A 30-years-old man presented with a seven to eight months history of a painless, progressively increasing hard mass on the palmar aspect of the right middle finger, located at the level of the PIP joint, causing mild restriction of the movement at the same joint. There was no history of trauma.

What is your diagnosis?

Diagnosis: Bizarre Parosteal Osteochondromatous

The radiograph shows a well-defined, oval bony mass with smooth margins, just adjacent to the ventral cortex of the shaft of the middle proximal phalanx, with no evidence of corticomedullary continuation. The longitudinal axis of the mass is parallel to the middle of the proximal phalanx. There is no evidence of any underlying periosteal reaction [Figure 2]. These findings are diagnostic of bizarre parosteal osteochondromatous proliferation (BPOP), also known as Nora’s lesion.

Nora et al first described BPOP in 1983, when it came to be known as a Nora’s lesion or disease.\textsuperscript{[1]} Many benign lesions like osteochondroma, florid reactive periostitis, myositis ossificans and periosteal chondroma mimic this lesion.\textsuperscript{[1-3]} BPOP is a rare lesion commonly affecting the hands and feet of young adults, in the second and third decades, without...
any sex predilection. The most common sites in decreasing order of frequency, are the proximal and middle phalanges followed by the metacarpal and metatarsal bones,[4] though it can also arise in other tubular bones and the skull and jaw bones. BPOP arising from the sesamoid bone of the great toe has also been reported.[5]

BPOP typically presents with a painless or occasionally a mildly painful mass that grows over a period of few weeks to months. Stiffness of the adjacent joint or other mechanical symptoms may be seen. On plain radiographs, it appears as a well-defined bony mass,[4,6] like an osteochondroma, except that it lacks the characteristic orientation away from the nearby physis seen in the latter.[2] The lesion may be sessile or pedunculated. There is absence of continuity between the cortex and medullary cavity of the parent bone and the lesion[1,4] and this can be better delineated by a CT scan, which helps to distinguish BPOP from osteochondroma. Also, the well-formed cartilage cap, characteristic of osteochondroma,[2] which on T2W MRI images, appears hyperintense to bone, is absent in BPOP. On MRI images, BPOP appears as a hypointense lesion on T1W images and hyperintense lesion on T2W images, similar to other neoplastic lesions. However, in contrast to malignant lesions, BPOP exhibits no periosteal reaction and has normal underlying bone and adjacent soft tissue.

Despite the high rate of recurrence, BPOP is considered a benign lesion. Metastases have never been reported in the literature.[4,7] Histologically, the lesion contains highly cellular, disorganized and irregular cartilage, a proliferation of bizarre fibroblasts and disorganized bone with spindle shaped fibroblasts in the intertrabecular spaces. There is no “columnation” of the cartilage as in osteochondroma.[4]

No treatment is required, if a BPOP is asymptomatic, as the lesion is benign. If the patient is symptomatic (pain or compromised function), definitive treatment is by surgical excision with wide margins to minimize recurrence. When surgery is contemplated, nuclear bone scan must be performed prior to surgery, since recurrence typically occurs in lesions that have increased uptake.[8]

Florid reactive periostitis is similar in appearance to BPOP, but there is a history of trauma and it is mostly seen in the hands. Plain radiographs show the lesion adjacent to the bone rather than arising from it. Soft tissue swelling is seen as well as marked periosteal reaction. There may be new bone formation in the soft tissues and subtle cortical thinning.[9,10]

References


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