A Case Study: Bizarre Parosteal Osteochondromatous Proliferation (Nora's Lesion)

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STATEMENT OF PURPOSE

Nora's lesions or "bizarre parosteal osteochondromatous proliferations" (BPOP) are rare lesions of osteochondromatous origins. Few cases have been reported in the literature and its pathology has remained ill-defined. Due to its rare presentation, it is frequently misdiagnosed, and therefore often mistreated. In this report, we describe a case of a Nora's lesion and the associated surgical management with long-term follow-up results.

LITERATURE REVIEW

BPOPs were first described in a case report of 35 patients by Nora et al in 1983. Less than 200 cases have been presented in the literature. The lesion affects patients of any age and affects both sexes equally.

Clinically, the lesions can be mildly painful. Trauma is purported to play a role. The tumor affects the long tubular bones of the hands and feet especially the phalanges. The hands are four times more commonly affected than feet. This is a benign lesion of the bone that can be mistaken for malignant processes due to the high rate of recurrence ~50%, rapid growth, and atypical histologic appearance. The lesion is an exophytic outgrowth from the cortical surface consisting of bone, cartilage, and fibrous tissue.

Radiographs may show a "well-marginated mass of heterotopic mineralization arising from the periosteal aspect of an intact cortex, without medullary changes". CT and MRI are non-specific but may show an ossified mass with well-defined margins and heterogeneous intensity, respectively. Diagnosis can be confirmed by histopathology, which may show "spindle shaped or stellate small chondrocytes scattered in a myxoid stroma"; the basal area may be composed of "immature bony trabeculae with high osteoblastic activity".[3]

Due to its rare clinical presentation and aggressive features it must be distinguished from malignant lesions. Treatment consists of excisional biopsy.

CASE STUDY

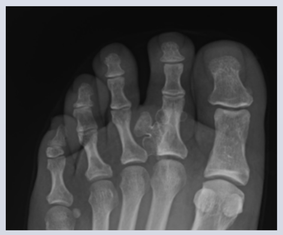
Subjective

A 30-year-old female presented with a one-year history of a soft tissue mass on the left second digit after stubbing her left second toe. She then noticed progressive growth of the lesion. Outside hospital radiographs were taken, which showed a fracture of her proximal phalanx. The patient continued to relate pain and presented to Olive View-UCLA Medical Center after stubbing her toe again. The patient had an unremarkable past medical and family history, negative for any benign or malignant masses and negative for an immunocompromised state or prior infection history. She was not on any medications.

Physical Exam

Lower extremity exam revealed an enlarged left second digit with a palpable mass extending into the second interspace. The mass was firm, non-mobile, and did not transilluminate. The prominence was mildly painful on palpation. No erythema, increased warmth, or open lesions. Neurovascular status was intact. No pain with range of motion and no gross structural deformities of her foot or ankle were noted. No noted lymphadenopathy.

Imaging



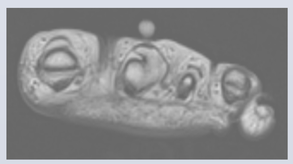


Fig. 1,2 XR and T1 sagittal MRI

Initial radiographs showed a presumed bizarre parosteal osteochondromatous proliferation arising from the lateral cortex of the proximal phalanx of the left second toe. MRI revealed an exophytic mass extending off the lateral aspect of the proximal phalanx of the second digit with continuity between the medullary cavity of the phalanx and the mass. The mass had characteristics most consistent with a pedunculated osteochondroma.

CASE STUDY

Treatment

The patient was subsequently scheduled for an excisional biopsy of the osseous tumor. Intraoperative findings consisted of a bony mass in the second interspace, protruding off the proximal phalanx of left second digit. The lesion was sent to pathology for histological analysis and was read as lamellar bone with areas of enchondral ossification and fibrous tissue. The intraoperative features and radiographs were not typical for osteochondroma; however, histological features were more consistent with a Nora's lesion.

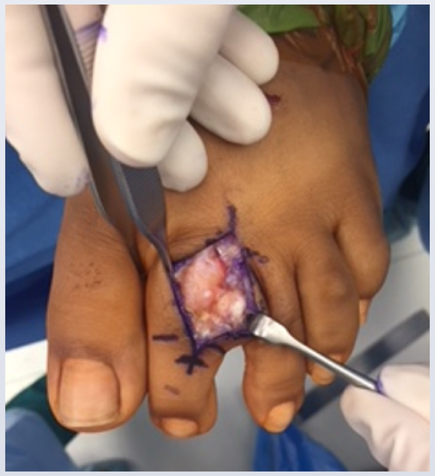
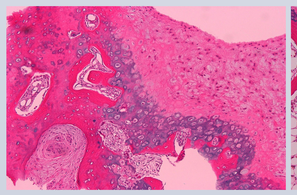


Fig. 3 Intra-operative findings

Post-Operative Care

Patient was initially non-weightbearing in a splint and crutches. Sutures were removed at two weeks and a dehiscence of the surgical site had occurred. Wound biologics were applied. At two months post-operatively, the dehiscence from the surgical site was healed. At one-year follow up, no recurrence was noted.



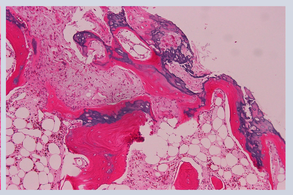


Fig. 4, 5 Histopathology of lesion

ANALYSIS & DISCUSSIONS

BPOP is a lesion that remains a challenging diagnosis due to its extremely rare presentation and its radiographic similarity to more common osteochondromatous tumors. These lesions have high recurrence rates between 29-55% in a two-year interval[1]. Despite a high tendency of recurrence and sometimes atypical histological appearance, no malignant transformation, metastases, deaths or associated systemic disease have been reported so far in patients[1].

Bizarre parosteal osteochondromatous proliferations rarely occur but should be considered as a differential diagnosis when encountered with a benign appearing bony mass in long bones of the hands and feet. Conservative treatment in general is considered inadequate and excisional biopsy is generally indicated especially when symptomatic. Because of high recurrence rates (20-55%), close follow-up is recommended.

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