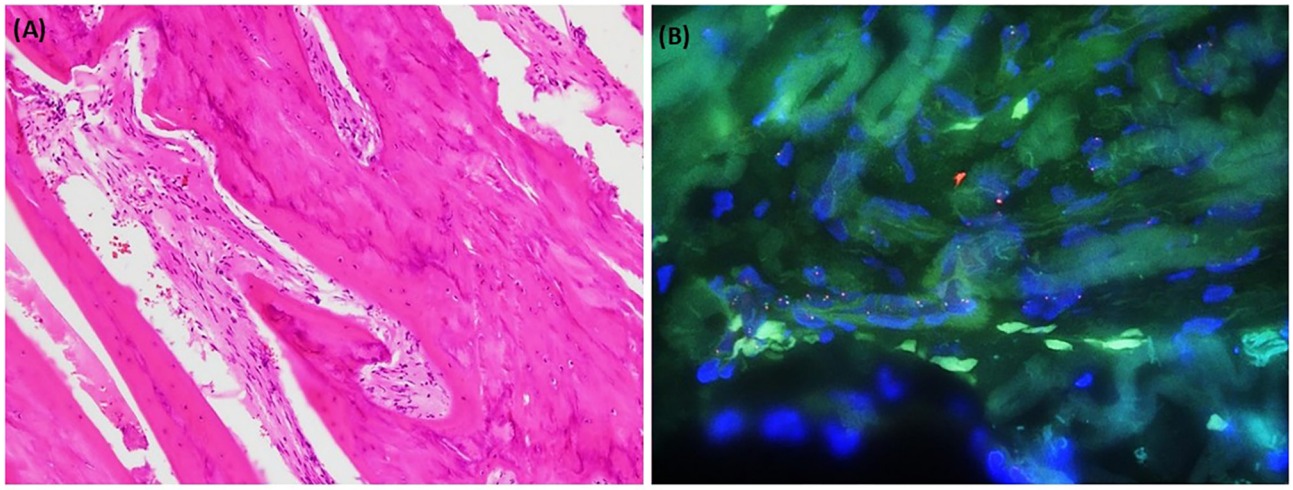

Figure 2. *Histopathological examination and immunohistochemistry results of the patient. (A) Open biopsy revealed a normal density of cells but increased bone density at the specimen surface, accompanied by a transition to cartilage without atypia. (B) Absence of MDM2 expression supported exclusion of parosteal osteosarcoma.*

when located near a joint or neurovascular structures (9).

Previous reports by Nodomi *et al.* (4) and Ishihara *et al.* (7) have described cases with very similar clinical and radiological features to those found in the present case. These cases (Table II) highlighted the potential diagnostic pitfalls associated with surface-based lesions of the distal femoral metadiaphysis in children and further justify the need for a careful diagnostic and surgical approach when malignancy cannot be confidently ruled out preoperatively.

The visualization of a peripheral cartilaginous component or "cartilage cap" in approximately 50% of parosteal osteosarcoma cases in imaging may become a significant diagnostic pitfall, as this can closely simulate the morphology of a sessile-type osteochondroma (1, 2, 8). Moreover, the thickness of the sessile-type osteochondroma cartilage cap in the pediatric population can be 1-3 cm and become thinner in the older population (2, 6). In our case, MRI failed to demonstrate a clearly defined cartilage cap, which is atypical for pediatric osteochondroma, which contributed to the diagnostic dilemma. Therefore, the cartilage cap alone is not diagnostically valuable when differentiating parosteal osteosarcoma and osteochondroma (5, 6).

Radioactive tracer imaging modalities such as bone scintigraphy and thallium scintigraphy provide essential data regarding the lesion's metabolic activity, yet they often present overlapping features that complicate the distinction between benign and malignant entities. The mild-to-moderate bone scintigraphy uptake observed in this six-year-old patient reflects active bone turnover, a finding common in both growing sessile osteochondromas and parosteal osteosarcoma (10). While bone scintigraphy is highly sensitive for detecting bone lesions in early stages, it is widely recognized as non-specific and unsuitable for definitively differentiating benign from malignant tumors. Similarly, the mild thallium scan uptake provides an equivocal signal; although thallium accumulation generally depends on cell viability, sodium-potassium pump activity, and blood flow – making it a useful tool for evaluating tumor grade and chemotherapy response (11) – it is susceptible to false-positive results in benign lesions characterized by hypervascularity or inflammatory changes.

For pediatric populations, interpretation is significantly complicated by the physiologically intense uptake in the physes, which represent normal growth centers. Furthermore, recent studies (12) emphasize that bone scintigraphy is a reliable method to evaluate

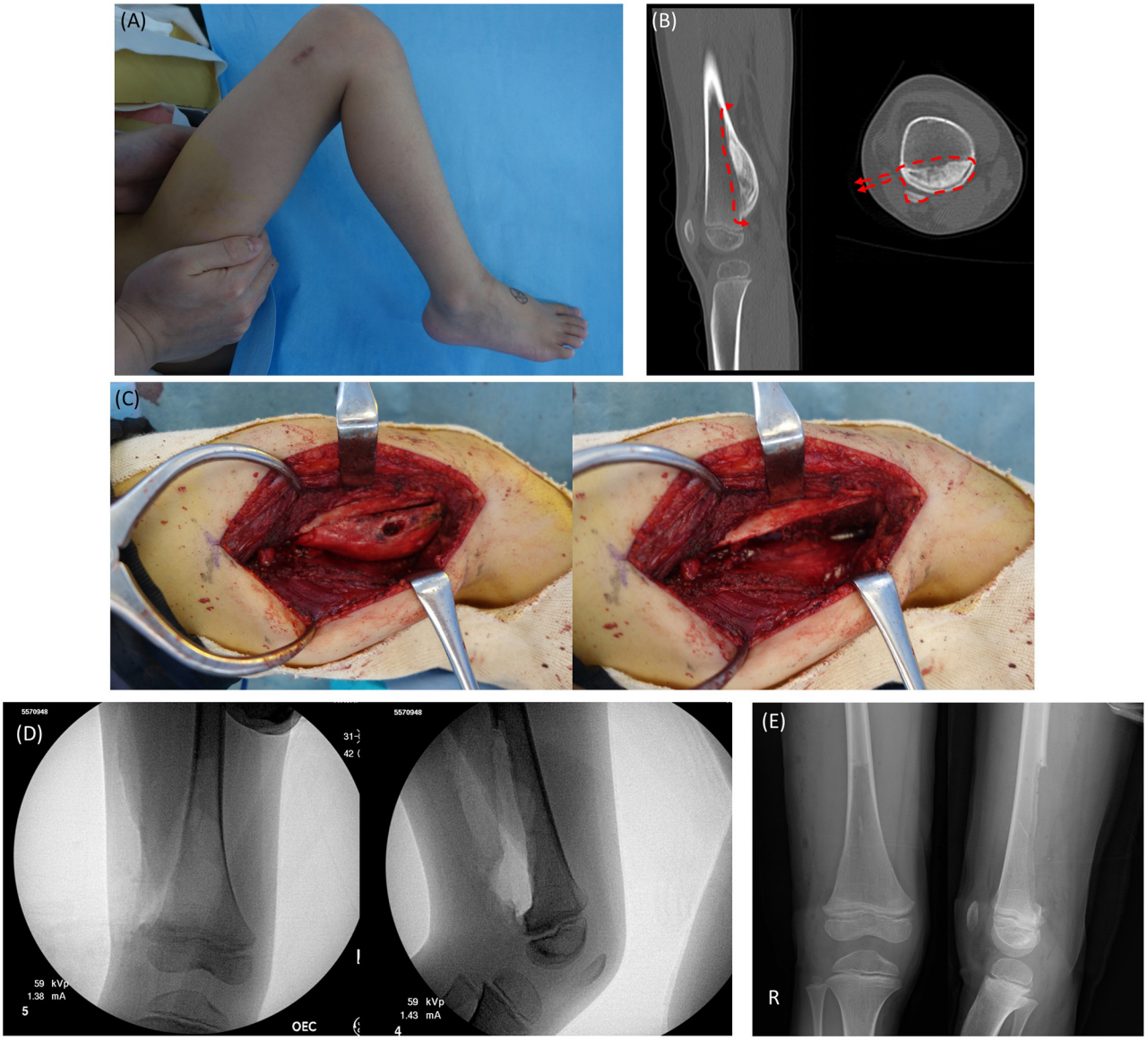

Figure 3. *Preoperative condition and intraoperative picture of the patient. (A) The range of motion of the right knee is limited. (B) Planned marginal excision. (C, D) Intraoperative condition pre- and post-tumor removal using image intensifier guided hemicortical osteotomy. (E) Postoperative plain radiograph showed a hemicortical lesion in the resected area without any bony reconstruction.*

the individual longitudinal growth potential of the epiphyses. While parosteal osteosarcoma typically manifests as a focal "hot spot", a benign osteochondroma in a skeletally immature patient may also show increased accumulation due to endochondral ossification during skeletal development (10). Thus, while bone and thallium scintigraphy show tumor metabolic activity, they cannot clearly tell if it is benign or malignant on their own.

Histologically, the cartilaginous cap of parosteal osteosarcoma lacks the organized, polarized columnar arrangement of chondrocytes seen in true osteochondroma, instead showing an irregular distribution of cells with

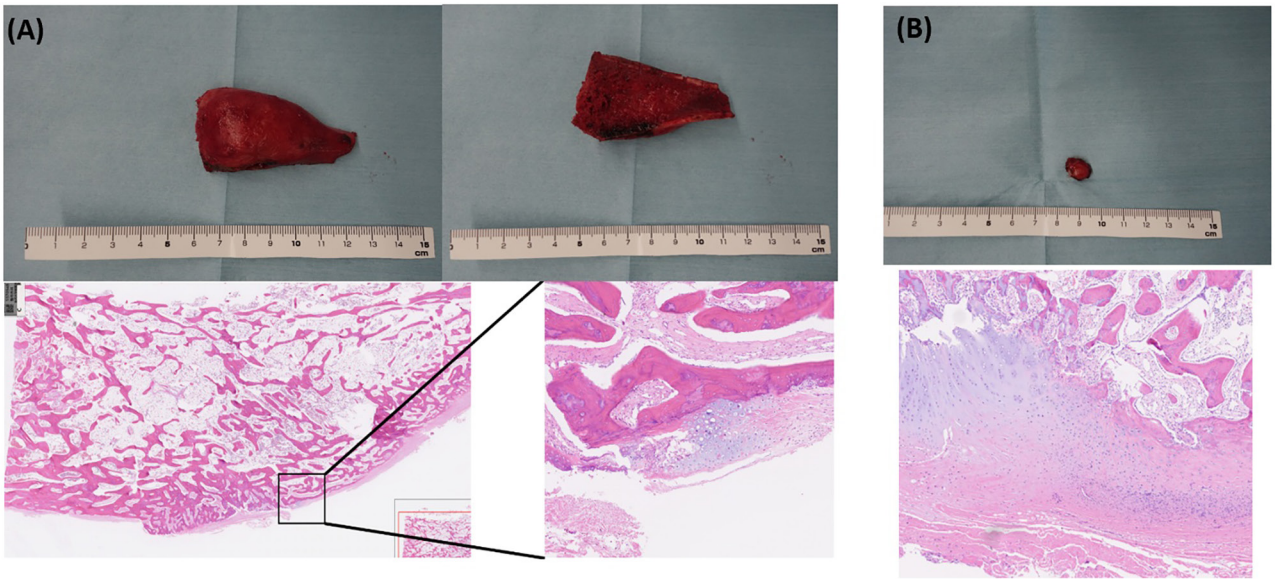

Figure 4. *Histopathological findings of the resected specimen. (A) Thickened bone trabeculae with focal cartilaginous transition are observed without cytologic atypia, consistent with osteochondroma. (B) The extraosseous lesion shows a benign bony nodule with partial cartilaginous transition and no evidence of malignancy.*

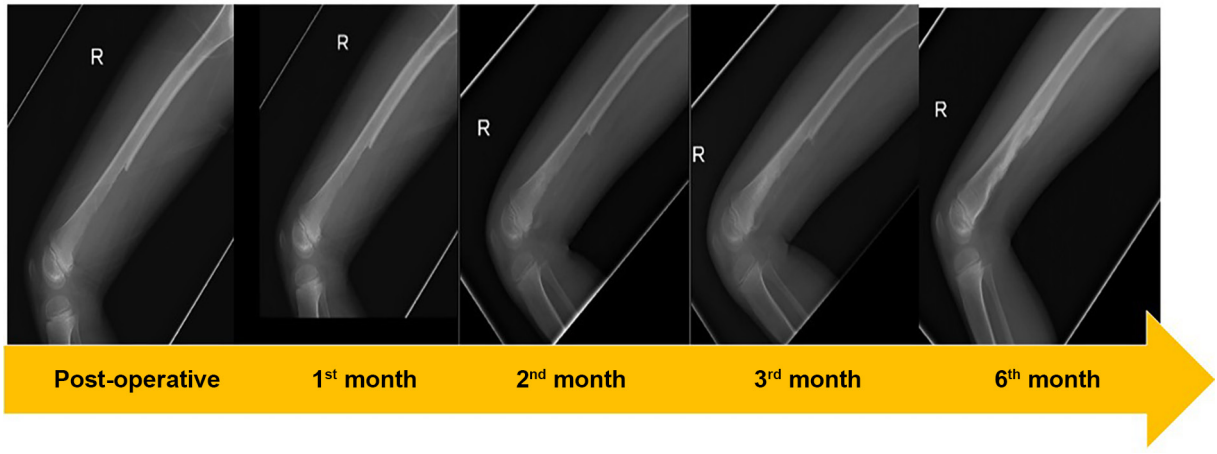

Figure 5. *A serial plain radiograph taken during six months showed that the hemicorticotomy defect is gradually remodelled despite no reconstruction and a full weight-bearing state of the patient.*

plump nuclei and occasional atypia. Furthermore, the underlying architecture of parosteal osteosarcoma consists of a diagnostic hypocellular spindle-cell stroma with minimal atypia and parallel-arranged bone trabeculae, a feature absent in benign osteochondroma (13). In our case, the diagnostic uncertainty became harder when histological examination showed a normal density of cells but increased bone density at the specimen surface, accompanied by a transition to cartilage cap without atypia. Given the diagnostic overlap between these two entities, immunohistochemical evaluation was performed in our case. The absence of MDM2 expression, in conjunction with clinical, radiological, and histological features, effectively excluded low-grade osteosarcoma (14, 15).

Table II. *Comparison of previous reports to present case.*

	Nodomi *et al.* (4)	Ishihara *et al.* (7)	Present case
Age (years)	9	12	6
Sex	Male	Male	Male
Duration of symptoms (months)	6	12	12
Pain	Present	No	No
Location	Posterior distal femoral metadiaphysis	Posterior distal femoral metadiaphysis	Posterior distal femoral metadiaphysis
Plain radiograph	A broad-based, irregular, highly ossificated mass. String sign (–), corticomedullary continuity (+)	A broad-based, regular, highly ossificated mass. String sign*(–), corticomedullary continuity (+)	A broad-based, regular, highly ossificated mass. String sign (–), corticomedullary continuity (+)
CT scan	Cortical lesion with irregular surface	Cortical lesion with homogenous calcification, smooth surface	Cortical lesion with homogenous calcification, smooth surface.
MRI	Medulla and soft tissue involvement was suspected, heterogenous enhancement	Focal enhancement on FDG-PET/MRI scan in deep bone marrow and in the stalk of polypoid lesion.	Unclear border in the bone-tumor margin, clear border in soft tissue
Extraosseous lesion	No	Yes	Yes
Cartilage cap in imaging	Not stated explicitly	Present	No
Bone scintigraphy/thallium scintigraphy	Not available	Not available	Mild-moderate uptake
Final histopathological result	Low-grade parosteal osteosarcoma	Low-grade parosteal osteosarcoma	Osteochondroma, without any atypical cells
Treatment	Limb-sparring surgery with marginal surgical margins, reconstruction using processed frozen bone, lung metastasectomy	Wide resection, vascularized fibula reconstruction	Excision, no reconstruction
Outcome (follow-up)	Alive with disease at 4.5 years	Disease-free at 2 years	No recurrence at 6 months

*String sign: radiolucent cleavage plane between tumor and cortex (9).

Although the cortical defect after marginal excision involved less than 25% of the axial plane, internal fixation was deemed unnecessary based on biomechanical stability and preservation of sufficient cortical bone. This decision is supported by the "50% rule" or Harrington criteria, which states that a critical loss of torsional integrity is typically observed only when a cortical defect approaches or exceeds 50% of the width of the femur (16-18). A study of fourth-generation composite femur also showed that a defect less than 33% cortical loss maintains a level of structural stability where the resistance to torsional load remains linearly related to the polar moment of inertia of the remaining bone stock (18). Despite these experimental defects were created on the lateral cortex rather than the posterior aspect, the underlying biomechanical principle remains applicable, as overall torsional resistance is primarily determined by the quantity and continuity of residual cortical bone rather than defect orientation alone.

Furthermore, recent *ex vivo* studies on human femora demonstrate that neither monocortical nor bicortical plating efficiently improves torsional stability in the presence of distal femoral defects (16). While plating may provide some stabilization against axial loading, it fails to restore maximum torque, angle, or torsional stiffness to a relevant extent because the bone continues to deform and fail at the defect site. In fact, the inclusion of plates can introduce additional risks, such as increased soft tissue damage, potential contamination of the contralateral cortex, and the development of stress risers at screw holes, which may actually facilitate fracture paths (16). Consequently, for a sub-critical defect of less than 25%, the intrinsic strength of the remaining bone is considered sufficient to withstand physiological torsional

loads without the need for invasive and potentially counterproductive metal augmentation.

During follow-up, the patient demonstrated a full recovery of daily living, including sitting in the Seiza position without pain or limitation, alongside a progressive bone consolidation enclosing the defect in six months. Despite the absence of internal fixation, no mechanical complications or pathological fractures were observed, which is consistent with the "50% rule". This approach also spared the patient from implant-related complications and the need for future hardware removal during growth. Moreover, there was no clinical or radiological evidence of tumor recurrence, which was concluded as an excellent example of integrating diagnostic caution, oncological principles, and biomechanical judgment in managing pediatric surface bone lesions.

Conclusion

This case describes a posterior distal femoral metadiaphysis surface bone tumor accompanied by an extraosseous lesion that demonstrated imaging findings similar to previously reported cases and therefore required careful differentiation from parosteal osteosarcoma. Because these entities share overlapping clinical and radiological characteristics, reliance on a single diagnostic modality may lead to misdiagnosis and inappropriate treatment. A comprehensive and stepwise evaluation integrating imaging, histopathology, and immunohistochemistry allowed accurate diagnosis and appropriate surgical management, especially in growing bone.

Conflicts of Interest

All Authors declare no conflict of interest in relation to this report.

Authors' Contributions

Conceptualization was performed by RW and SM. Investigation, formal analysis, and methodology were carried out by SM, YT, SMi, TH, HY, and YA. Data curation and visualization were conducted by RW and SM, with resources and project administration provided by SM. The original draft was written by RW. Writing – review and editing were performed by SM, SMi, TH, HY, and YA. Supervision and validation were undertaken by KH and SD. Funding acquisition was not applicable. All Authors read and approved the final manuscript.

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Artificial Intelligence (AI) Disclosure

During the preparation of this manuscript, a large language model (GPT-5.2, OpenAI) was used solely for language editing and stylistic improvements in select paragraphs. No sections involving the generation, analysis, or interpretation of research data were produced by generative AI. All scientific content was created and verified by the authors. Furthermore, no figures or visual data were generated or modified using generative AI or machine learning–based image enhancement tools.

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