Case Report

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Bizarre Parosteal Osteochondromatous Proliferation Arises from Proximal Femur: A Case Report

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Abstract

Bizarre parosteal osteochondromatous proliferation (BPOP) of long bones is extremely rare and often confused with osteochondroma. Careful correlations of clinical, imaging, and pathological findings give the diagnosis. A 17 years male patient presented with a progressive occasionally painful swelling over the lateral aspect of the left proximal thigh of 2 years duration without any history of trauma. The mildly tender, hard bony femoral mass was having lobulated surface. Imaging showed heterogeneously opaque bony mass without medullary canal continuity. Cartilage cap was thin with an irregular surface. During operation cortical surface of the femur was intact. It was covered with thick perichondrium, but the cartilaginous cap was thin. Histopathology demonstrates production of bluish tint cartilage with irregularly arranged chondrocytes of different stages of maturation and lamellar bone with intertrabecular spindle cells without cytological atypia. Appropriate diagnosis of BPOP can only be made when it is clinically suspected, careful radiological evaluation is made. That guides surgeon for meticulous excision to prevent recurrence. Finally, careful histopathological examination confirms the diagnosis.

Key words: Bizarre parosteal osteochondromatous proliferation, Nora's disease, Long bone

INTRODUCTION

Bizarre parosteal osteochondromatous proliferation (BPOP) also known as Nora's disease is a rare condition. It involves small bones of hands and feet.[1] Very few of them develop from long bones.[2] Florid periostitis, Turret exostosis, and BPOP are believed to be different stages of post-traumatic osteochondral proliferation.[1] However the incidence of trauma may be absent in few occasion.[3] Many a time the clinical, radiological and histopathological (HP) features are confused with that of solitary osteochondroma, parosteal osteosarcoma, chondrosarcoma and heterotrophic ossification.[1] Unless meticulously excised it has a strong tendency for recurrence.[3,4]

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This case is being reported not only because of its rarity but careful evaluation of clinical and imaging features give strong suspicion to guide surgeon for meticulous resection to prevent recurrence. Confirmation of diagnosis is obtained after careful HP study. Post-operative counseling and the need for adequate vigilance are also important issues.

CASE REPORT

A 17 years male patient presented to the Outpatient Department of our hospital in October 2013 with a progressive swelling over the lateral aspect of the left proximal thigh of two years duration. It was associated with occasional mild pain which is independent of exertion. Family history and history of trauma are non-contributory. Hard mildly tender non-indurated and lobulated swelling was found over the proximal thigh of size 7 cm and 6 cm in vertical and transverse dimensions, respectively (Figure 1a). It was attached to the bone but free from overlying structures. X-ray shows lobulated bony outgrowth from proximal femoral shaft just distal to greater trochanter and

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directed laterally (Figure 1b and c). In magnetic resonance imaging, the heterogeneous surface was covered with a thick membrane which was non-continuous with shaft periosteum. It did not show any medullary continuity (Figure 2a-c). Under spinal anesthesia operation was done in the lateral position and using vertical lateral incision. After excision of the mass, the intact cortical surface under the bed of the swelling can be seen (Figure 3a). A wound was then closed in layers after achieving hemostasis. Cut surface of the excised specimen showed heterogeneous trabeculations without a medullary cavity (Figure 3b and c). The patient was allowed active movements on day one and permitted weight bearing on the third post-operative day. Stitches were removed at the 14th post-operative day.

HP demonstrates mature bone formation and plenty of spindle cells without evidence of malignancy in the intertrabecular areas with bluish tint cartilage formation with different sized irregularly arranged cartilage cells (Figure 4).

The patient was followed up at 2 weeks, 1 month, and monthly up to 6 months. X-ray at 6 months did not show any recurrence (Figure 5).

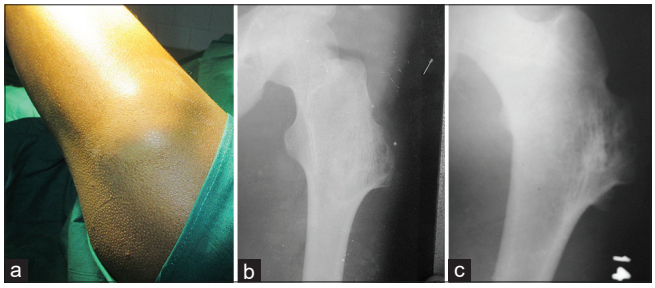

Figure 1: (a) Clinical picture of the lesion over lateral aspect of proximal thigh of left side, (b) X-ray picture in anteroposterior view, (c) X-ray picture in lateral view

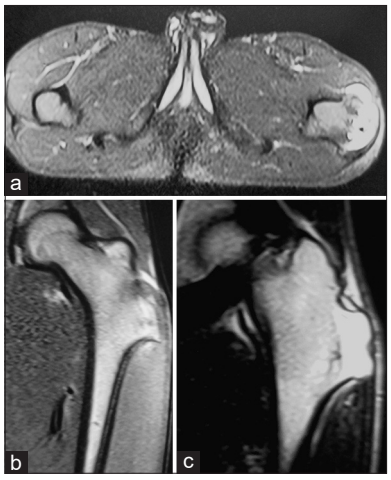

Figure 2: (a) Magnetic resonance imaging (MRI) picture in transverse section, (b) MRI picture in coronal section, (c) MRI picture in coronal section showing junction of intact cortical bone and the mass

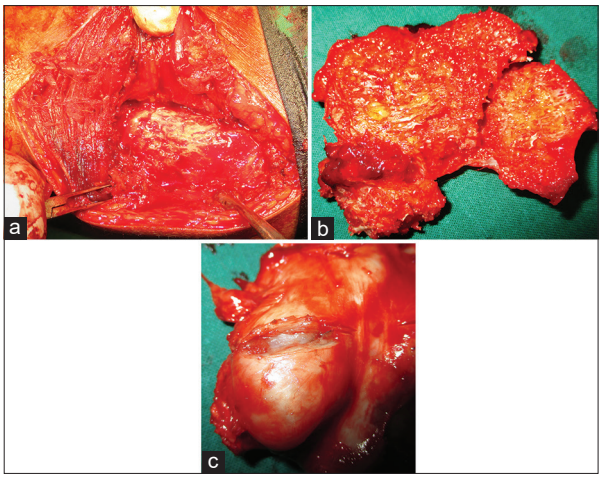

Figure 3: (a) Peroperative picture showing intact cortical surface, (b) excised specimen without medullary canal, (c) specimen showing thick perichondrium but thin cartilage cap

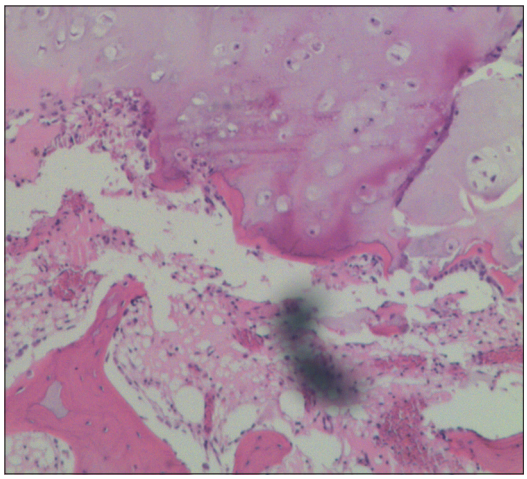

Figure 4: Pinkish bone with intertrabecular spindle cells without evidence of malignancy and cartilage formation at the interface with bluish tint and cartilage cells are irregularly arranged and of different stages of maturation

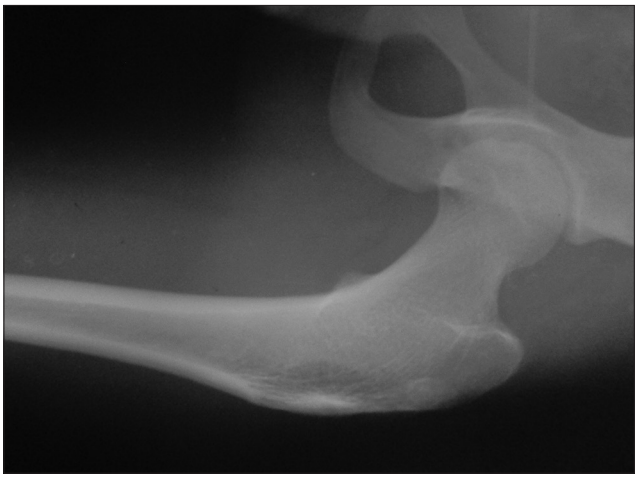

Figure 5: Follow-up X-ray after 1 year without evidence of recurrence

DISCUSSION

The diagnosis, in this case, was based on clinical, imaging, and supported by histopathology. Possibilities, in this case, are non-hereditary solitary exostosis,[5,6] BPOP,[1-4,7,8] Turret exostosis, and florid reactive periosteitis.[9] Myositis ossificans are also excluded in relation to this case.[10] Parosteal osteosarcoma, secondary chondrosarcoma are also confusing at times in histopathology findings.[1]

Non-hereditary solitary exostosis usually presents within the 3rd decade. They are directed toward diaphysis and may have medullary canal is in continuity with that of the parent bone. Metaphyseal osteochondroma is believed to stop growing with skeletal maturity. Hameetman *et al.* demonstrated mutation of tumor gene and also correlated with solitary exostosis. The traditional theory of "skeletal dysplasia" is shifting toward the theory of "cell-of-origin."[5,6] This might be the one explanation of continued growth after skeletal maturity. In this case, radiological findings and specimen examination do not show very thick cartilage cap. HP study distinctly demonstrates irregularly arranged cartilage cells at different stages of maturation which distinctly differ from exostosis.

Adler *et al.* redefined BPOP, also known as Nora's lesion, as the proliferation of bone and usually emanates from the intact cortical surface of short bones and rarely in long bones.[2] It is assigned to be reactive heterotrophic ossification. Though generally correlated with trauma, few authors denied it and genetic mutation supposing tumor genesis has been recently suggested.[3] Usual age group of the sufferer is in the 3rd or 4th decades.[7] In the early stage, cortical continuity may not be there but ultimately develops it. In some series there is some male preponderance.[4] Imaging in most occasions resembles sessile variety exostosis. BPOP apparently arises from the periosteal tissues through a process of cartilaginous metaplasia. Microscopically, it is composed of hypercellular cartilage with calcification with basophilic tintorial character and ossification with pinkish appearance.[8] Cancellous bone undergoes maturation and presence of bizarre spindle cells in the intertrabecular space creates confusion with parosteal osteosarcoma but, in this case, they are without cytologic atypia.[1] There is thinning of cartilage cap in long-standing cases. These features are similar to the present case. 50% local recurrence is reported in the literature.[3] This patient did not have recurrence in early follow-up. Secondary chondrosarcoma is not a possibility in this case as there was no primary lesion and HP features are not in its favor.

LeClere *et al.* believe Turret exostosis and florid reactive periostitis are benign osteo-cartilaginous lesions arise from reactive periosteum following relatively mild trauma.[9] Microscopically, the central area of mature bone from endochondral ossification characterized by thin hypocellular peripheral rim of cartilage and absence of periosteum over the surface. No history of trauma can be obtained in this case.

Myositis ossification develops in more peripheral from bone and ossification starts from periphery to center.[10]

CONCLUSION

As biopsy is not usually done prior to excision, a careful clinico-radiological examination creates strong suspicion for the diagnosis of BPOP for an atypical variety of bony outgrowth arising from short and ends of the long bones. This suspicion is important as BPOP has a strong tendency for recurrence, unlike solitary exostosis. Meticulous resection can minimize the chance of recurrence.

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