Bizarre Parosteal Osteochondromatous Proliferation (Nora Lesion) in Upper and Lower Limbs: A Report of Three Cases and Review of Literature

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Learning Point of the Article:

BPOP is a rare benign lesion of the bone which poses difficulty in diagnosis but once diagnosed, you can expect good clinical outcome with excision.

Abstract

Introduction: Bizarre parosteal osteochondromatous proliferation (BPOP) is a rare bone pathology affecting small bones of hand and feet. This benign lesion needs to be distinguished from many malignant bone tumors as it poses a diagnostic dilemma due to its clinical, radiological, and histological picture. We report three cases of BPOP affecting the hand and foot.

Case 1: A 21-year-old gentleman presented with painful swelling in the long finger of the right hand. A plain radiograph showed a radio dense mass which was later excised and diagnosis confirmed in histopathology. There was no recurrence in 2 years of follow-up.

Case 2: A 5-year-old boy presented with painful swelling over the right ankle with no history of antecedent trauma. Following radiological evaluation, the patient was successfully treated with excision.

Case 3: A 35-year-old lady presented with a painful swelling on the dorsal aspect of her hand which was gradually increasing in size. After radiological evaluation, the patient was successfully treated with excision and lesion confirmed to be BPOP on histological examination. She was symptom free without recurrence in up to 2 years of follow-up.

Conclusion: Nora's lesion is a rare pathology requiring high index of suspicion. Excision is the recommended mode of treatment. All our cases responded well with excision with immediate pain relief following surgery and no recurrence in up to 2 years of follow-up.

Keywords: Nora, bizarre parosteal osteochondromatous proliferation, neoplasm, tumor, benign.

Introduction

Bizarre parosteal osteochondromatous proliferation (BPOP) is a rare bone pathology affecting small bones of hand and feet [1]. First described by Nora et al. in 1983, over 200 cases have been reported since then world over [2, 3]. The clinical presentation of this condition is that of a bony swelling and poses a diagnostic dilemma due to its aggressive radiological appearance and confusing histological picture. It needs to be distinguished from malignant bone tumors such as parosteal osteosarcoma and chondrosarcoma or benign pathologies such as myositis ossificans and osteochondroma [4-7]. We report three cases of BPOP, all of which presented as painful swelling and were successfully treated with resection following which diagnosis was confirmed with histopathology.

Case Report

Case 1

A 21-year-old gentleman presented in the outpatient department (OPS) with a painful swelling on the right hand. He denied any history of trauma. Clinical examination revealed a

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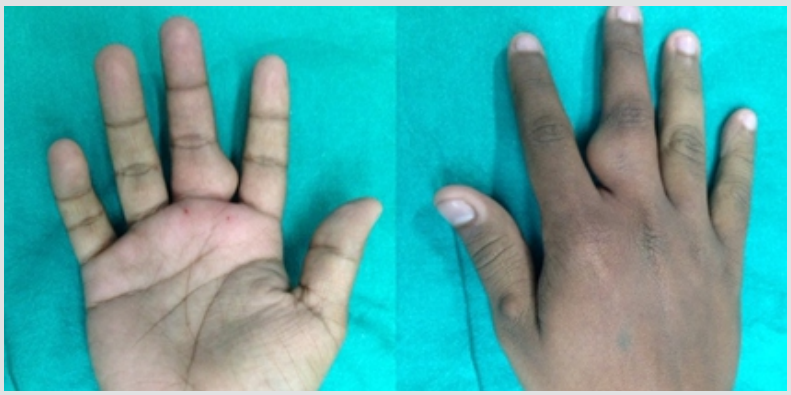

Figure 1: Clinical picture of swelling on the long finger.

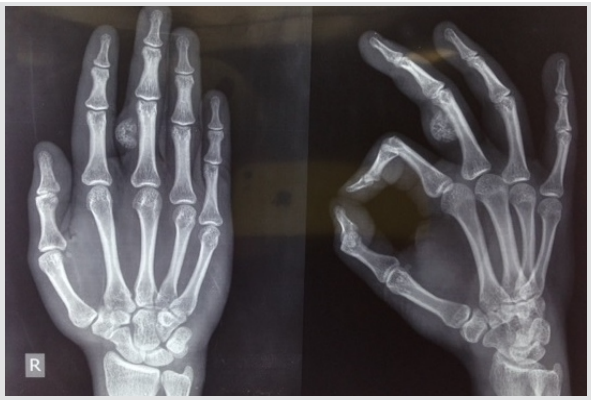

Figure 2: Plain radiograph of the affected hand in anteroposterior and oblique projection views showing dense mass.

bony hard swelling measuring about 2 cm in length and 1 cm in width over the palmar and radial aspect over the proximal phalanx of the middle finger in the right hand (Fig. 1). A plain radiograph was obtained in anteroposterior and oblique projection views which showed a dense mass measuring about 2 cm adjacent to the proximal phalanx of the long finger, the margins were well circumscribed and cortical erosion was noted on the adjacent phalanx (Fig. 2). The mass was excised under regional anesthesia using a lateral incision. The bony mass was not adherent to the underlying bone and was send the mass for histopathological evaluation which showed bony trabeculae intermixed with fibrous and chondroid cells (Fig. 3). The patient was on regular follow-up and no recurrence was noted in up to 2 years of follow-up.

Case 2

A 5-year-old boy presented to the OPD with pain and swelling of right ankle for 6 months. He gave no history of antecedent trauma, fever, or other joint swelling. Clinical examination revealed circumferential diffuse ankle effusion with limitation of ankle dorsiflexion. Skin was normal color and no local rise of temperature noted. Plain radiograph in anteroposterior and lateral projection view showed an irregular dense round mass over the anterior aspect of the talus with overlying cortical erosion of the talus (Fig. 4). Subsequent magnetic resonance imaging showed a well formed mass hypointense in T1W and hyperintense in T2W images lacking any cortical continuity with underlying talus (Fig. 5). Excision biopsy was done using a dorsal incision directly overlying the mass and proceeds were send for histopathological evaluation. The microscopic section showed circumscribed lesion with a fibrous capsule, a fibrous myxoid spindle cell stroma, and hypercellular cartilaginous cap with endochondral ossification (Fig. 6). The patient was symptom free and had no recurrence in up to 2 years of follow-up.

Case 3

A 35-year-old lady presented to the OPD with complaints of a painful swelling over on her right hand. She noticed the swelling 3 years back and the swelling had gradually increased in size since then. On clinical evaluation, the swelling was bony hard in consistency, located over the dorsal aspect of the head of second metacarpal, and measured 2 cm in length and 2 cm in width. Radiograph revealed a radiodense mass over the second metacarpal head and cortical erosion over the adjacent bone was noticed. Further evaluation with computed tomography scan showed a well-circumscribed mass with clear corticomedullary differentiation, overlying the dorsal and distal aspect of second metacarpal bone with close cortical approximation with it (Fig. 6). The mass was excised in toto using a dorsal incision and send for histopathology. Histopathology examination confirmed the absence of

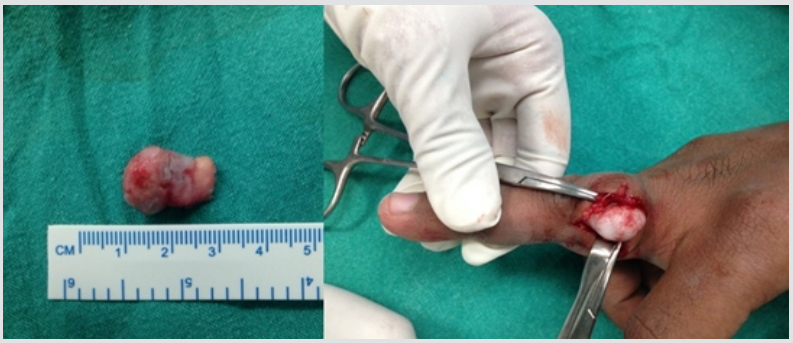

Figure 3: Intraoperative picture of the approach and excised lesion.

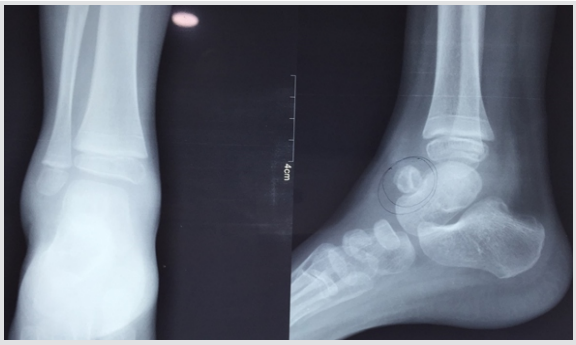

Figure 4: Anteroposterior and lateral projection view of ankle showing radiodense lesion adjacent to the talus.

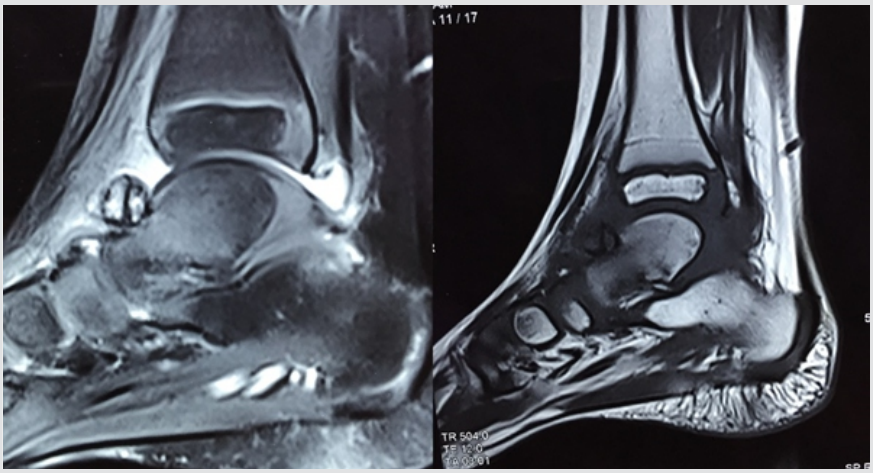

Figure 5: Magnetic resonance scan showing well-formed mass, hypointense in T1W and hyperintense in T2W images lacking any cortical continuity with underlying talus.

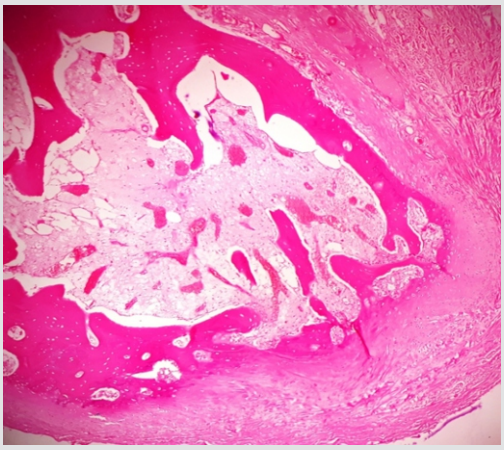

Figure 6: Microscopic section of the excised lesion showing circumscribed lesion with a fibrous capsule, a fibrous myxoid spindle cell stroma, and hypercellular cartilaginous cap with endochondral ossification.

malignant changes and showed osteoid seams with cartilaginous cap, hence confirming BPOP. The patient was on regular follow-up for 2 years and was symptom free in this period.

Discussion

BPOP is also termed as Nora's lesion after the first clinical report of 35 cases by Nora et al. in 1983 [2]. The lesion is rare and affects small bones of hand and foot but there have been isolated reports on long bones and skull [4,8,9]. Young individuals in the second and third decades are more affected with no gender preponderance [4,6]. The exact etiology is unknown [1] and multiple theories are postulated including reactive bone formation following trauma [4], but majority of patients fail to give a history of trauma [10-12]. The clinical presentation is that of a slow-growing painful swelling.

Nora's lesion poses significant diagnostic challenge as it can give confusing radiological picture. The surface location of the lesion may resemble parosteal osteosarcoma [2]; the cartilaginous component would be confused for an osteochondroma [6]. Lack of corticomedullary continuity and occurrence in small bones of hand and feet may aid the diagnosis but there has been reported literature with corticomedullary continuity and atypical location and hence radiology solely cannot be relied on for diagnosis and histopathological evaluation is mandatory [13].

The prevalence of the pathology is difficult to be assessed due to the lack of large volume studies. The available case series and reports show high local recurrence. Two of the largest series reported showed recurrence rates of 51% and 29% [2,4]. En bloc excision including the pseudocapsule, any periosteal tissue beneath the lesion, and decortication of the underlying bone if it looks abnormal reduces the chances of local recurrence [14]. The lack of local adjuvant therapy to prevent the recurrence adds to the treatment challenge in Nora's lesion. Despite high local recurrence and occasional cellular atypia in the histopathological evaluation, there has been no incidence of malignant transformation, metastates, or death associated with this lesion [15].

The etiological factors resulting in Nora's lesion are a matter of debate. The classical location in the small bones of hand and feet resulted in the categorization of BPOP in the spectrum of reactive periostitis. The identification of the presence of basic fibroblastic growth factor expression backed by immunohistochemical and molecular analysis strengthened the theory of reparative process after injury to periosteum [16]. However like the two cases in our report and many other reported cases denies any history of trauma and high rate of recurrence strongly stand against the reparative theory. A balanced translocation t (1;17) (q32;q21) was identified by Nilsson et al. in cytogenetic analysis of five patients with Nora's lesion [17]. Endo et al. in a case report described a similar cytogenetic analysis in a 39-year-old woman with Nora's lesion [18]. These suggest a neoplastic etiology for BPOP.

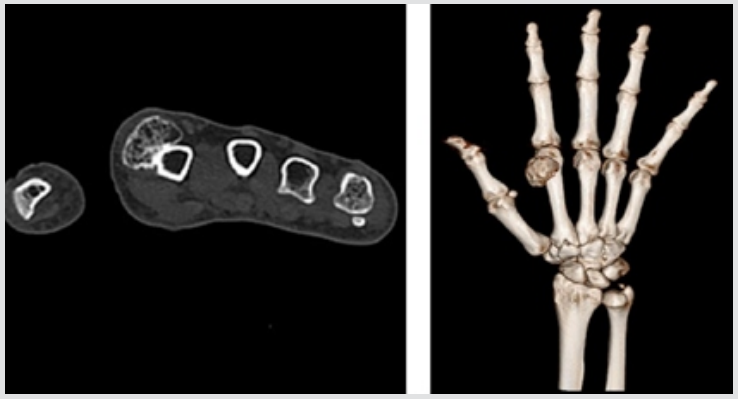

Figure 7: Computed tomography image showing the well-circumscribed lesion in the axial section and 3D image.

Conclusion

Nora's lesion is a rare pathology requiring high index of suspicion. Careful clinical and radiological analysis backed with histopathological examination aids in diagnosis. The etiological factors that cause the lesion are yet to be identified and there are significant diagnostic challenges posed by the nature of the lesion. Excision is the recommended treatment and all our cases had good results well with excision and no recurrence in up to 2 years of follow-up.

Clinical Message

BPOP may create a confusing clinical scenario as it mimics bone tumor. Strong clinical suspicion is needed to arrive at proper diagnosis and excision gives good results. We demonstrate the clinical presentation and management of this bizarre lesion through a series of cases in upper limb as well as lower limb. The evidence base and literature review are also provided.

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