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TEST YOURSELF: ANSWER

Painless hand mass

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Answer

Bizarre parosteal osteochondromatous proliferation (Nora lesion) of the metacarpal.

Discussion

Radiograph (Fig. 1) and CT (Fig. 2) images show a well-corticated osseous lesion originating from the dorsomedial surface of the second metacarpal neck. There is a slight irregularity of the underlying cortex, but no periosteal reaction or bone destruction. Although no visible cleavage plane separates the lesion from the second metacarpal, the medullary space of the lesion is noncontiguous with that of the underlying bone, and it lacks characteristic orientation away from the joint space as is seen with an osteochondroma. Clinically, a firm, immobile, nontender mass was palpated in the second intermetacarpal space. The overlying skin was normal in appearance. Because the patient had a limited range of motion at the second and third carpometacarpal joints, surgical excision was performed. Histopathology demonstrates fragments of bone and cartilage with areas of endochondral ossification and surrounding bland fibroblastic proliferation. The interface between forming bone and cartilage has a characteristic purple-blue staining quality (Fig. 3).

First described by Nora et al. in 1983 (described in Abramovici and Steiner [1]), bizarre parosteal osteochondromatous proliferation (BPOP), also referred to as a "Nora lesion," is a rare, benign, locally aggressive osteochondromatous exostosis composed of bone and cartilage in a fibrous myxoid cell stroma without cellular atypia. Histologically, BPOP is characterized by the presence of a hypercellular fibrocartilaginous cap containing large, bizarre binucleate chondrocytes, with admixed areas of endochondral ossification maturing to trabecular bone. Bony trabeculae are histologically immature, with high osteoblastic activity and irregular calcification.

Bizarre parosteal osteochondromatous proliferation most commonly arise from the periosteum of the metacarpals and metatarsals, with the hand affected four times more commonly

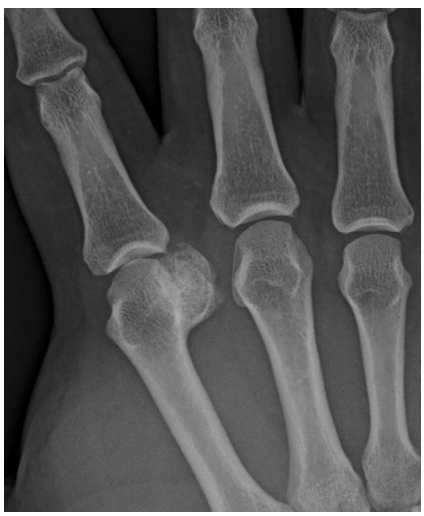

Fig. 1 Anteroposterior radiograph of the right second to fourth metacarpals and proximal phalanges

The case presentation can be found at doi: 10.1007/s00256-016-2543-z

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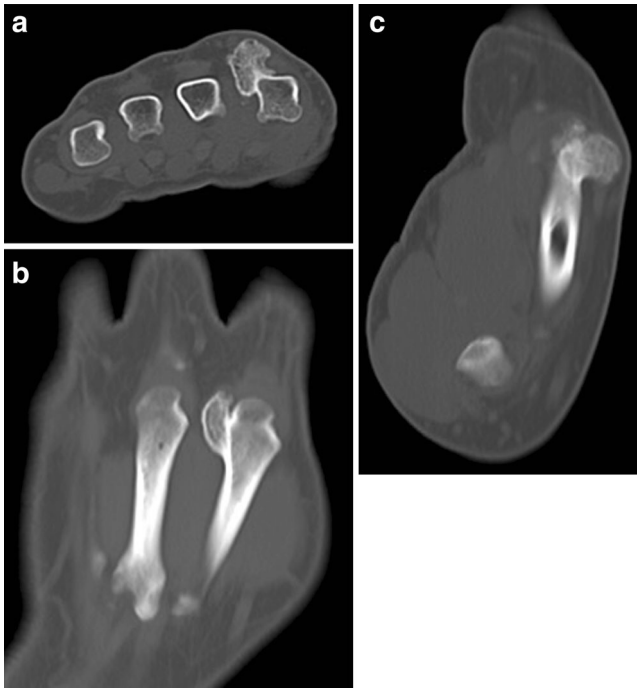

Fig. 2 **a** Axial, **b** coronal, and **c** sagittal unenhanced CT of the right hand at the bone windows

than the foot. Rare cases have been reported in the long bones, calvarium, maxilla, pelvis, clavicle, tibial sesamoid, and iliac crest. Clinically, BPOP are characterized by rapid growth and locally aggressive behavior. Most affected patients are in the second and third decades of life, with the youngest reported patient presenting at age 3 months [2]. There is no gender predilection. Because they have a high recurrence rate (20–55%) after marginal resection, resection of the pseudocapsule surrounding the lesion, any underlying periosteal tissue, and any abnormal-appearing areas in the underlying host bone is recommended [1, 3].

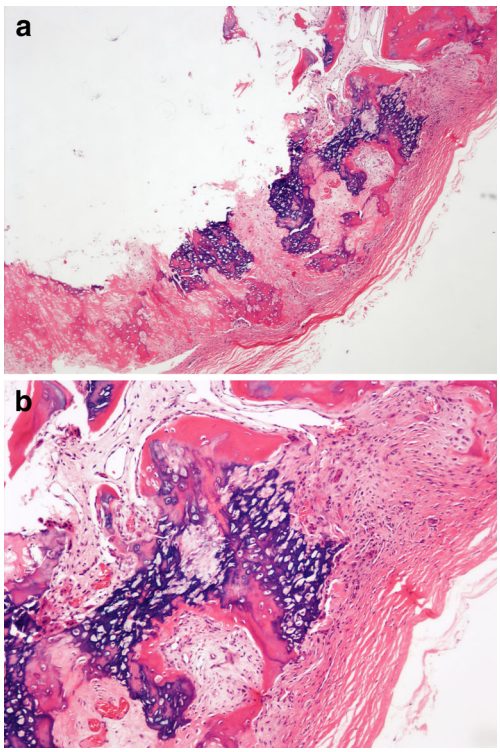

Fig. 3 Histopathological photomicrographs (hematoxylin and eosin staining). **a** ×40 and **b** ×100

Radiographically, BPOP manifests as a well-marginated bony exostosis oriented toward or parallel to the joint space, contiguous with, but not disruptive of, the native cortex, and lacking corticomedullary continuity. Underlying bony architecture and surrounding soft tissues are preserved. MRI may demonstrate a cartilage cap and overlying pseudocapsule, although the latter is better appreciated histologically [4]. When present, this cartilage cap is less well formed than that of an osteochondroma. Marrow signal may appear hypointense or isointense to muscle on T1-weighted sequences, and hyperintense on T2-weighted sequences on the surface, with more signal heterogeneity in the deeper part of the lesion, radiologically suggesting a more aggressive lesion. The underlying native cortex, medullary bone, and adjacent soft tissues are normal in signal intensity. Radionuclide bone scans typically show increased uptake of Tc-99 m MDP [4].

Radiographic findings alone are sufficient to diagnosis BPOP in typical cases. The potential for considerable variation in imaging and histopathological features, which may overlap with other parosteal osteochondromatous proliferations, such as osteochondroma, turret exostosis, florid reactive periostitis, and surface osteosarcoma, may necessitate histological sampling or excision. There are reports of histologically confirmed BPOP lesions demonstrating corticomedullary continuity, cortical destruction, and intramedullary edema on imaging [3]. Distinguishing BPOP from radiologically similar entities, both benign and malignant, is the major challenge to radiologists. Familiarity with these various entities is essential for generating an appropriate differential diagnosis. Osteochondromas are a common imaging finding, seen in up to 3% of the population. Most arise in the appendicular skeleton, typically the distal femur, proximal tibia, and humerus, with approximately 10% occurring in the small bones of the hands and feet. These cartilage-capped osseous exostoses may be sessile or broad-based and are classically distinguished by their corticomedullary continuity with underlying bone. They are developmental lesions that cease growth at skeletal maturity and are usually discovered in the first three decades of life. While typically painless, some may become symptomatic secondary to impingement, trauma, or, rarely, malignant degeneration to chondrosarcoma. Other osteochondroma-like lesions may occur in the hands and feet, including turret exostosis and florid reactive periostitis; although their etiology is unknown, a relationship with the preceding trauma has been suggested. Both typically present with pain and swelling at the lesion site, and are primarily

significant for their clinical and radiographic simulation of infection or osteosarcoma. In contrast to BPOP, the phalanges are involved more often than the metacarpals. Depending on lesion maturity at the time of imaging, the radiological appearance is variable. Initial radiographs may only demonstrate soft-tissue swelling at the affected site; as healing progresses, aggressive lamellated periostitis (more common with florid reactive periostitis) and the development of broad-based, dome-shaped osseous excrescence (often with turret exostosis) are seen [5]. Bone scintigraphy shows markedly increased uptake initially that gradually decreases with maturation. High-grade surface osteosarcomas, which account for less than 1% of all osteosarcomas and approximately 10% of juxtacortical osteosarcomas, very rarely involve the short bones of the hands and feet, preferentially affecting the femur, humerus, tibia or fibula. These high-grade, bone-forming tumors often incite an aggressive periosteal reaction and result in destruction of the underlying cortex; MRI often reveals edema in the adjacent cortex, and intramedullary or soft-tissue invasion beyond the visible bony matrix. Demographics are similar to those of conventional osteosarcoma, with a slight male predominance and peak incidence in the second decade of life.

The etiology of BPOP is unknown. It has been theorized that lesions might represent a proliferative response to periosteal injury [6], with some investigators suggesting that BPOP might reflect one clinicopathological manifestation in a spectrum of related parosteal osteochondromatous proliferations, including florid reactive periostitis and turret exostosis, which are distinguished by their degree of differentiation [7]. This "unitary hypothesis" has been called into question given the inconsistent history of antecedent trauma. Zambrano et al. suggested that BPOP could represent a neoplastic process, identifying karyotypic abnormalities of chromosomes 7 and 12 in one lesion, and nonclonal abnormalities of chromosomes 2, 8, and 14 in another [8]. Cytogenetic analyses by Nilsson et al. and Endo et al. revealed balanced chromosomal translocations at t(1;17) (q32;q21) and t(1;17) (q42;q23) respectively, raising the possibility that t(1;17) might represent a distinct translocation point that is unique to BPOP [9, 10]. Subsequent studies, however, fail to confirm these findings, indicating that BPOP may not result from a single chromosomal aberration or distinct translocation point [3].

Although the etiology, natural history, and clinical course of BPOP are not completely understood, typical imaging findings may allow a confident diagnosis and spare patients invasive procedures. The potential for radiological and histopathological overlap with other benign and malignant entities may necessitate histological analysis in certain cases. Accordingly, multidisciplinary management by clinicians, radiologists, and pathologists is indicated.

Compliance with ethical standards

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Conflicts of interest The authors declare that they have no conflicts of interest.

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