Unveiling the Rarity: A Case Report on Distal Femur Parosteal Osteosarcoma

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ABSTRACT

The parosteal osteosarcomas represents a rare low grade malignant but less metastasizing tumour. They often present with a simulating clinic-radiological features of other similar conditions like osteochondroma. Local wide Excision with hemi-cortical resection usually behaves better without recurrence. We are unveiling one of such case who reported with progressively increasing swelling, pain in popliteal fossa and difficulty on squat for the last 2 years. Its location at distal femoral metaphysis and radiographic appearance was little confusing for osteochondroma. Whereas MRI findings suggested it as of a parosteal osteosarcoma. The wide local excision was successful with no recurrence at 2 years of follow-up.

Keywords: Osteosarcoma, Neoplasms, Bone Tissue, Margins, Resection.

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INTRODUCTION

The Parosteal osteosarcoma (POS) constitutes 4.3% of all osteogenic sarcomas, that characteristically behave as a low-grade osteosarcoma with a low-metastatic potential. The POS characteristically present as a large mass, arises from cortical bone of metaphyseal region, commonly from distal femur[1,2]. POS affects women more than males with 2:1 ratio and mostly affecting during third to fourth decades of life[1]. Due to its low grade, and well-differentiated nature, POS seldom or rarely involves the medullar canal, which may occupy less than 25% of the canal's width[3]. Wide Local excision often behaves successful without recurrence. However, POS with intramedullary extension may necessitate an extensive wide local excision and a reconstruction with allograft or hemi to complete prosthetic replacement based on individual case requirements[4-5]. To accomplish the targets of recurrence prevention a precise diagnosis based on radiologic findings, CT-MRI evaluation significantly needed.

CASE PRESENTATION

A 22 years old female with no known comorbid presented with complaints of a progressively increasing hard swelling in popliteal region of right femur. Earlier most she noticed a small swelling arising from back of knee 2 years back. That was non tender and not disturbing her movements and was overlooked with off and on taking over the counter pain killers. For the last 6 months she got apprehensive as the mass becoming prominent, painful, tender on deep touch, developed restricted flexion at knee, difficulty to sit in squat and

Tashahud (kneeling). This was progressively increasing and she started feeling hard tender mass. She had no history of trauma, fever or a constitutional illnesses. She did not have a relevant family history for malignancies.

The clinical examination revealed a tender hard swelling in central part of right popliteal region, which was not adherent to skin, fixed to distal femur and become prominent with forceful complete extension of the knee. There was fixed flexion deformity of knee for 20 degrees and range of flexion from 20^0 -120°, with extension lag of 20^0. The muscle power and sensation were within normal limits. The initial clinico- radiologic findings were confusing, rather supporting diagnosis of osteochondroma Figure 1). MRI however, appreciated well-defined heterogenous enhancing abnormal signal intensity lesion. There was no intracortical or medullary continuity and soft tissue extenxion. Overall finding most likely represented neoplastic lesion like parosteal osteosarcoma.

Local wide excision of tumour was carried out, through a S-shaped incision at popliteal fossa. With a meticulous dissection, tumour was resected with thickened soft tissue and periosteal mass along with

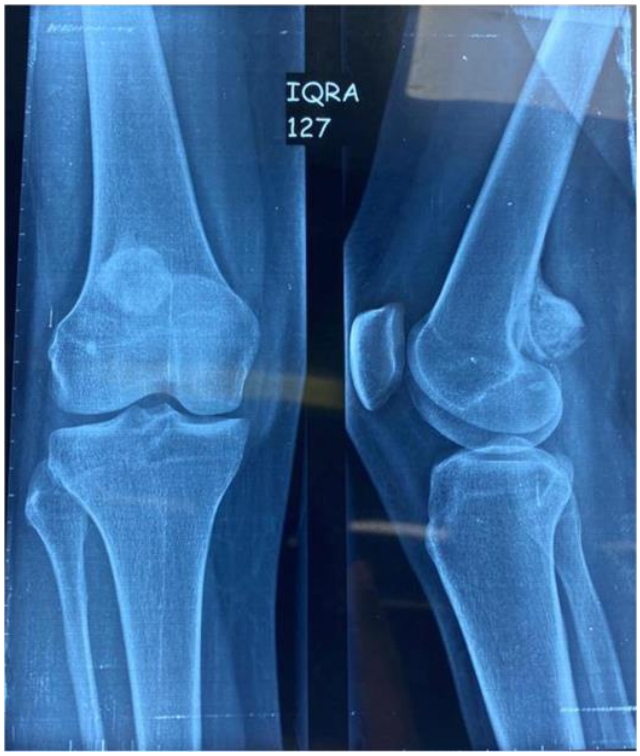

Figure 1: Radiograpghic picture revealed well circrumscibed mass fixed to posterior central cortex of distal femur. A maximal radiodensity near tumour base while blurred outer margin is well appreciated.

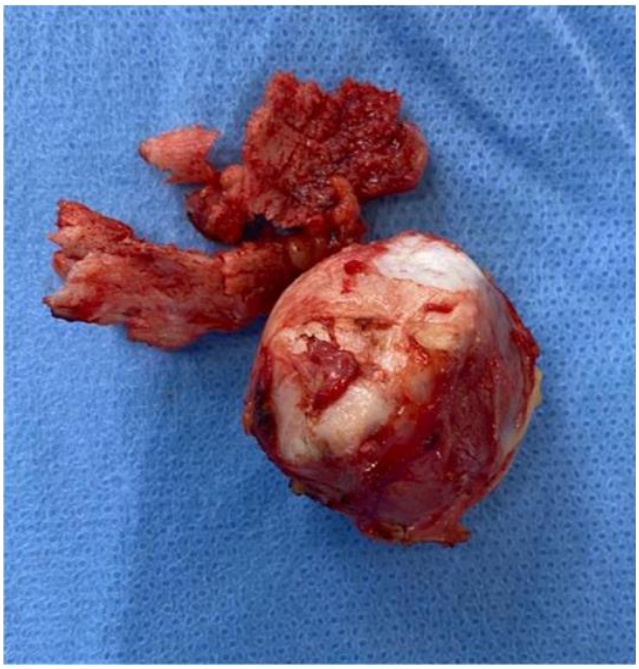

Figure 2: Wide Local excised globular mass along with excised hemicortex, thickened soft tissue and periosteal mass. The base of tumour tissue was granular hard mass.

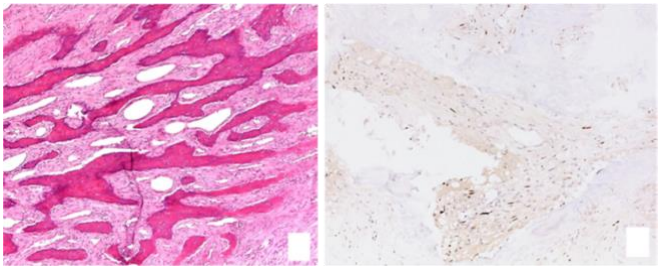

Figure 3. A. Characteristic microscopic arrangement of parallel bone trabecula and the intervening low-grade fibroblastic component (H&E; 50X). B. MDM2 nuclear positivity (IHC; 50X). **Source:** Picci P, Manfrini M. Diagnosis of musculoskerletal tumours and tumour like conditions. 2nd ed. 2020. Switzerland: Spriger Nature.

hemicortictomy of popliteal surface of distal femur (figure 2). The surgery was uneventful, wound closed in layers without placement of a drain. The underlying base of mass was granular, extending within medullary canal to some extent that was scooped out as well (figure 2). The histopathology report revealed a low-grade parosteal osteosarcoma. Margins could not be assessed as tumour was present in both nodular bony tissue and separate tissue. Lymph nodes or any area of necrosis was not identified. The oncology consult did not advised chemo-radiotherapy. Her subsequent clinico-

radiologic followups for the last two years were uneventful. She regained full ROM, pain free and was able squat and sit in tashahud at ease.

DISCUSSION

The Parosteal Osteosarcoma (POS), Osteochondroma (OCM), Bizarre Parosteal Osteochondromatous Proliferation (BPOP) and Juxtacortical Myositis Ossificans (JCMO) represent few distinct simulating primary bone tumors[5]. Osteochondromas are typically benign arising from peripheral part of the growth plate, characterized by having covered with a cartilage-capped bony projection. That typically grow away from joint margin. Whereas the Parosteal Osteosarcomas are low-grade malignant tumor originating from the periosteum of juxta-metaphyseal or from meta-diaphyseal regions of long bones. Mostly remain centric. Distal femur popliteal surface being its favored location in 60% Cases. The BPOP have characteristic predilection for long tubular bones, whereas POS exceptionally may affect flat bones, small tubular bones of hands and feet and vertebrae[5]. Clinically all these Tumours most commonly presents as a long indolent, painless and indurated mass that usually discovered when it has achieved large dimensions. The presented case has similar natural history hence was confused with as a malignant transformed OCM.

The parosteal osteosarcomas often present as a circumferential or mushroom shaped, exophytic and lobulated mass with a broad base attachment to the cortex of the host bone [Figure1]. The base of POS display radiodensity while the outer margin tends to be blurred. Tumour mass body often display appearance ranging from ground glass to ivory density with overall look of mesh of trabecula (steel-wool pattern). POS grow either longitudinally along the long axis of the bone or may encircle it. Elevation of periosteum and reactive periosteal bone formation are typically absent. This exclusive appearance of POS is well evident on CT scans and MRI in an approximately 60% of dedifferentiated parostal osteosarcomas[1,6-7]. The combination of Xray, CT and MRI scans allows significantly high accuracy in the determination of the extent of intramedullary infiltration of the tumor, thus assisting in the planning of conservative surgeries and to avoid surgical morbidity[7]. The findings in presented case were not so defined hence a confusion for aggressive OCM was considered an option in deferential diagnosis.

The cut surface of POS appears whitish, hard, or gritty and may contain areas of cartilage [Figure 2]. Microscopically it exhibits with atypical osteoblastic cells producing osteoid and chondroid matrix. That appear as long fascicles of deceptively bland spindle cells embedded in a collagenous stroma and intervening well-formed bone trabecula. The fibrous fascicles are hypocellular, the spindle cells exhibit minimal nuclear atypia and inconspicuous mitotic figures. [Figure 3][1,6-7].

A marginal excision (hemicortical resection) with preservation of the articular surfaces has been reported as an optimal approach to excise the localized POS involving distal femur. The marginal excision with histologically negative picture have been reported sufficient for prevention of distant metastases and a long-term survival rate of at least 90%[5,13,3,5,8-9]. Whereas, with wide margin excision, the resultant hemicortical defect may need reconstruction with allograft to provide structural support to facilitate an optimal early functional outcomes[3,8-9]. Moreover, Kamal, A.F., Kodrat[5] reported no significant and consistent correlation between local recurrence and medullar involvement (<25%) that necessitate a radical excision reconstruction with a endo-prosthesis[4,10]. Neoadjuvant and adjuvant chemotherapy has also been recommended by some investigators to prevent local and distant metastasis in complicated cases[4].

In this reported case MRI did not revealed an intramedullary extension, marginal hemicortectomy excision was carried out. The histopathology revealed un-clear margins. The patient however showed no recurrence till 2 year follow-up. Moreover, patient has been advised for a quarterly clinic-radiologic surveillance for a long term follow-up.

CONCLUSION

The parosteal osteosarcoma might be misdiagnosed as a fast growing osteochondroma, Bizarre Parosteal Osteochondromatous Proliferation or Juxtacortical myositis ossificans. Adequate clinic-radiologic evaluation and planned excision of clear margins are therefore crucial in preventing local recurrence. The wide-margin excision usually suffice good without significant recurrence. The presented case example illustrates for a meticulous pre-operative planning that did not revealed an intra-medulary extension. The wide local marginal excision however, eliminated tumour without recurrence so far till 2 years followup

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