Bizarre Parosteal Osteochondromatous Proliferation of the Second Metatarsal Bone (Nora’s Lesion)

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Parosteal bone tumors involving the small bones of the hand and the foot are rare. Their clinical and imaging features may imitate other bone tumors, leading to incorrect diagnosis and management. Histological examination of the lesion will establish the correct diagnosis.

Bizarre parosteal osteochondromatous proliferation was first described in 1983 by Nora et al. In a report of 35 Mayo Clinic cases, Nora et al described the histological structure of the lesion and classified it as benign tumor of bone.

In a recent review of 150 bone tumors of the feet, Bakotic and Huvos found 3 cases of bizarre parosteal osteochondromatous proliferation accounting for roughly 2% of all bone tumors of the feet.

This article reports a case of a 20-year-old woman with a bizarre parosteal osteochondromatous proliferation of the second metatarsal of the left foot.

**CASE REPORT**

A 20-year-old woman presented with an 8-month history of an enlarging mass of the second metatarsal of the left foot. There was no history of trauma. The mass was occasionally painful and caused difficulty in fitting shoes.

Clinical examination revealed a palpable mass over the second metatarsal. There was no tenderness on palpation.

Plain radiographs showed a well-circumscribed lesion arising from the cortex of the distal third of the second metatarsal (Figure 1). There was no periosteal reaction.

With a presumptive diagnosis of a juxta cortical aggressive tumor, a wide resection of the tumor was performed, leaving the posterior cortex intact. The dimensions of the excised lesion were 3×2×1.2 cm. Histological examination revealed isolated chondroid areas with hypercellularity, marked nuclear swelling and binucleated cells, as well as osseous elements associated with marked osteoblastic activity and osteoid formation. The fibrous interval stroma had no osteoblastic activity (Figure 2). The diagnosis of bizarre parosteal osteochondromatous proliferation of bone (Nora’s lesion) was made.

Postoperatively, partial weight bearing was recommended for 4 weeks. At the latest follow-up 27 months postoperatively, the patient was asymptomatic with no difficulty in wearing shoes or walking and there was no evidence of local recurrence (Figure 3).

**DISCUSSION**

Bizarre parosteal osteochondromatous...
proliferation of bone is a rare benign bone neoplasm usually seen in young adults, although other age groups cannot be excluded. The lesion involves the small tubular bones of the hands and feet (76%). Less frequently, the lesion is located in the femur, humerus, fibula, radius, and skull. According to the literature, the tumor is commonly seen in phalanges and its appearance in the metatarsal, as in the present patient, is rare. Previous trauma does not appear to be a definite etiological factor, however, it is highly possible that the traumatic insult is simply not recalled by the patient.

Figure 2: Irregular mixture of bone (arrow head), cartilage (arrow) and fibrous tissue (H/E 3 100) (A). Cellular cartilage with oddly bizarre nuclei (H/E 3 200) (B).

The typical clinical characteristic of this tumor is the lack of pain at rest as well as during palpation. The mass grows slowly and manifests itself with discomfort rather than pain, as occurred in the present patient.

The radiographic appearance of this bone lesion is typically a well-circumscribed area, ranging from 0.4–3 cm in diameter, without periosteal reaction and osteolysis of the cortex. The mass gives the impression that it originates from the cortical bone without expansion to the intramedullary space. The lesion may expand into the soft tissue, as it did in the present case; however, it does not infiltrate the soft tissue.

Radiographic differential diagnosis includes conventional osteochondroma, subungual exostosis, florid reactive periostitis, myositis ossificans, turrett exostosis, peripheral chondrosarcoma, perioskeletal and parosteal osteosarcoma, and periosteal chondrosarcoma. Osteochondromas are uncommon in the small bones of the distal extremities and are not characterized by an intact cortex underneath as in the Nora’s lesion. Because of its parosteal location, bizarre parosteal osteochondromatous proliferation of bone should be distinguished from parosteal osteogenic sarcoma. Parosteal osteosarcoma is rarely found in the hands and feet. It typically appears as a dense lobulated mass attached by a broad-based pedicle to the cortex and has heavy mineralization with a sclerotic appearance on plain films.

Bone scintigraphy typically shows intense radioisotope uptake at the site of the lesion. Computed tomography defines the extent of the lesion. Computed tomography findings include a mass with well-defined margins, intensely calcified or ossified, arising from the cortex of the affected bone and showing a wide base. There is no continuity with the medullary canal of the bone from which it originates, and cortical flaring of the affected bone is absent.

Magnetic resonance imaging shows lack of bone marrow involvement and no significant soft-tissue expansion. The absence of soft-tissue swelling and cortical destruction, as well as lack of stress fracture or medullary involvement by the tumor, are all useful in defining the true nature of these lesions. Magnetic resonance images show low signal on T1-weighted sequences and high signal on STIR imaging. On fast spin-echo T2-weighted imaging, the lesion is usually of high signal, and sometimes the lesion is of intermediate signal centrally and uniform high signal peripherally on both fast spin-echo T2 and gradient imaging sequences. The variable signal on fast spin-echo T2 and gradient imaging sequences may be related to the relative amounts of cartilage present within the lesion.

Histological features include a cartilaginous tissue with abundance of large chondrocytes, some of which are binucleated. The lesion does not as a rule lack cytologic atypia. For this reason, it is
often misdiagnosed as low-grade sarcoma. Binucleation, nuclear enlargement, and pleomorphism are all seen in a large fraction of cases. There are areas with enchondral ossification of the cartilaginous cap. The osteoblasts lining the bony trabeculae, although they lack signs of cellular atypia, may be quite prominent and not necessarily uniform. Spindle cells are present in the spaces between trabeculae with no signs of cellular atypia.

Bizarre parosteal osteochondromatous proliferation has a distinct histologic appearance that permits differentiation from other benign lesions or low-grade malignant tumors. In osteochondroma, chondrocytes often are arranged in parallel lacunar spaces, whereas in Nora’s lesion the cartilage is disorganized and irregular. Osteochondroma also contains cancellous bone and hematopoietic marrow that is shared with the host bone. Parosteal osteosarcoma can present with a cartilage cap, but the stroma displays cytologic atypia. The cornerstone of parosteal osteosarcoma is the spindle-cell proliferation of cytologically bland-looking spindle cells and normal bony trabeculae.

Immunohistochemical and molecular analysis of FGFR, VEGF, and ChM-I strengthened the concept that bizarre parosteal osteochondromatous proliferation is a reparative process, which is similar to enchondral ossification in the growth plate.

Wide resection is the treatment of choice for bizarre parosteal osteochondromatous proliferation. Incomplete tumor excision is associated with remarkably high rates of local recurrence ranging from 22% to 55%. Despite this high recurrence rate, there have been no reported metastases, and no adjuvant therapy is recommended. Time interval between excision and local recurrence ranges from 2 months to 2 years. The time interval between excision of a recurrent lesion and second local recurrence ranges from 3 to 13 years. Although osteochondromas may recur after incomplete excision especially in a growing child, local recurrence after removal of the lesion may suggest the diagnosis of a Nora’s lesion. In conclusion, because of its rarity, bizarre parosteal osteochondromatous proliferation can be confused with both benign and aggressive neoplastic processes, and incorrect treatment measure may ensue. Therefore, it is important that this lesion be recognized early and treated appropriately.

REFERENCES