

### **Bizarre parosteal osteochondromatous proliferation (Nora's lesion) of the phalanx**

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**B**izarre parosteal osteochondromatous proliferation (BPOP) or Nora's lesion, named after the pathologist who described it in 1983, is a rare tumorous lesion with an aggressive growth pattern that affects primarily the small tubular bones of the distal extremities and often recurs after excision. There were 35 Nora's lesion reported cases emerging from the bones of the hands and feet. The largest published series of 65 cases, with 17 lesions involving the long bones of the upper and lower extremities.<sup>1</sup> With this exception, most authors found isolated cases, Age ranges from 8-74 years, however, most patients are between 20-35 years of age, with the lesion affecting males and females in equal proportions. Usually, the presenting symptoms are due to bony mass, with a variable growth rate (months-years) and infrequent pain. In 15-20% of cases, there was trauma in the patient's history. Evidence of radiologic demonstration of temporal development of bizarre parosteal osteochondromatous proliferation supports the theory that BPOP is caused by trauma.<sup>2</sup> Since the histological findings of BPOP are similar to those of florid reactive periostitis, and subungual (Dupuytren's) exostosis, some authors have postulated that all these conditions are reactive proliferative lesions representing different phases of reactive process, and BPOP apparently arises from the periosteal tissues through a process of cartilaginous metaplasia. Whether BPOP is a reactive proliferative lesion or a neoplastic lesion, however, remained controversial until a recent translocation, t (1; 17) (q32; q21) was detected using chromosome banding and fluorescence in situ hybridization analysis. Few more studies detected, to break point translocation at t (1; 17) (q42; q23)<sup>3</sup> and at t (1; 17) (q32; q21). These results suggest that t (1; 17) is a distinct translocation of bizarre parosteal proliferation, and that BPOP is a neoplastic lesion, rather than a reactive proliferative process, and further studies are being conducted to determine whether any of these translocation are involved in the formation of a fusion gene. Further, cytogenetic analysis on Dupuytren's exostosis of great toe, identified a balanced translocation t (X; 6),

suggesting that Dupuytren's exostosis and BPOP are the distinct clinicopathological entity with a different molecular pathogenesis.

A 27-year-old Egyptian patient of BPOP, mechanic by profession, was presented to our hospital with a painful bony swelling in the left middle finger of one year duration. Plain radiographs of the lesion and other routine relevant investigations were carried out, and a provisional diagnosis of exostotic bony lesion was arrived. On gross examination, an excision biopsy of the lesion showed a 0.6 cm thick cartilage capped bony mass measuring 1.6 x 1.3 x 0.8 cms (**Figure 1**).<sup>1</sup> On microscopic examination of the lesion, we observed a differential diagnosis of traumatic osteochondroma and bizarre osteochondromatous proliferation. The patient developed recurrence of similar lesion at the same site after one year. On the following radiological investigation, an excision biopsy was carried out, and the specimen subjected to histopathological examination, which confirmed the diagnosis of bizarre osteochondromatous proliferation.

The following specific radiological findings help to distinguish BPOP from many of its mimickers including solitary osteochondroma: 1. The tumor appears to arise from the surface of the cortex directly. 2. There is no continuity between the central part of the tumor and the medulla of the underlying bone. 3. There is no flaring of the cortex of the underlying bone.<sup>4</sup> The absence of continuity between the central part of the lesion with the cortex and medulla of the parent bone can be well-demonstrated on computed tomography images. Isotope bone scans have shown intense tracer uptake in the lesion, and



**Figure 1** - Gross picture of the excised specimen showing shiny pearly white cartilage capped bony mass measuring 1.6 x 1.3 x 0.8 cms, with cartilaginous cap measuring 0.6 cms.

## Clinical Notes

the appearance progresses to a more characteristic mature bone architecture. Magnetic resonance images of the lesion revealed that the tissues within the lesion have signal characteristics that differ sharply from the normal bone and cartilage. The lesions are of low signal intensity on T1 weighted sequences, and on fast spin echo T2 weighted and short-tau inversion recovery sequences, the lesions are of high signal intensity, which is a typical pattern of a neoplastic lesion.

Histologically, BPOP has 3 distinct components with variable degree of representation: 1. Hyper cellular cartilage with calcification, with the calcified cartilage having a characteristic basophilic tinctorial quality. 2. Cancellous bone undergoing maturation. 3. spindle cell struma without cytological atypia. The most important lesions that present differential diagnostic problems are chondrosarcoma, parosteal osteosarcoma, and florid reactive periostitis. However, with radiological and clinical findings with appropriate history, supplemented with histopathological examination can help guide the therapy and improving patient management, as BPOP has a high recurrence rate of 51% initial recurrence, 22% second recurrence. Probably, our case had the first recurrence. The recommended treatment of choice is excising the pseudo capsule over the lesion, any periosteal tissue beneath the lesion, and decorticating any abnormal appearing areas in the underlying host bone, as this procedure has less recurrence rate than simple excision of the lesion.<sup>5</sup>

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