Case Report

Nora Lesion (Bizarre Parosteal Osteochondromatous Proliferation): An Ultrasound Diagnosis with Magnetic Resonance Imaging Correlation

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Abstract

Advanced cross-sectional imaging techniques are firmly established as a means of evaluating musculoskeletal disease, and ultrasound (US) is increasingly being used for the assessment of a diversity of tendon, joint, and soft-tissue abnormalities. A benign condition – bizarre parosteal osteochondromatous proliferation (BPOP) – arises from the periosteum, typically in the small bones of the hands and feet, and grows as a surface bone lesion in the surrounding soft tissue. Proliferations can become symptomatic, exercising mass effect on adjacent structures that may require operative management. As a bone-forming process, BPOP may occasionally assume worrisome histologic features that mimic sarcoma, and a pronounced tendency to recur after primary excision. A solitary mass was growing in the middle finger of a young woman that curtailed proper hand function. With US, a partially ossified formation was revealed in the proximal phalanx situated on the outer surface of the bone. There was faint acoustic shadowing distal to the lesion, and a hypoechoic halo was seen covering part of the abnormal tissue growth. Importantly, the lesion caused significant limitation of motion of the finger, on the dynamic flexion US images with the displacement of the flexor tendon and compression of a digital nerve. To restore the range of motion in the finger, surgical excision of the juxtacortical mass was performed and histology yielded a diagnosis of BPOP. We describe the US features of digital BPOP, which were found to correspond closely to those of computed tomography and magnetic resonance imaging.

Keywords: Bizarre parosteal osteochondromatous proliferation, computed tomography, juxtacortical, magnetic resonance imaging, Nora lesion, phalanx, ultrasound

Introduction

In 1983, Frederick Nora *et al*.[1] first reported a reactive, surface-based osteocartilaginous process affecting the tubular bones in the hands and feet of 35 patients. Named after pathologist Nora and frequently referred to as Nora's lesion, this benign proliferative entity was described as bizarre parosteal osteochondromatous proliferation (BPOP). The hand, especially the metacarpal bones, is affected four times more commonly than the foot, although the condition can occur in other sites including the phalanges, long bones, calvarium, and pelvis.[2,3] Patients present with swelling or a painful palpable mass. Because BPOP is characterized by aggressive histologic features resembling those of malignant bone tumors and a high recurrence rate after excision, diagnosis of BPOP is notably challenging. The use of ultrasound (US) is firmly established as a means of evaluating musculoskeletal abnormalities, including various lesions in soft tissue.[4,5] We present the preoperative imaging work-up and differential diagnosis, in a patient with a cortically-based lesion and local aggressive behavior, favoring malignancy.

Case Report

A 31-year-old woman presented with a 1-month history of deep palpable sclerosis over the palmar surface of the swollen

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proximal phalanx. She complained that the growing, painless lesion curtailed the motion of the numb middle finger. On physical examination, the patient had decreased active and passive flexion at the right proximal interphalangeal (PIP) joint, measuring 40°. Localized paresthesia suggested direct pressure of the proper palmar digital nerve of the median nerve. A radiologic evaluation was prompted. Radiographs revealed a lobulated osteosclerotic lesion arising from the volar and ulnar surfaces of the proximal phalanx [Figure 1]. US examination was performed with a broadband, multifrequency linear transducer (6–12 MHz) on an Aplio a-series (Canon Medical Systems Corporation) US machine. US demonstrated a focal area of moderately increased echogenicity with some acoustic shadowing surrounded by a hypoechoic border, in the phalangeal diaphysis [Figure 2]. On the dynamic flexion-extension images, the mass appeared to jeopardize the flexor tendons, causing painful impingement and decreased flexion of the PIP. Computed tomography (CT) images disclosed a parosteal, corticated hypodense mass. The ossified lesion originated directly from the cortex of the phalanx, lacking any medullary continuity with the underlying cancellous bone [Figure 3]. Magnetic resonance (MR) imaging was performed. An ovoid area of decreased signal intensity on T1-weighted and increased signal intensity on T2-weighted images was seen attached to the volar midshaft of the proximal phalanx. The cartilage around the lesion resulted in the mass having a low signal intensity border on T1-weighted MR images and a high signal on T2-weighted MR images. Similar to the US findings, the cortically-based extraosseous mass was seen interposed between bone and the flexor tendons and exerted a mass effect on the neighboring, ulnar-sided digital neurovascular bundle [Figure 4]. Compression deformity of the tendinous and neurovascular structures caused by the ossified lesion resulted in motor-sensory disturbance and functional deficit of the middle finger.

Surgical excision of the lesion was then performed. Histopathologic examination disclosed a well-marginated osteocartilaginous mass contiguous with underlying cortical bone, with no medullary continuity with the parent bone. The interfaces between bone and hypercellular cartilage were characterized as grossly irregular. Individual chondrocytes were enlarged, bizarre, and binucleate [Figure 5]. The final pathologic diagnosis was BPOP or Nora lesion of the third digit. Approximately 2 years after surgery, there has been no evidence of local recurrence or distant metastasis.

Discussion

BPOP lesions are uncommon, with fewer than 200 cases sporadically reported in the literature. With regard to epidemiology, BPOP has no gender predilection, and the highest incidence is in the second and third decades of life. The etiology of these osteochondromatous proliferations is unknown; however, a relationship with preceding trauma has been suggested.[1,2,6] Our patient recalled a history of remote hand injury while participating on the high school baseball team, which was probably unrelated to the current situation.

BPOP is a benign proliferative entity that histologically is characterized by the presence of atypical hypercellular cartilage, which forms irregular interfaces with immature bone in a fibrous cell stroma. The atypical cartilage harbors enlarged, bizarre, and binucleate chondrocytes that may mimic chondrosarcoma or surface osteosarcoma.[1,2,7] Interestingly, immature bone exhibits a distinct blue tinctorial quality.[1,2] Chromosomal aberrations have been identified on cytogenetic analyses of BPOP cases, although solid evidence of a true neoplastic origin is missing.[8] Whether related to periosteal injury or a neoplastic process with a remarkable tendency to recur (up to 50%) after excision, BPOP may evolve from florid reactive periostitis to an overly calcified mass.[6,9-11] As such, a "unitary hypothesis" has been postulated suggesting a continuum among the periosteal reactive processes.[6,10] Although uncommon, BPOP has become an important entity to recognize for two principal reasons: it often mimics benign or malignant bone neoplasms, and it may cause mass effects, leading to neurologic, vascular, or tendon complications. In addition, in terms of treatment, correct

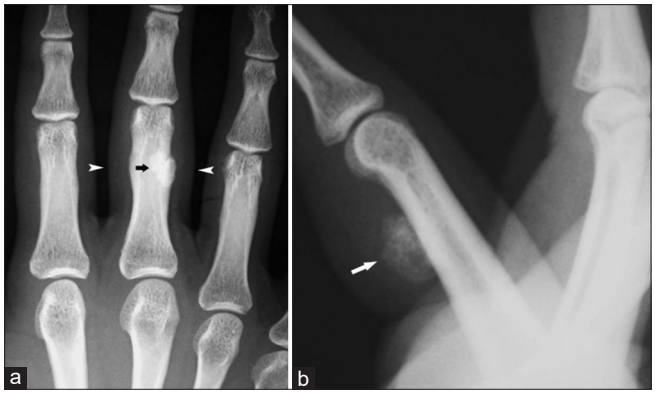

Figure 1: (a) Posteroanterior radiograph of the right second to fifth digits shows a diffusely sclerotic lesion projecting over the proximal phalanx of the middle finger (arrow). Note soft-tissue swelling in the affected finger (arrowheads). (b) The lateral radiograph reveals an ossified lesion (arrow) projecting on the volar aspect of the finger

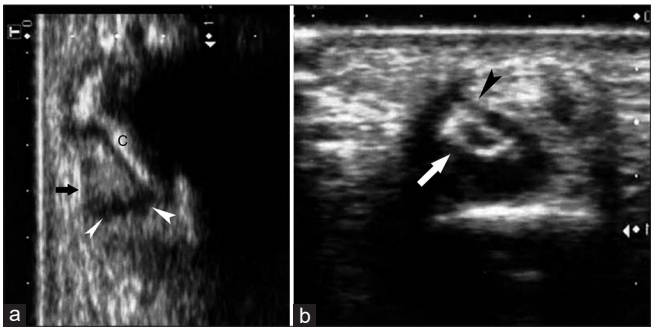

Figure 2: (a) Longitudinal ultrasound image shows a lobular echogenic mass (arrow) with an outer echolucent border (arrowheads) that is superficial relative to the cortex, (c) of the phalanx, and (b) transverse ultrasound image depicts the corticated mass (arrow) arising directly from the bony cortex of the phalanx (arrowhead)

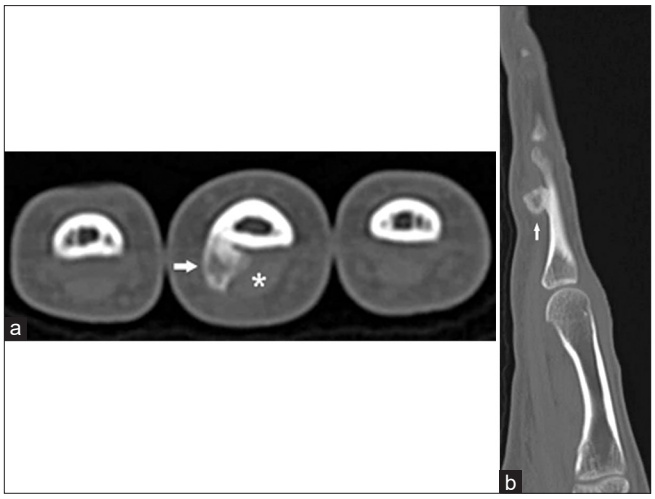

Figure 3: (a) Axial CT image (bone windows) at the level of the proximal second to fourth phalanges shows an ossified mass (arrow) arising from the bone cortex causing displacement of the flexor tendon (asterisk). (b) The sagittal MPR CT image in the bone reconstruction algorithm of the affected middle finger shows the broad-based ossified lesion (arrow) originating from the cortical surface of the bone. Note the sparing of the cancellous bone. CT: Computed tomography, MPR: Multiplanar reconstructed

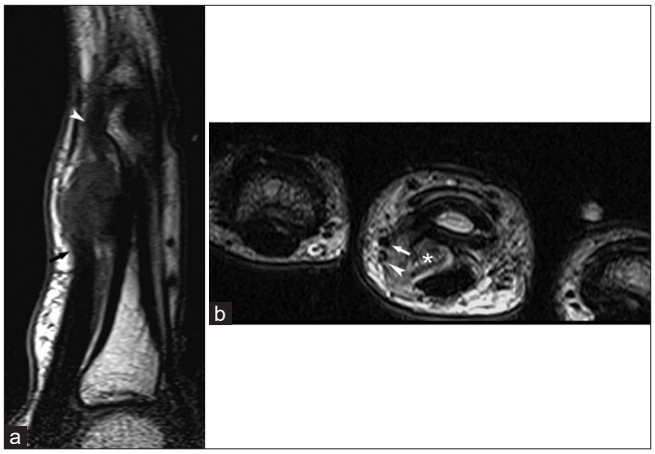

Figure 4: (a) Sagittal T1-weighted MR image of the middle finger shows the bone surface mass abutting the flexor tendons (arrow) and the volar plate (arrowhead). (b) Axial T2-weighted MR image of the third digit at the level of the proximal second to fourth phalanges shows the juxtacortical mass (asterisk) interposed between the phalanx and the deformed tendon. Note the adjacent digital artery (arrow) and nerve (arrowhead). MR: Magnetic resonance

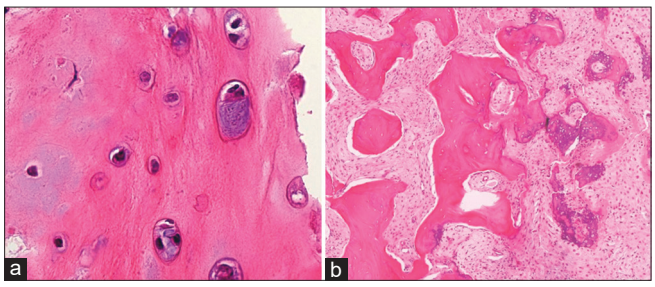

Figure 5: Histopathological photomicrographs (hematoxylin-eosin staining). (a) ×25. Note irregular osteochondral interfaces interspersed with fibrous tissue. (b) ×200. Bizarre, enlarged, binucleate chondrocytes

diagnosis is warranted as BPOP requires aggressive resection and possible reconstruction, and the patients need to be specifically informed of an increased rate of recurrence.[9] Patients with BPOP often have an initial presentation of soft-tissue swelling or pain (or both) over a juxta-articular lesion. Our case is unusual in that the juxta-articular mass exercised mass effect on an adjacent tendon and nerve. The tentative clinical diagnosis favored at the time of presentation, given the patient's age, rapid growth, and locally aggressive behavior was a malignancy. Radiography revealed a large, exophytic bony formation arising in the proximal phalanx. US played a pivotal role in displaying a bone surface mass lesion of mixed osseous and cartilaginous tissue consistency that compromised the flexor tendon while limiting the motion of the finger. Both CT and MR imaging allowed the assessment of the integrity of the underlying cortical bone and facilitated the depiction of the tendon and regional neurovascular structures. The mass was removed to restore the motion of the finger and the diagnosis of BPOP was confirmed by results from histopathology.

Imaging findings were fundamental to the diagnosis of BPOP in this individual case. In general, the radiologic differential diagnosis of BPOP from benign parosteal osteochondromatous proliferations should focus on osteochondroma, florid reactive periostitis, turret exostosis, subungual exostosis, and myositis ossificans. Other malignant processes include chondrosarcoma and parosteal (surface) osteosarcoma.[1,7] Osteochondroma is more common and may arise in the small bones of the hands and feet in 10% of the cases, whereas BPOP is rare, and usually involves the metacarpal and metatarsal bones.[3,12] The former points away from the joint and is characterized by corticomedullary continuity with the parent bone and an overlying cartilage cap, whereas the latter does not and typically is noncontiguous with the underlying bone and also lacks a hyaline cartilage cap.[10,11] Unlike BPOP, however, osteochondroma does not exhibit cytologic atypia of the cartilage. Florid reactive periostitis is signified by an aggressive lamellated periosteal reaction, and turret exostosis involves a broad-based and dome-shaped osseous excrescence. Subungual exostosis has a characteristic location and is dissimilar to BPOP, it lacks the irregular chondroid material. Myositis ossificans is usually separated from the adjacent bone and contains orderly and mature bone. Although rapidly growing osteochondromatous lesions may occasionally exhibit locally aggressive behavior that prompts differentiation from malignant tumors including bone sarcomas, the latter only rarely affects the small bones of the hands and feet and are characterized by predominant cortical and medullary bone destruction, and metastatic potential.[3,12] Finally, in BPOP cases, profound soft-tissue swelling may be present, especially if the affected bone is superficial (as with the small bones in the hand and foot); however, documentation of largely intact bone eliminates the diagnosis of osteomyelitis.

Because BPOP is a surface bone lesion, US may prove especially useful in direct visualization of BPOP and rapid characterization of the questionable digital mass. Importantly, due to the potentially interactive nature of US, the application of pressure to the finger producing digital flexion and extension

may identify the source of the reproduced clinical symptoms, which closely approximates the clinical situation. Our case represents a classic presentation of BPOP: A young patient with an ossified mass arising from the periosteum of the phalanx. Details of this case that make it noteworthy include the presence of compromised motor-sensory function in the swollen finger secondary to mass effect, which coupled with the imaging appearances of BPOP eliminates the possibility of malignancy. On US examination, the osteocartilaginous formation representing BPOP appears as a relatively echogenic mass surrounded by an outer hypoechoic border that grows from the cortical surface of the bone. Although US features, in some cases, can be helpful in determining a specific diagnosis of BPOP, sonographic findings can strongly suggest the presence of a parosteal osteochondromatous proliferation. Finally, because BPOP is a surface bone lesion, US may efficiently allow for dynamic assessment of the cause of a decreased range of motion in affected areas, as in this case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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