Bizarre Parosteal Osteochondromatous Proliferation of the Femur (Nora’s Lesion) - A Case Report -

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- Abstract -

We report a case of bizarre parosteal osteochondromatous proliferation of the right femur in an 18-year-old man. Roentgenograms showed a calcific mass attached to the underlying cortex with a broad base. Histologically, the lesion showed hypercellular cartilaginous tissue with maturation into trabecular bone, which contained spindle cells and lymphocytes in the intertrabecular spaces. Bizarre parosteal osteochondromatous proliferations are a form of heterotopic ossification and should not be mistaken for osteosarcoma or chondrosarcoma.

Key Words: Bizarre parosteal osteochondromatous proliferation, Femur

INTRODUCTION

Bizarre parosteal osteochondromatous proliferation of bone (BPOP; Nora’s lesion) is a rare, benign bone lesion, which was first demonstrated in 1983 by Nora et al. It was first regarded as occurring only in the small bones of the hands and feet, but in 1993, Meneses et al. reported 65 cases including those involving long bones. Bizarre parosteal osteochondromatous proliferation can often be mistaken for other benign bone tumors, such as osteochondroma. It is also confused with malignant tumor due to its high recurrence rate and aggressive histologic findings. In order to make a proper diagnosis, it is necessary to correlate the clinico-radiologic and pathologic characteristics appropriately.

We present a case of BPOP occurring in the left femur and discuss the clinicopathologic characteristics of this lesion.

CASE REPORT

A 18-year-old man was presented with complaints of a painful swelling of the right knee. He had had a painless growth at the same area of the knee for about 2 years,
and suffered from an aggravation of pain and swelling about one month ago. He could not recall any history of trauma. Physical examination revealed diffuse swelling and tenderness on the distal aspect of the right femoral area. Plain radiographs (Fig. 1) revealed a relatively-demarcated calcified mass arising from the medial cortical surface of the metaphyseal area of distal femur.

On magnetic resonance imaging (MRI), the mass revealed low signal intensity in T1W image (Fig. 2A) and intermediate signal intensity in T2W image (Fig. 2B). The cortex of the underlying bone was intact, and directly contacted with the mass, which did not involve the marrow nor have significant soft tissue extension. At the time of excisional biopsy, the mass was covered with cartilage cap resembling osteochondroma and well demarcated from the surrounding tissue.

On histologic examination, the mass showed a disorganized intermingling of cartilage, bone, and fibrous tissue. The cartilage cap (Fig. 3A) revealed hypercellular chondrocytes with nuclei of variable size and mild to moderate atypia (Fig. 3B). The osseous component consisted of trabeculae of bone with wide variation in configuration and width (Fig. 4). The trabeculae were arranged at various angles to one another without any apparent organization. The trabecular bone is focally lined by osteoblasts with uniform size and lack of atypia. A proliferation of benign fibrous tissue was interspersed between the trabeculae. The cartilage-bone interface demonstrated typical irregular maturation of cartilage into bone, resembling fracture callus. There was also a distinct blue tinctorial characteristic at the interface of the cartilage and ossified bone. The diagnosis based on the pathological specimen was consistent with bizarre parosteal osteochondromatous proliferation (Nora’s lesion).

After surgical excision, he have had no evidence of recurrence for 48 months.

DISCUSSION

BPOP was originally described as occurring only in the hands and feet, but later literature by Meneses et al. found that the lesion also involved the long bones of extremities in about one fourth of cases. In Korea, Kim et al. reported 5 cases of BPOP including one recurrence case. Clinically, BPOP occurred equally in both sexes and the ages of the patients at diagnosis ranged from 8 to
74 years, with a median of 34 years\textsuperscript{2,3}. These lesions are usually small ranging from 0.4 to 3cm in diameter and the rate of growth varies from months to years\textsuperscript{2,3}. Instead of pain or redness, the presenting symptoms are due to bony tumefaction.

The differential diagnosis of this lesion includes various benign and malignant lesions such as osteochondroma, subnugal exostosis, heterotopic chondroossification\textsuperscript{6}, florid reactive periostitis, myositis ossificans, and parosteal osteosarcoma. BPOP often looks like more common osteochondroma because of paracortical position and cartilage cap. However, in osteochondromas, chondrocytes lack atypia and often are arranged in parallel lacunar spaces. On the other hand, BPOPs do not have continuity with underlying medullary cavity. Radiologic correlation is helpful. Parosteal osteosarcoma has been described in hands\textsuperscript{5,7} although it is more common in long bones, and may be histologically similar to BPOP. In long bones, the lobular architecture of BPOP, distinct blue tintorial characteristic of the osteoid, and slender, irregular bony trabeculae are helpful in differentiation. BPOP is small, and does not invade the adjacent tissue nor metastasize. Subungal exostosis is usually painful and has a characteristic location. Histologically it is differentiated from BPOP due to lack of the chondroid material. Myositis ossificans is recognized by the maturation and ossification first found at the periphery of the tumor. The cartilage usually does not form a cap. The lesion is usually separated from the bone and radiologically often shows associated periosteal reaction on the adjacent bone. Some authors consider florid reactive periostitis as a form of myositis ossificans\textsuperscript{6}. It shares similar histologic characteristics with BPOP, but bizarre cartilaginous proliferation is not prominent. Another authors proposed a term, proliferative periosteal processes of phalanges, for those separately designated previously as

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Fig. 2. Mass with low signal intensity in T1 weighted MR image (A) and intermediate to high signal intensity in T2 weighted MR image (B).
proliferative periostitis, BPOP, subungal exostosis and turret exostosis\(^8\). They hypothesized that these entities developed from the same initial stimulus, often related to trauma, that leads to hemorrhagic subperiosteal proliferation, and their different appearances and reflections of temporal factors, breaching of the periosteum, and local anatomic features\(^8\).

The recurrence rate of BPOP is very high. In earlier studies, more than half of the patients developed recurrent lesions and most of them were within 2 years of the first excision\(^2,3\). Despite the high local recurrence rate and the bizarre histologic appearance, BPOP is a benign lesion with no reported metastasis or deaths. The optimal treatment is complete removal by simple excision.

REFERENCES

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