# Case Report of a Novel Transformation of an Osteochondroma to a Nora's Lesion (Bizarre Parosteal Osteochondromatous Proliferation)

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## Purpose

Bizarre parosteal osteochondromatous proliferation, otherwise known as Nora's lesion, is a rare benign bone tumor with a high local recurrence rate and diagnostically challenging growth patterns, radiologic features, and histologic appearance. There have been no documented cases to present of tumor transformation, malignant or otherwise. This case report describes a novel transformation of an osteochondroma of the right 2nd proximal phalanx to a Nora's lesion in a 52 year old female.

## Literature Review

Bizarre parosteal osteochondromatous proliferation (BPOP) is a rare benign bone tumor, first described by Nora et al. in 1983 and is thus otherwise known as Nora's lesion. It was originally described as having a predilection for the metatarsals, metacarpals, and proximal phalanges of the hands and feet, but with increased reporting has been found in multiple long bones and the skull (1, 2). BPOP affects males and females in equal proportions, occurring most commonly in the third and fourth decades of life (2). Nora's lesions tend to be under 4cm, and symptoms are most often related to the tumefaction, particularly when it is fast growing, despite being benign (2, 3). Dhondt et al. defined the radiologic features of Nora's lesion as a well-marginated mass of heterotopic mineralization arising from the periosteal aspect of an intact cortex, without medullary changes, identifying it as a distinct radiologic entity along a spectrum of florid reactive periostitis and turret exostosis (3). MRI findings include low signal intensity on T1, high signal intensity on T2 and STIR, and normal appearances of the cortex, medullary cavity, and adjacent soft tissues (4). The most common differential diagnosis is osteochondroma, due to the similar surface location and cartilaginous component (4). Other differentials include subungal exostosis, parosteal osteosarcoma, and heterotopic chondro-ossification. Despite being a distinct imaging entity, radiologists still confuse BPOP with a variety of other benign and malignant lesions due to reported variable patterns. Ryback et al. described a series of four histologically confirmed Nora's lesions with cortico-medullary continuity similar to osteochondroma, and Rushing et al. presented a case of a BPOP causing heel pain mimicking plantar fasciitis and osteosarcoma (5, 6). For these reasons, diagnosis remains histological. On histology, BPOP demonstrates three distinct components with variable degrees of representation: hypercellular cartilage with calcification and ossification, with the calcified cartilage having a characteristic basophilic tinctorial quality ("blue bone"); cancellous bone undergoing maturation; and spindle cell stroma without cytologic atypia (7) (Figure 1).

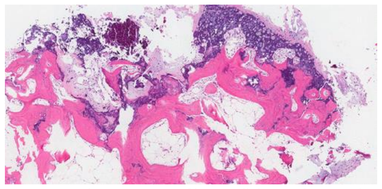

Figure 1. Specimen demonstrating irregular maturation of hypercellular cartilage into bone, producing bone with the characteristic blue quality ("blue bone") (from Zhang et. al. 2017)

Similar to the variability in imaging presentation, there have been reports of variable histology, particularly cellular atypia in a confirmed benign BPOP, leading to new studies exploring the role of cytogenetic testing with promising translocation breakpoints for diagnostic confirmation (8). Due to the benign nature of the lesion, the mainstay of treatment remains local resection. However, local recurrence rates are reported between 20-50%, with some case series reporting up to three recurrences of the same lesion in the same patient (1, 2, 3). Despite the high rate of recurrence and similar characteristics to malignant lesions, there is currently no documented cases of tumor transformation, malignant or otherwise.

## Case Study

A 52 year old female presented to the clinic in April 2018 with a painful, rapidly growing mass of the plantar lateral aspect of the right 2nd proximal phalanx. She had noticed progressive swelling and discomfort over the past few months that had worsened just prior to presentation due to abutment with the 3rd digit. Of note, patient had a history of an osteochondroma of the same site, which had been resected by another surgeon of the same practice without complication 10 years prior in February 2008. Patient could not recall any precipitating event or trauma. The remainder of her history and physical exam was within normal limits.

Radiographs of the right foot revealed a mixed sclerotic and lytic exophytic bone lesion arising from the lateral aspect of the base of the right 2nd proximal phalanx (Figure 2). The entire contour of the lesion was appreciated and was more sclerotic proximally near the base of the bone and lucent distally. MRI revealed a 2.4 x 1.1 x 1.4 cm heterogeneously enhancing mass with a broad bony cortex base with exuberant osseous proliferation, no definite involvement of the medulla, and surrounding soft tissue enhancement (Figure 3). Differential diagnoses at that time included BPOP, parosteal osteosarcoma, or atypical osteochondroma with malignant transformation. For this reason as well as the patient's history of prior tumor at the same site, the case was discussed with an orthopedic oncologist and pathologist, and it was decided to proceed forward with an incisional bone biopsy.

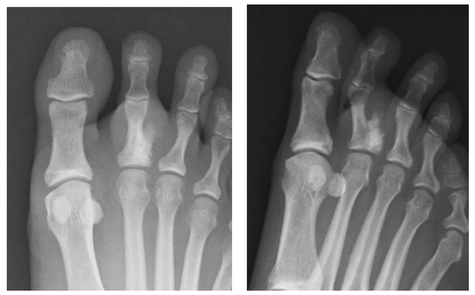

Figure 2. A) AP view. B) Medial oblique view.

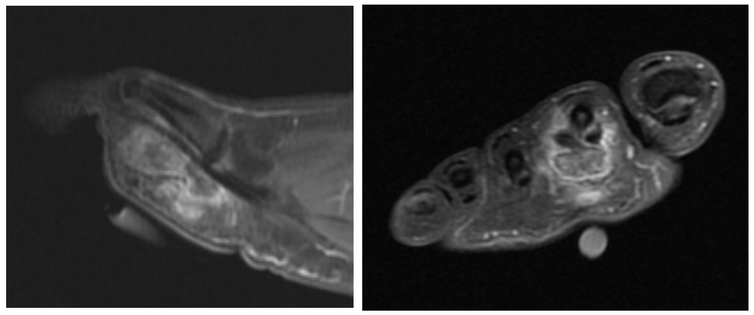

Figure 3. A) Sagittal T1 FS. B) Coronal T1 FS.

Incisional bone biopsy of the lesion in May 2018 was performed through a lateral skin incision of the 2nd digit aided by intraoperative fluoroscopy. Pathology report revealed an irregular proliferation of actively remodeling woven and lamellar bone with thickened trabeculae, lined by osteoclasts and osteoblasts with a fibrovascular stroma and transition to fibrocartilage at one end with no cytologic atypia, consistent with a diagnosis of BPOP. The same pathologist was able to review the report and slides from the resected tumor 10 years prior and confirmed the diagnosis of osteochondroma at that time.

## Case Study Continued

Pathology report was discussed with the patient at follow up, and surgical options of local resection vs. digital amputation were discussed. Patient decided to proceed with local resection which was performed in June 2018 through the same incision with resection of half the proximal phalanx (Figure 4). The post-operative course was uncomplicated and patient returned to normal shoe gear with complete resolution of her symptoms.

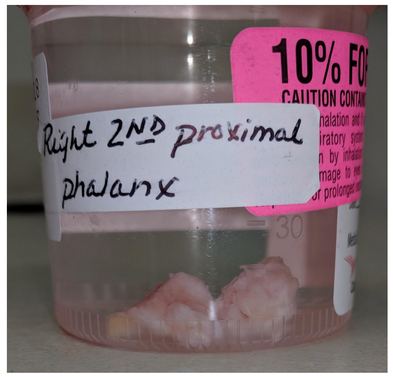


Figure 4. Complete tumor resection with a margin of normal proximal phalanx

## Discussion

Nora's lesion, is a rare, benign bone tumor and diagnostically challenging etiology of pain and deformity in the podiatric population. Imaging variability, sometimes rapid growth, atypical histological appearance, and a high recurrence rate, combined with an unfamiliarity with the pathology confounds the diagnosis of BPOP, raising concerns for malignancy, leading to further often unnecessary workup. Although a high incidence of local recurrence exists, tumor transformation, malignant or otherwise, of BPOP has not been previously reported. This case report illustrates a novel incidence of transformation of an osteochondroma to a Nora's lesion. Despite the variability and often challenging presentation, BPOP remains a treatable benign process. Awareness and increased reporting of BPOP can improve diagnosis and thus effective and timely treatment of this rare entity.

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