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Case Report

Nora's Lesion in the Proximal Phalanx, Diagnostic Dilemma and Treatment-Case Report and Review of Literature

Abrar S Hashishi[1], Abdullah Al Mulhim[2], Nasreen Mohamed[2], Fouad AlAdel[3] and Emad Al Absi[1]*

[1]*Department of Orthopedic Surgery, King Fahad Specialist Hospital, Saudi Arabia*

[2]*Department of Pathology and Laboratory Medicine, King Fahad Specialist Hospital, Saudi Arabia*

[3]*Medical Imaging Department, King Fahad Specialist Hospital, Saudi Arabia*

***Corresponding Author:** Emad Al Absi, Department of Orthopaedic Surgery, King Fahad Specialist Hospital, Saudi Arabia.

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Abstract

Nora's lesion, also known as "bizarre parosteal osteocartilaginous proliferation" (BPOP), was first described in 1983 by the pathologist Nora. It is a rare, reactive, mineralizing mesenchymal lesion that typically affects the surfaces of bones in the hands and feet. We are reporting a case of 43-year female patient with left hand middle finger swelling and pain for nine months. X-ray and MRI showed aggressive looking lesion in the phalanx and the pathology was not diagnostic in the beginning, it shows osteogenic lesion suspicious for osteosarcoma. So, the pathology specimen was sent to a referral center overseas till the diagnosis of bizarre parosteal osteochondromatous proliferation was confirmed. Tumor was resected and decortication of the phalanx was done. Patient recovered smoothly and had excellent function post-surgery and no signs of recurrence in two years follow up.

Keywords: *Bizarre Parosteal Osteocartilaginous Proliferation; Phalanx Tumor; BPOP; Nora's Lesion; MRI: Magnetic Resonance Image; CT Scan; Computerized Axial Tomography*

Introduction

Nora's lesion, also known as "bizarre parosteal osteocartilaginous proliferation" (BPOP), was first described in 1983 by the pathologist Nora. This lesion is defined as a proliferation of the bone. It is one of the rare diseases of bone with a diagnostic challenge. The disease most commonly occurs in proximal phalanges, metatarsals, or metacarpals and few cases has reported to occur in skull and long bones including humerus, ulna, radius, fibula, and femur. These lesions arise in the periosteum and histologically resemble osteochondroma or osteosarcoma. We report here a case of phalangeal lesion with a diagnostic challenge and was initially diagnosed as osteosarcoma and complete healing after resection without recurrence.

Case Report

A 43-year female photographer, medically free referred to the orthopedic oncology clinic for further evaluation and management regarding left middle finger bone lesion. Patient reported history of pain and swelling for the last nine months gradually increasing till it started to affect her function and finger movement. There was no history of trauma. Her family history is significant for cancer as her mother died of liver cancer with and bone metastasis, one sister died of stomach cancer and her other sister had breast cancer. She went to a central hospital and was referred to the orthopedic oncology clinic as suspicious lesion of malignancy. On examination, left hand middle finger swelling of the proximal phalanx dorsal and volar, no hotness, tender. Distal neurovascular was intact and range of motion is markedly restricted. Systemic exam shows no lymph adenopathy or any other significant abnormalities.

Plain radiographs showed the lesion at the proximal phalanx (Figure 1A and 1B). Further evaluation by CT Scan showed a periosteal based lesion at the dorsum of the middle phalanx (Figure 1C). MRI was done and showed the lesion with soft tissue mass at the dorsum of the phalanx with low signal in T1 weighted images and heterogeneous bright signal in T2 weighted images with heterogeneous contrast enhancement and areas of breakdown. The mass causes cortical destruction of the proximal phalanx and crosses the metacarpophalangeal join with fine dorsal cortical erosions (Figure 1D and 1E). Whole body bone scan showed intense uptake of the lesion at the middle phalanx (Figure 1F).

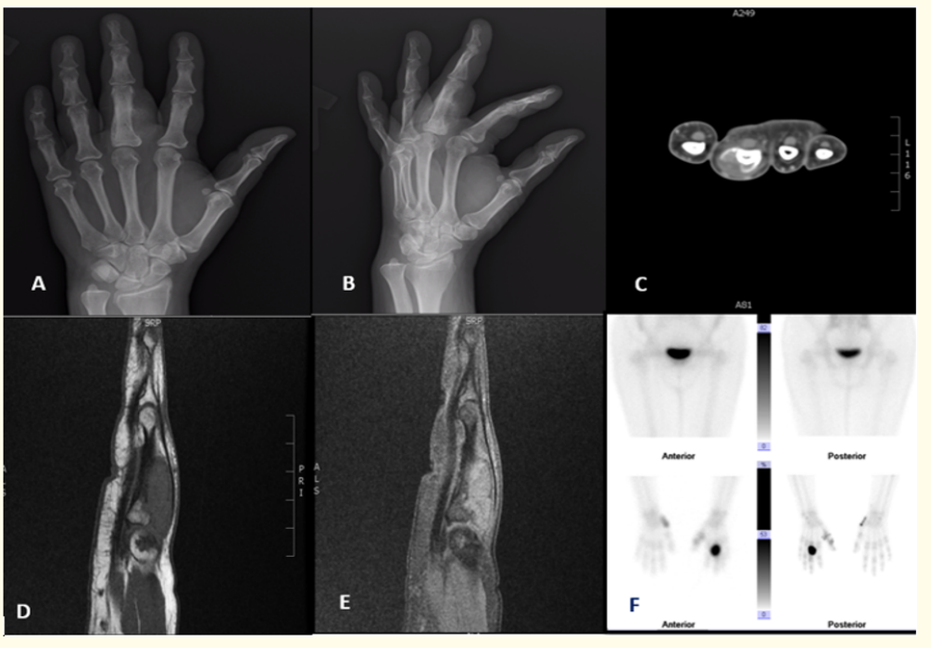

Figure 1: *(A, B): Palin radiographs of the hand shows proximal phalanx periosteal lesion in the metaphyseal and diaphyseal area with bone formation. C: CT Scan shows periosteal based lesion at dorsal aspect of the proximal phalanx with adjacent periosteal reaction causing bone erosions. (D, E): MPH of the finger shows soft tissue mass at the dorsum of the proximal phalanx with mild changes in the dorsal cortex. F: Whole body bone scan showed focal increased activity in the lesion.*

Open biopsy was done and showed osteogenic lesion and was reported as suspicious for osteosarcoma. The case sent to a referral center overseas for second opinion and the result came back as highly suggestive of bizarre parosteal osteocartilaginous proliferation "BPOP". Due to marked symptomatology and impairment of function surgery was planned. The tumor was approached via dorsal approach and extensor tendon splitting and intraoperatively it was well encapsulated over the dorsum and both medial and lateral aspects of the phalanx (Figure 2A). Tumor was resected and shaved form all over the phalanx and the bone surfaces were covered by bone wax for adequate hemostasis (Figure 2B). Post operatively wound healed nicely and she was started on physical therapy to regain range of motion. She eventually gained full extension and flexion of the finger with excellent grip strength (Figure 2C and 2D). On two years follow up patient remained free of symptoms and with good function and radiographs didn't show any evidence of recurrence of the tumor (Figure 2E and 2F).

The specimen was sent to the pathology lab and the final diagnosis was consistent with bizarre parosteal osteocartilaginous proliferation, Nora's lesion (Figure 3).

Discussion

Bizarre Parosteal Osteocartilaginous Proliferation, BPOP, was first described by Nora and colleagues. They reported 35 cases of what they described as 'peripheral skeletal osteochondromatous lesions', which were histologically and radiologically distinctive [1]. It is a rare, reactive, mineralizing mesenchymal lesion that typically affects the surfaces of bones in the hands and feet, typically the proximal and middle phalanges as well as the metacarpal and metatarsal bones [2]. The hands are affected four times more than the feet. The disease has no gender preference [1]. Patients affected are commonly in their 20s and 30s [4,5]. Chromosomal anomalies associated with BPOP include t(1;17) (q32;q21) [6] and t(1;17) (q42;q23) [7].

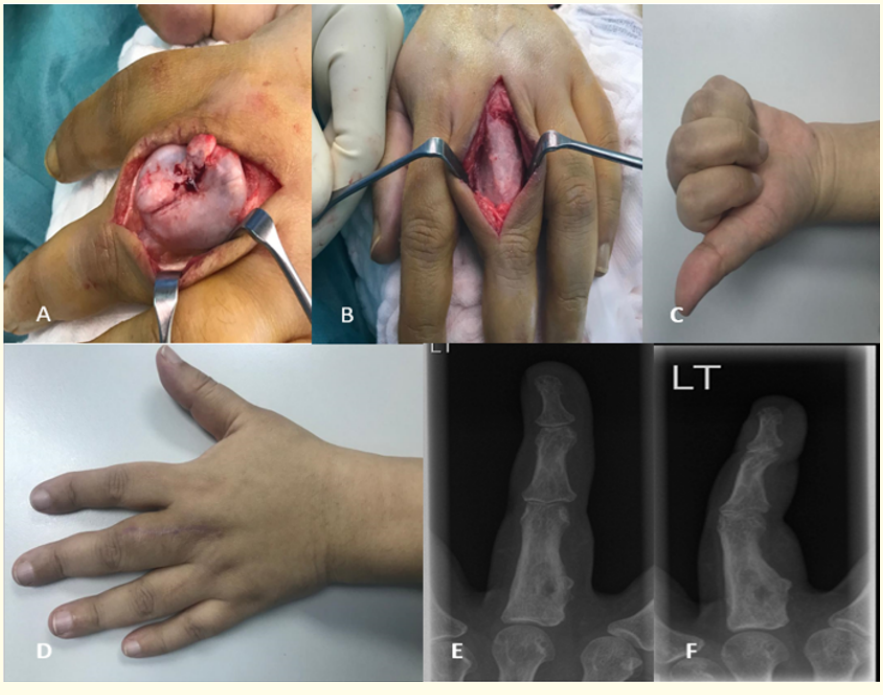

Figure 2: *A: Intraoperative image shows the tumor with a thin capsule covering the soft tissue mass. B: Tumor resection and the bone surface covered with bone wax. C, D: Range of motion after two years post surgery with full flexion and extension. E, F: Radiographs two years post-surgery.*

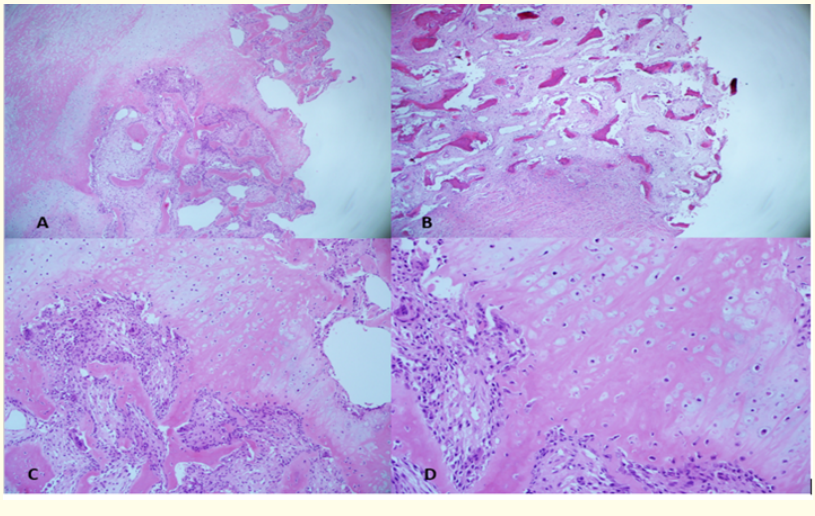

Figure 3: *Histological examination of the resected specimen shows fibrous and osseocartilaginous tissue (A, B). Most of the bone trabeculae are rimmed by osteoblasts and multinucleated osteoclast type giant cells C. The chondroid components reveals increased cellularity with scattered binucleated cells D.*

The typical clinical presentation is a painless swelling that grows slowly over months, sometimes associated with mild pain [11,12]. The most interesting aspect of BPOP are the radiographic and microscopic features which may be confused with several other osteochondromatous lesions, including both benign and malignant entities, such as osteochondroma, low grade parosteal osteosarcoma, myositis ossificans, florid reactive periostitis, and Turret exostosis. BPOP has an osteochondroma-like configuration with a typical ''stuck on'' appearance and lacks the cortical and medullary continuity seen in osteochondroma [13]. Osteochondroma, on the other hand, presents with a narrow or broad-based stalk that merges directly with the underlying cortex and medullary cavity [13]. Histologically, both lesions are covered peripherally by a cartilaginous cap [15]. The bony trabeculae are oriented at ninety degrees in osteochondromas [15], while BPOP demonstrates an irregular orientation of the trabecular bone, which also has a characteristic ''bluish'' appearance [13]. Low-grade parosteal osteosarcoma can also have features similar to BPOP both histologically and radiographically, although parosteal osteosarcomas are exceedingly rare in the small bones of the hands and feet. Parosteal osteosarcomas very commonly contain either a cartilage cap or islands of cartilage deeper within the tumor; however, this low-grade variant of osteosarcoma typically contains relatively mature bone deposited in an organized, variably cellular fibrous stroma.

When BPOP compared to Turret exostosis, Turret exostosis would have a mature osteocartilaginous cap overlying a transitional area with signs of endochondral ossification and bone tissue trabeculae lined by osteoblasts without atypia whereas florid reactive periostitis would show a picture of proliferating spindle cells with minimal osteocartilaginous growth [17].

BPOP has an atypical histological appearance, explaining the inclusion of the word "bizarre" in its name. Typical microscopic findings of BPOP include highly cellular, disorganized, and irregular cartilage, which is associated with the proliferation of bizarre-appearing fibroblasts, disorganized bone and spindle shaped fibroblasts in the intertrabecular space and bizarre binucleated chondrocytes of soft tissue [8,9]. The bluish staining of the calcified cartilage matrix is a distinctive feature that has been interpreted as "blue bone" [10]. The disease is usually benign in nature with no documented metastatic spread or mortality [1 as well it is usually affecting single site with no multiplicity]. To date only one case of malignant transformation in BPOP has been reported by Choi., *et al*. [16], although it is not clear from their report whether BPOP underwent transformation to fibrosarcoma or there were two distinct entities arising in the same location. indicated that BPOP should be considered a neoplastic rather than a reactive process based on the atypical cellular features often found in the biopsy samples [7].

In general, observation alone is considered adequate for asymptomatic BPOP, but simple excision is indicated for patients with pain or a functional disorder. Resection of the capsule of the lesion and decortication of the underlying cortical bone is reportedly important to reduce recurrence rates [18,19].

Conclusion

BPOP is a rare, reactive, mineralizing mesenchymal lesion that typically affects the surfaces of bones in the hands and feet. Radiologically, may be similar to other lesions like osteochondroma and parosteal osteosarcoma. The final diagnosis needs pathological testing to confirm the diagnosis. It can be treated surgically or conservative. When treated surgically Resection of the capsule of the lesion and decortication of the underlying cortical bone is needed as it has high risk of recurrence.

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