Atypical Radiological Presentation of a Bizarre Parosteal Osteochondromatous Proliferation (Nora’s Lesion) Affecting the Index Finger: A Case Report

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ABSTRACT

We are reporting a rare case that presented to our hand and upper limb surgery at Royal Rehabilitation Center at KHMC of Bizarre Parosteal Osteochondromatous (BPOP) The importance of this case lies in the fact that most BPOP lesions in the literature have been described to have typical radiological appearance, but few describe the atypical presentation. Our case report also highlights the need to make a staging system of the disease using a combination of plain radiograph, MRI and clinical features.

Keywords: Atypical radiological presentation, bizarre parosteal osteochondromatous proliferation, Nora’s lesion,

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Introduction

Bizarre parosteal osteochondromatous proliferation (BPOP) also known as Nora’s lesion, is a rare, benign exostotic osteochondromatous tumor of the hands and feet. Until now, around 200 cases of Nora’s lesion have been described in the literature. It presents as a painless swelling in soft tissues of the hand or feet. The most common site is adjacent to proximal phalanx of metacarpal or metatarsal bones. Similar lesions have been reported in radius, ulna, femur, tibia, nasal bone and skull. Patients’ ages range between 8 and 73 years although most patients are between 20 and 40 years. About 20% of patients have a history of trauma. Histologically, BPOP consist of irregular aggregates of hyaline cartilage, spindle cells, and new bone. BPOP is treated with surgical excision. These lesions have a remarkable tendency to recur, recurrence rates between 29% and 55% in a 2-year interval have been reported, and almost half of those patients have had a second recurrence.

Case Report

A 28-year-old man was referred to hand surgery clinic at Royal Rehabilitation Center at King Hussein Medical Center with a history of an enlarging mass in the left index finger. A radiograph was performed on the first visit and was unremarkable (Figure 1). Left hand and wrist MRI was performed and showed a 1.8x1.0 cm mass, hypointense in Table I and Table II weighted images and in close relation to the underlying tendons reaching the cortex but not invading it (Figure 2). Appearance was highly suggestive of giant cell tumor of the tendon sheath; MRI study was reported by DR. Jameel Shawaqfeh. Marginal excision of the mass was done and sent for histopathology. Histopathology of the specimen revealed multiple fragments of a lesion showing irregular maturation of cartilage containing enlarged bizarre...
binucleated chondrocytes into bone consistent with bizarre parosteal osteochondromatous proliferation (BPOP) (Figure 3). After 3 months, the mass re-appeared after excision Figure 4 but this time with its typical radiological presentation Figure 5 i.e., radiodense mass with intact underlying surface, and the patient underwent En-bloc negative margin excision (i.e. excision of the pseudocapsule over the lesion plus periosteal tissue beneath the lesion with decortication of any areas in the underlying bone that was abnormal).

**Discussion**

BPOP is a benign but locally aggressive tumor that has clinical similarities with osteochondroma and periosteal chondroma. In addition to their similar clinical presentation, these tumors can also have similar radiographic and histological appearances. On X-ray, BPOP appears as a well-circumscribed radio-dense mass arising from the cortical surface. The underlying cortex is always intact but occasionally shows slight surface irregularity. The radiological appearance of BPOP is dependent on the stage of evolution of the lesion. The first stage presents as a periosteal soft tissue swelling with or without the presence of a small amount of calcifications. In later stages the soft tissue mass progressively calcifies and finally ossifies. It usually takes about 6 months to make this classic evolution from soft tissue mass to ossified mass. X-rays alone are sufficient to diagnose BPOP in typical radiographic appearance and typical clinical findings. In our case, plain radiographs were initially unremarkable but later on started to develop this classic appearance and this made the diagnosis of BPOP lesion less likely at first presentation. Taking these concepts into account, histopathology becomes an even more important diagnostic tool for this disease.

**Conclusion**

We report a case of BPOP with atypical radiological presentation with diagnosed based on findings from postoperative histopathology. Due to local recurrence rates and its rarity, the Nora lesion will continue to be having a challenge for orthopedic surgeons. Additionally, at the present time, there is no standardized staging system or treatment regimen. Therefore, treatment and follow-up care should take place in specialized centers.

![Fig 1: anteroposterior and lateral radiograph at first presentation showed no calcification around the middle phalanx of index finger](image-url)
**Fig. 2A&B**: Hand MRI showing a 1.8x1.0 cm mass, hypoitense in T1 and T2 weighted images and in close relation to the underling tendons reaching the cortex of the middle phalange of the index finger but not invading it. Appearance was highly suggestive of giant cell tumor of the tendon sheath.

**Fig. 3A&B**: Histopathology slide of the specimen revealed multiple fragments of a lesion showing irregular maturation of cartilage containing enlarged bizarre binucleated chondrocytes into bone consistent with Bizarre parosteal osteochondromatous proliferation (BPOP).

**Fig 4**: hand photograph 3 months postoperatively showing mass reappeared from left index with old surgical scar over it.
Figure 5 A&B: anteroposterior and lateral radiograph of the hand at 3 months after surgical excision showing calcified mass near the middle phalanx of index finger not attached to underlying cortex

References