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CASE REPORT

A Recurrent Case Report of Bizzarre Parosteal Osteochondromatous Proliferation (Nora Disease) and Literature Review

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Introduction

Bizarre parosteal osteochondromatous proliferation (BPOP), as defined by Nora and colleagues [1] in 1983 (also called Nora lesion), is a rare lesion. About 160 cases of BPOP have been presented in the literature until now. The lesion is an outgrowth from the cortical surface consisting of bone, cartilage and fibrous tissue. It usually affects the proximal and middle phalanges, and the metacarpal or metatarsal bones. The hands are 4 times more commonly affected than the feet [2]; however, lesions in the long bones, skull, maxilla and metatarsophalangeal sesamoid have been reported [3-5]. The lesion affects patients of any age, but most are in their 20s and 30s with no sex predilection [6]. The benign lesion of the bone might be mistaken for malignant processes because of the high frequency of recurrence, the occasional quick growth and atypical histologic appearance. Along with a review of the literature, we discuss a patient with this rare lesion to illustrate the histologic, radiologic and clinical features as well as the different etiologic theories on BPOP.

Case Reports

A 30-year-old man, presented to our outpatient clinic with pain and a tender palpable mass at the middle phalanx of his right index finger (Figure 1a). He reported a recent increase in the size of the mass, and the pain was unremitting and related to touching or rubbing on. He denied any history of injury but he has a habit of chewing his right index finger for the past a few years when he got in nervous situations.

The clinical and imaging findings were characteristic of BPOP. We performed an incisional biopsy for him to confirm the diagnosis. In this case, radiographs showed calcified and osseous masses adjacent to the affected middle phalanx. The underlying bone had no cortical flaring or structural changes. The radiographs showed intensely calcified and ossified masses with well-defined margins. There was no continuity with the medullary canal of the bones from which the masses originated. The adjacent soft tissue in normal appearance, in keeping with the diagnosis of BPOP (Figure 2). Three months after excisional biopsy, recurrent lesion was seen on index same finger and radiographs in the same size with similar features of the initial mass over middle phalanx of the index finger (Figure 3a, Figure 4 and Figure 5).

The MRI scans showed that the masses were isointense lesions with muscle on T_1-weighted images.

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On T_2-weighted images, most area of the lesion had high intensity, and mixed with area showed heterogeneous intensity either in initial or recurrent images of the MRI. MRI T2- weighted images showing high signal mixed with a low-signal lesion extending from the middle phalanx of the index finger (Figure 6 and Figure 7).

In summary of imaging data of this 30-year-old man with bizarre parosteal osteochondromatous proliferation at the middle phalanx of his right index finger (Figure 1a and Figure 3a) Anteroposterior and lateral radiographs showing a calcified mass (arrow) on the middle phalanx of his right index finger without alteration of the underlying cortex. There is normal signal intensity of the cortex and the bone marrow of the underlying bone (Figure 2, Figure 4 and Figure 5). The anteroposterior and lateral radiographs displayed a similar feature on local recurrence 9 months after initial index excision surgery (Figure 5).

Histologic Findings

In our patient, the excised specimen was well circumscribed masses (Figure 1b and Figure 3b). In cross-section, the lesions consisted of a cartilage cap and bone tissue. This structure seemed to correspond with the MRI findings. Histologically, the superficial area of the masses showed fibrocartilaginous tissue with high cellularity. Spindle shaped or stellate small chondrocytes were scattered in a myxoid stroma. The cells varied in size, and some were binucleated. The basal area was composed of immature bony trabeculae

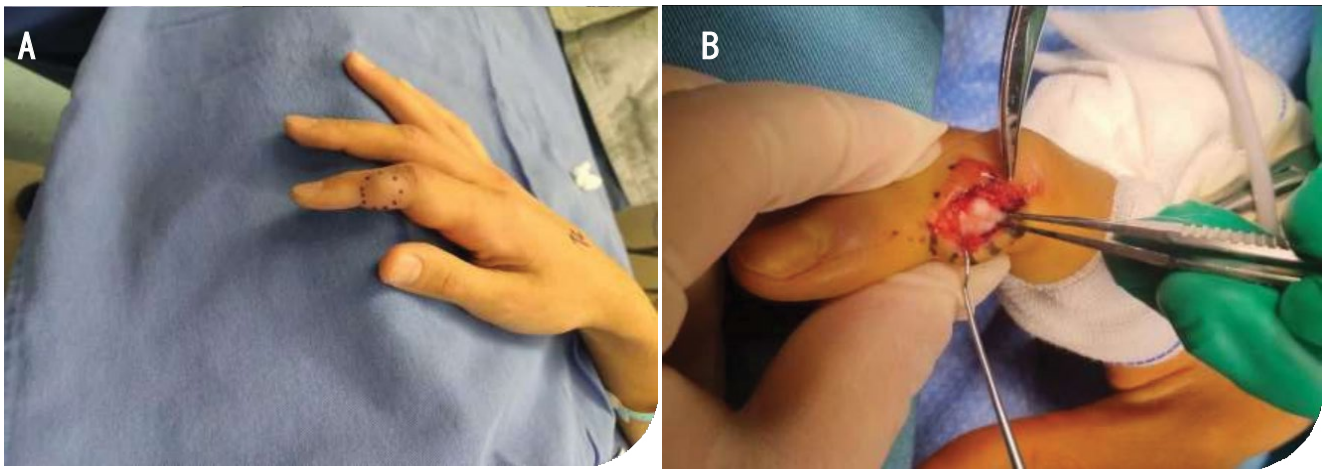

Figure 1: Photographs showing a mass on radial side middle phalanx of the Index finger of right hand (A); and index surgery of intraoperatively findings of the mass (B).

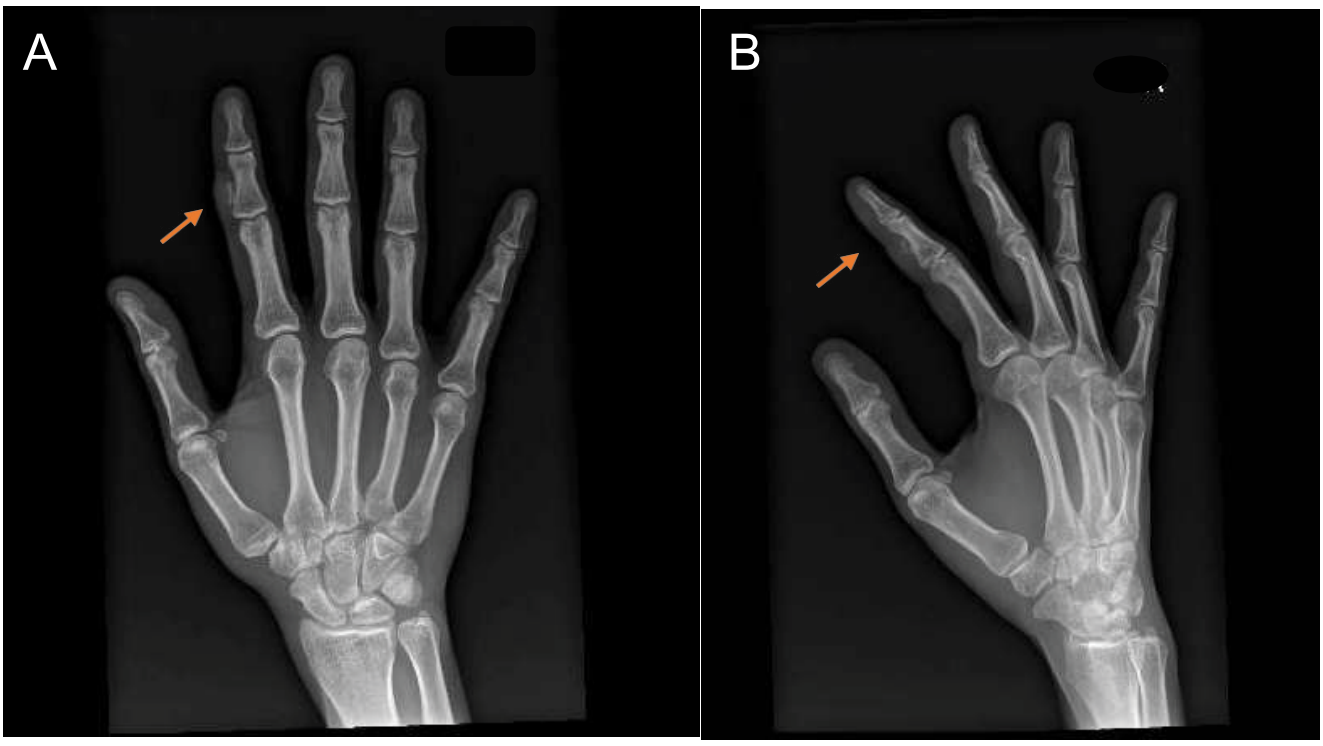

Figure 2: Initial AP oblique radiographs showed a calcified lesion over radial side of middle phalanx (arrows).

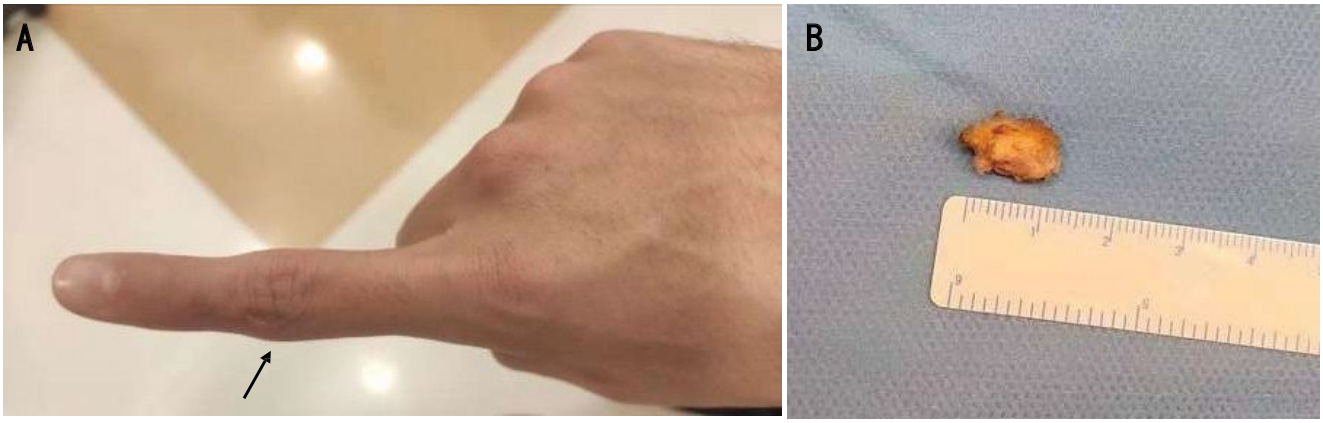

Figure 3: Nine months after index surgery, recurrent mass over the radial side of the middle phalanx of index finger seen (an arrow) (A); Surgical excision of the mass was shown in size of 15 mm × 10 mm × 4 mm solid mass (B).

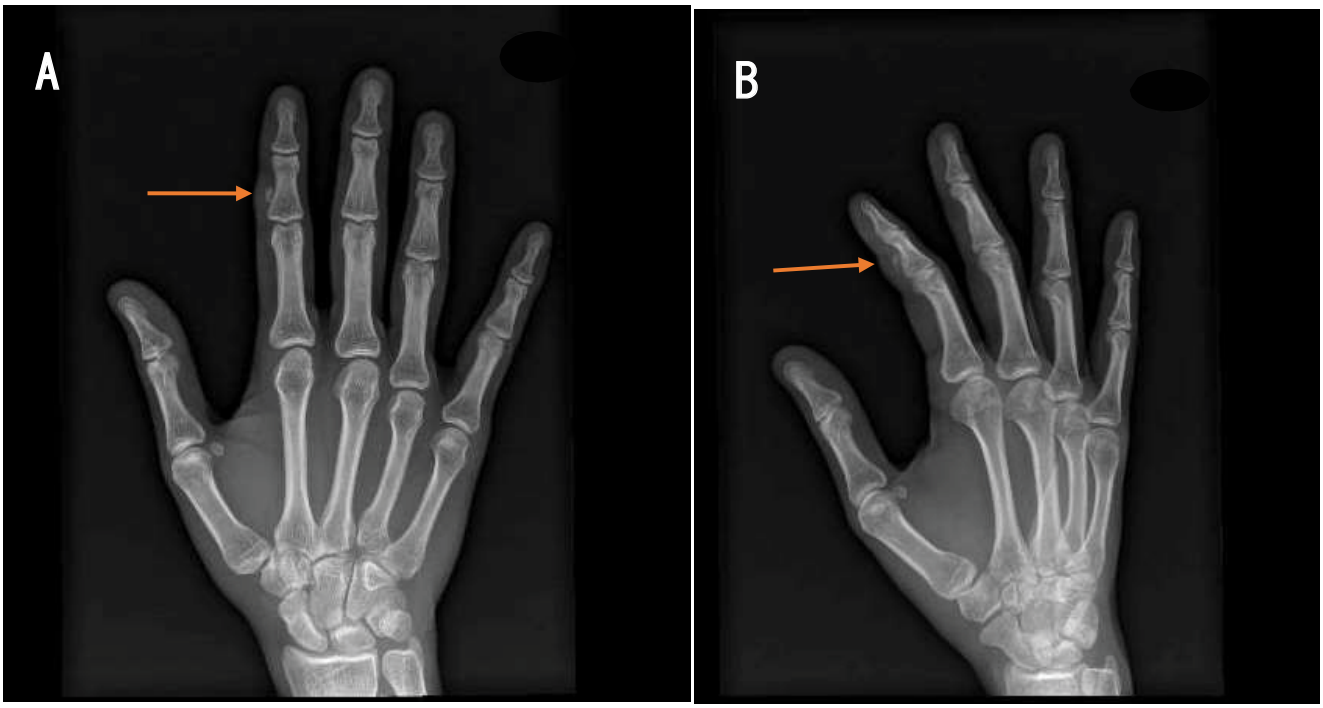

Figure 4: Three months post index surgery, AP (A) and oblique (B) radiographs showed the similar lesion at the same site and similar morphology.

with high osteoblastic activity. These formed trabeculae stained mostly deep blue with hematoxylin and eosin ("blue bone"). The spindle cells were arranged loosely among the trabeculae, which were apparently formed by a process of enchondral ossification. The cells showed neither atypical mitoses nor cytological atypia (Figure 8a, Figure 8b, Figure 8c and Figure 8d).

Clinical Results

Our patient experienced local recurrence; it occurred 3 months after excision. He experienced progressively enlarge mass with minor pain while touching on. At his most recent follow-up 12 months after surgery, he was free of pain and had no limitation of motion in his index finger, and the radiographs showed clear on radial side of index finger (Figure 9). In this case, we excised the pseudo capsule over the cartilage cap (Figure 1b) and any periosteal tissue beneath the lesion, and curettaged superficial cortices of the tumour bed. 12 months after the second excision, he was free of any evidence of local recurrence. The local recurrence had histology identical to that of the primary lesion.

Discussion

Bizarre parosteal osteochondromatous proliferation (BPOP) is an uncommon reactive mineralizing mesenchymal lesion that typically affects the surfaces of bones in the hands and feet, usually the proximal and middle phalanges, and the metacarpal and metatarsal bones [7]. 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

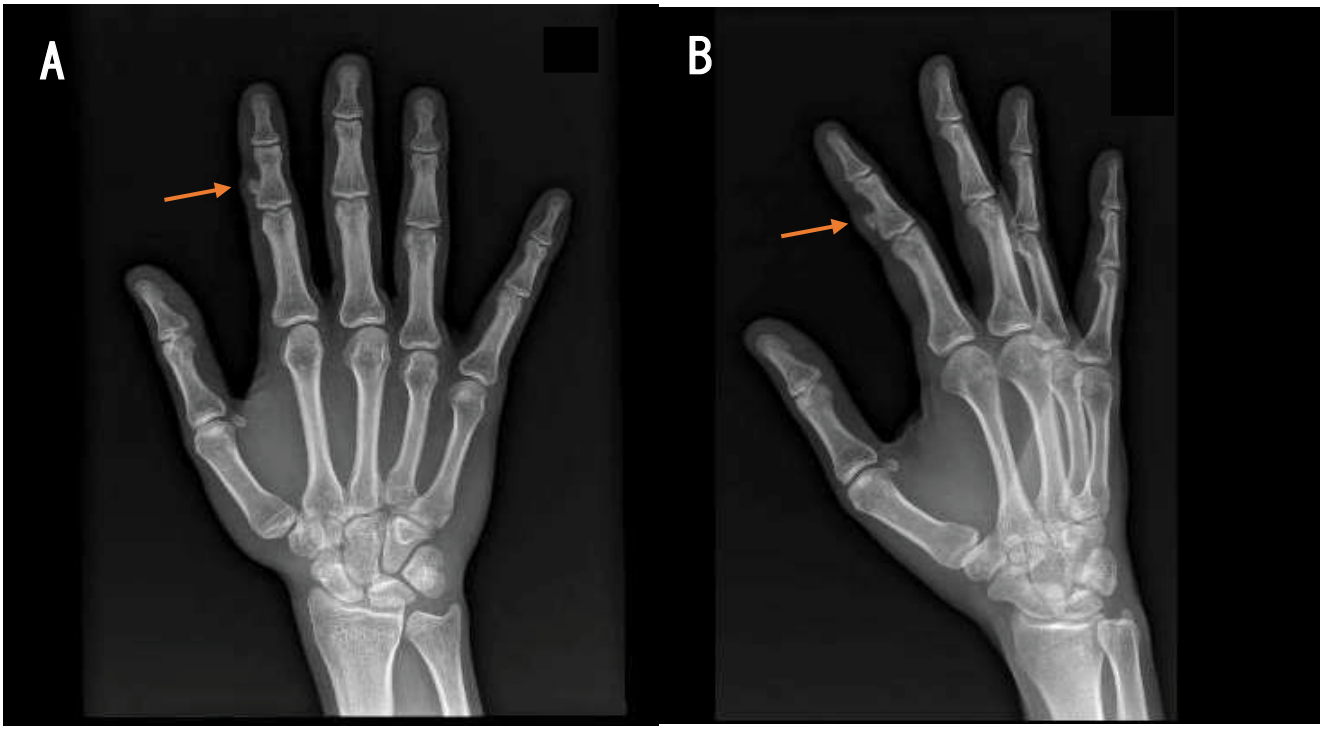

Figure 5: Ten months post index surgery, radiograph showed the similar lesion at the same site with increased calcification.

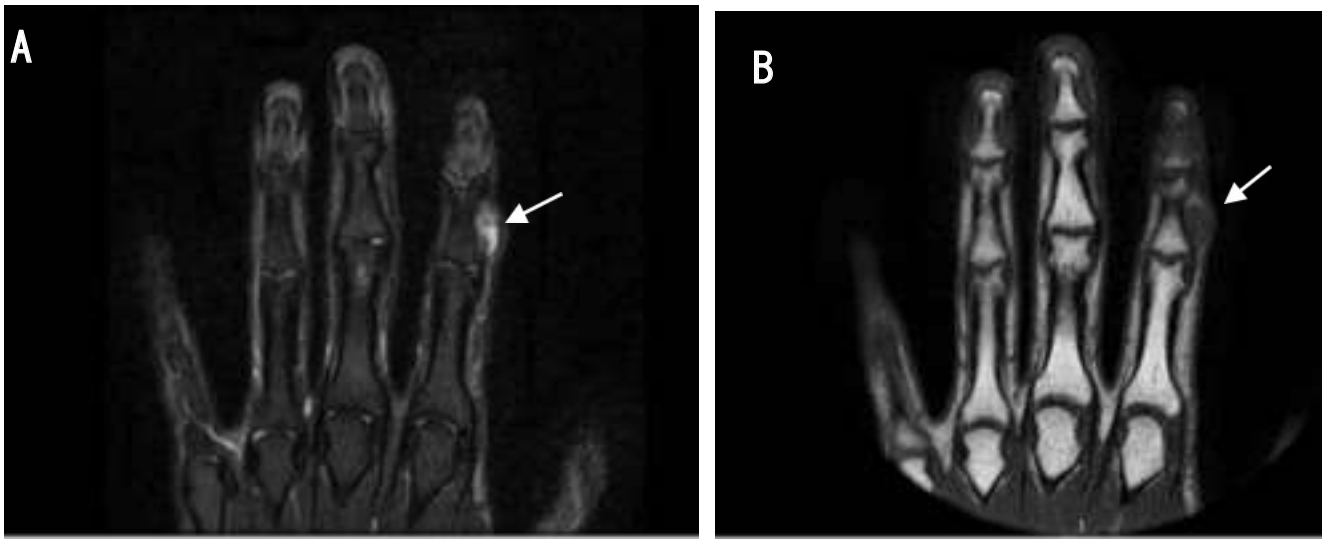

Figure 6: Preoperative index surgery, MRI showed T2-weighted magnetic resonance images (coronal) fat suppression, showing a high-signal lesion mixed with low signal calcification (arrow) extending from the middle phalanx of the index finger (A). T1-weighted MRI fat suppression sequence (coronal) showing a low-signal lesion (arrow) extending from the middle phalanx of the index finger (B) indicating rich-cellular components of the lesion.

patients have had a second recurrence [1,5,7]. Nora and colleagues [1] presented 35 cases of BPOP with 18 (51%) local recurrences. Meneses and colleagues [5] reported a recurrence rate of 55% in a series of 65 patients, and Dhont and colleagues [7] reported a recurrence rate of 29% in 24 patients. However, despite a high tendency to recur and a sometimes atypical histologic appearance, no malignant transformation, metastases, deaths or associated systemic diseases have been described so far in patients with BPOP [8].

Although BPOP has a characteristic clinical and histologic appearance, it may be confused with other benign and malign lesions. Owing to the parosteal location, BPOP must be distinguished from parosteal osteosarcoma, which is rarely found in the hands and feet [9]. The absence of cellular atypia helps to distinguish this lesion from osteosarcoma. The lesion might be mistaken for osteochondroma because of its surface location and cartilaginous component. Osteochondromas are extremely uncommon in the small bones of the distal extremities [2]. They show the typical continuity with the medullary canal and the cartilage does not show any signs of atypia. Rybak and

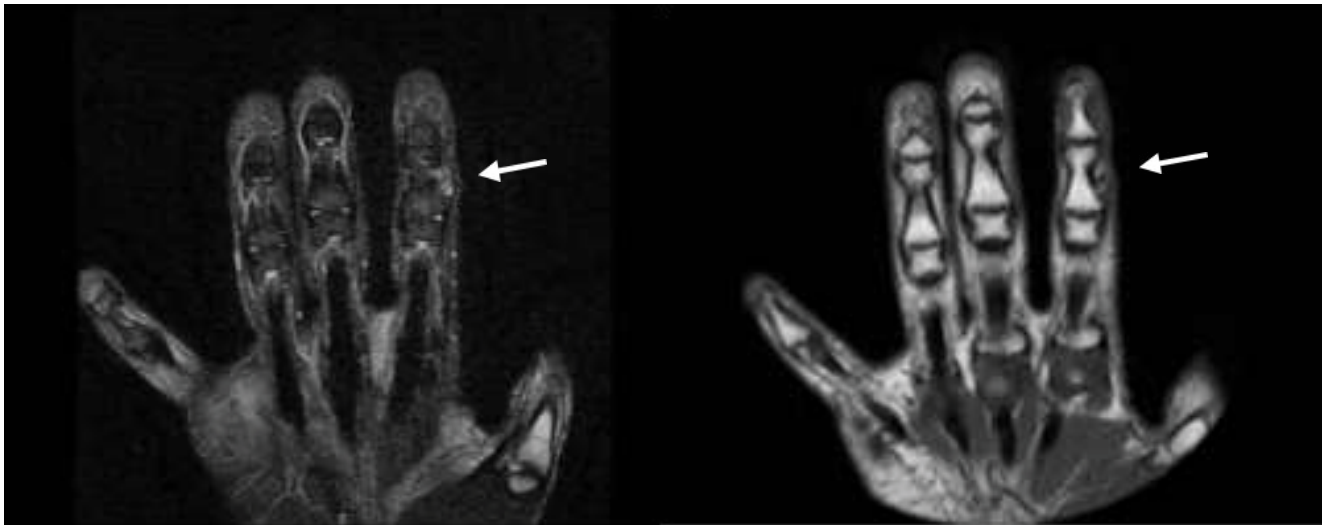

Figure 7: MRI prior to second surgery showed T2-weighted magnetic resonance images (coronal) fat suppression, showing less high-signal lesion mixed with low signal calcification (arrow) extending from the middle phalanx of the index finger (A). T1-weighted MRI fat suppression sequence (coronal) showing a low-signal lesion (arrow) mixed with low signal lesion (B) indicating less-cellular components of the lesion in recurrent lesion.

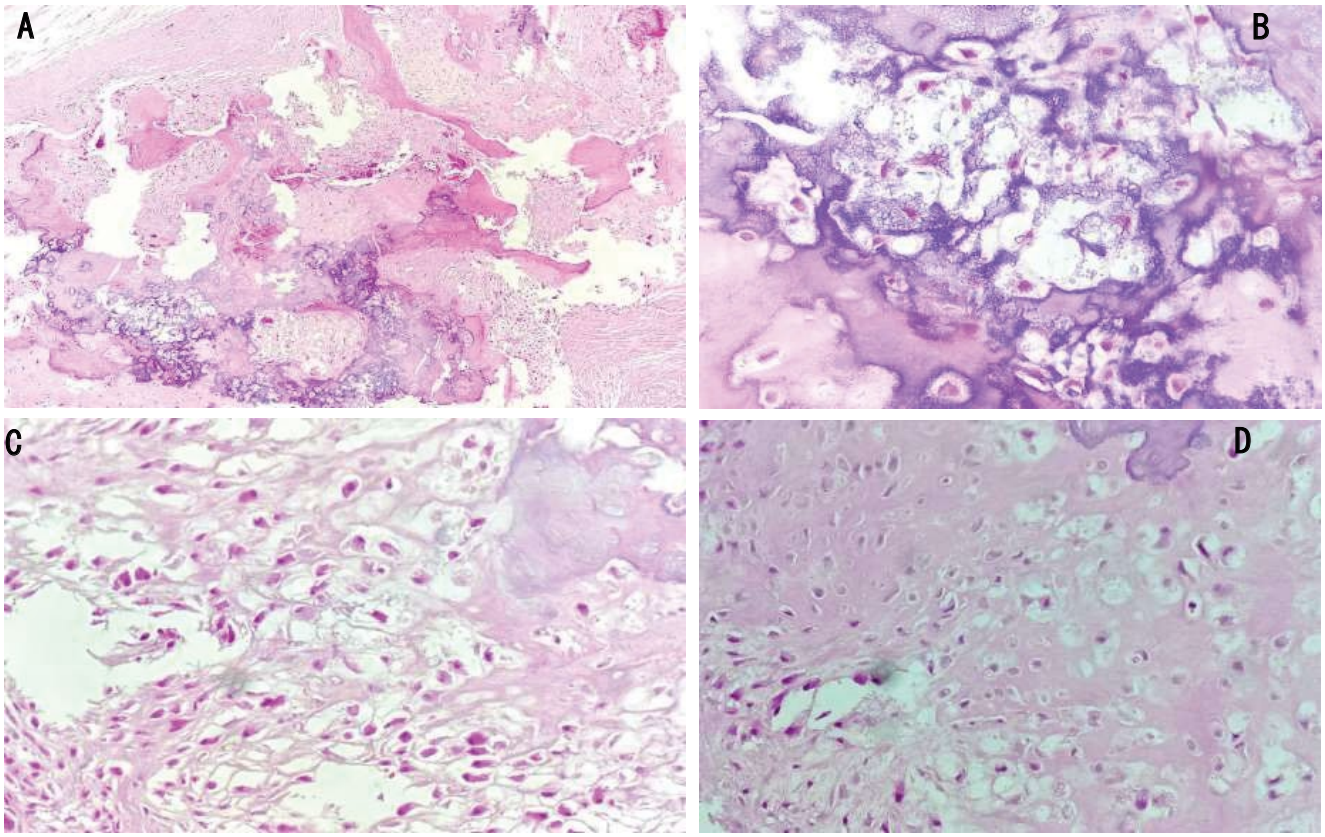

Figure 8: (A) Low power field revealed the lesion consisting of a cartilaginous cap and a poorly developed disorganized zone of enchondral ossification (hematoxylin and eosin stain; original magnification x100); (B) High power field revealed irregular maturation of cartilage in bone produces chondro-osteoid with characteristic blue quality (“blue bone”). Contains enlarged, bizarre, binucleated chondrocytes with matured bone; (C) Spindle cell proliferation between trabeculae bones without atypia but contained myxoid stroma; (D) High power view showed enlarged, bizarre, binucleated chondrocytes with matured bones.

colleagues [10] presented the cases of 4 patients with pathologically proven BPOP in which cortico-medullary continuity with the underlying bone was demonstrated on imaging. The absence of such a communication has been singled out as a critical imaging feature of BPOP. Rybak and colleagues [10] indicated that BPOP could not be identified by radiologic features alone. Histopathologic examination is the best method to identify this lesion and should be performed for definite diagnosis.

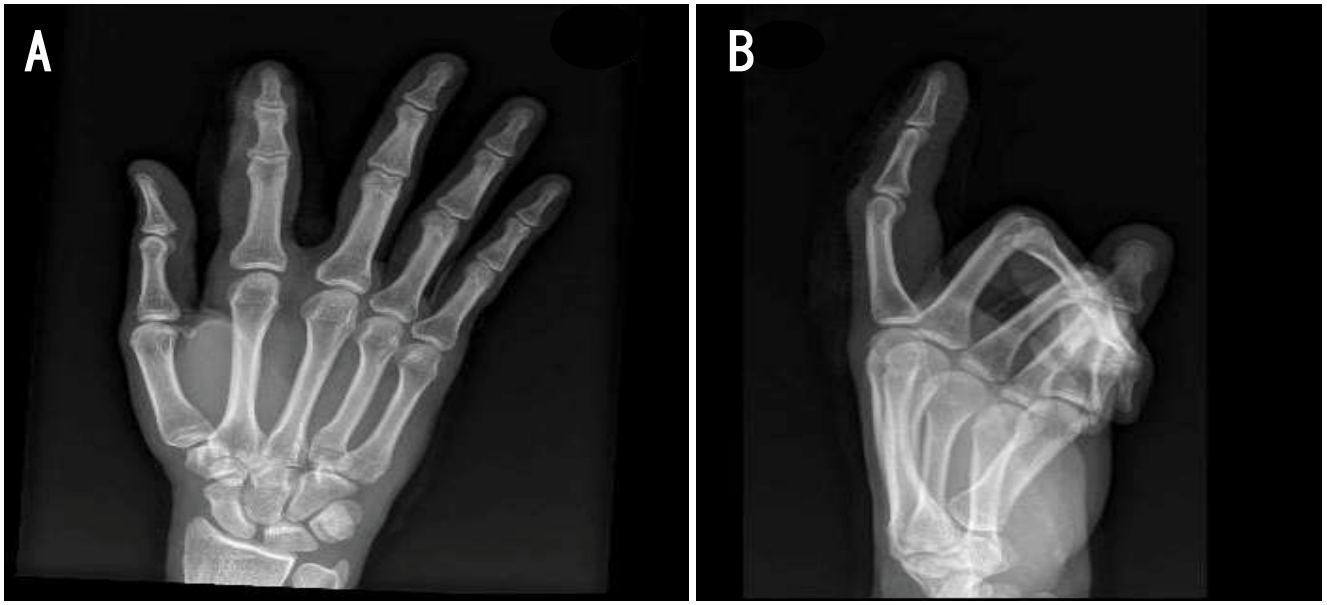

Figure 9: Eleven months after index surgery, re-excision of the lesion with a thin decortices was performed. Radiographs showing completely excision of the lesion a year after excision of recurrent lesion (Figure A and B).

Other benign, non-neoplastic lesions like periostitis ossificans may also simulate BPOP. This florid, reactive periostitis affects the bones of the hands in most patients, although other parts of the skeleton cannot be excluded. Turret exostosis is a dome-shaped parosteal bone proliferation located on the dorsal aspect of the phalanges. It has been proposed that BPOP, florid periostitis and turret exostosis are all part of the same lesion spectrum [11,12]. The lesion may represent an intermediate lesion between florid reactive periostitis and turret exostosis. Florid reactive periostitis may progress to BPOP, as described by Dorfman and colleagues [11].

Horiguchi and colleagues [6] report the expression of basic fibroblastic growth factor in nearly all chondrocytes: chondromedulin-I in the tissue of the cartilaginous cap and vascular endothelial growth factor only in the large chondrocytes near the osteocartilaginous interface of the lesion. Their findings suggest that the processes occurring in the cartilaginous cap of BPOP are similar to those of enchondral ossification in the growth plate, concluding that BPOP is a reparative process after periosteal injury. Immunohistochemical and molecular analysis strengthened this assumption which is more likely in our case who has a habit chewing his right index finger for the past a few years when he was in nervous. However, most patients do not report a history of previous trauma. Moreover, if BPOP is a reactive lesion, its remarkable tendency to recur after excision is difficult to explain. Orui and colleagues [13] reported the case of 1 patient with BPOP that occurred 2 years after bilateral leg erythema nodosum. Systemic or focal inflammation might have been responsible. Zambrano and colleagues [8] presented the cases of 3 patients with subungual (Dupuytren) exostosis and of 2 patients with BPOP. Their findings of consistent chromosomal rearrangements indicate that BPOP is a neoplastic, rather than reactive, process. The cytogenetic analysis of 5 patients with BPOP by Nilsson and colleagues [14] showed a balanced translocation t (1;17) (q32;q21). To investigate the specificity of this reciprocal translocation, they screened the karyotypes of more than 43 000 neoplasms and found no identical translocation. It seems to be a recurrent and pathogenetically significant aberration in BPOP. Endo and colleagues [15] described the case of a 39-year-old woman with BPOP arising in the proximal phalanx of her third toe. Their cytogenetic analysis is comparable with the findings of Nilsson and colleagues [14]. The occurrence of a translocation, as mentioned previously, supports the assumption that a neoplastic process may be the etiologic agent.

The true prevalence of BPOP is difficult to assess because most lesions are reported in case studies [2-4,6,8,10,14-16] and because larger, mostly histologic studies [1,5,7] are retrospective. Therefore, further work is needed to fully elucidate the etiology of BPOP.

Excision is the recommended therapy of symptomatic BPOP. Intralesional excision seems to have a great potential for local recurrence, but it preserves stability without deep decortication of the affected bone. En bloc negative margin excision by the excision of the pseudo capsule over the lesion and any periosteal tissue beneath the lesion and the decortication of any areas in the underlying host bone that appear abnormal has been shown to be beneficial in preventing local recurrence [16]. Wide resection could possibly lead to segmental amputation because of the anatomic conditions in the long bones of the fingers and toes, and it cannot be recommended as first-line surgical treatment.

Owing to high local recurrence rates and a lack of

adjuvant therapy options, the Nora lesion will continue to pose a challenge for orthopedic surgeons and clinical research. Therefore, surgical excision and follow-up carefully is compulsory.

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