

A lesion of juxtacortical origin...

Une lésion juxtacorticale...

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I. THE CASE

A 24 year-old woman was seen for a nine months history of a hard mass at the second phalanx of her right middle finger. She denied any history of injury. Antero-posterior and lateral radiographs showed a calcified well juxtacortical well circumscribed mass (Figure 1).



Fig. 1a: Lateral radiograph: Calcified well circumscribed lesion, developed at the palmar aspect of the middle phalanx base, with no adjacent bone or soft tissue abnormality

High resolution ultrasound of the right medius showed a calcified lesion surrounded by a thin hypoechoic cap (Figure 2).

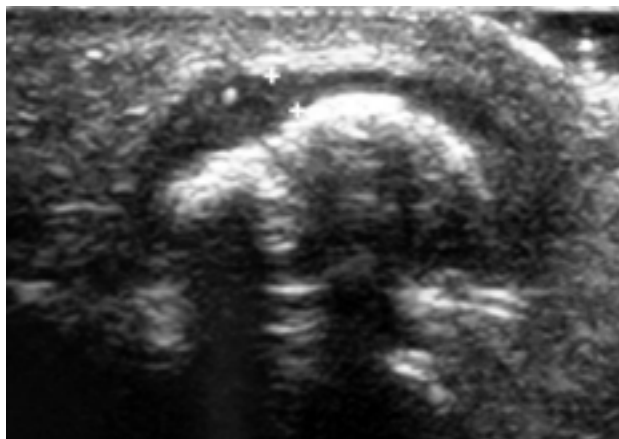


Fig. 1b: High resolution ultrasound in transversal view: calcified lesion surrounded by a thin hypoechoic cap

CT scan showed the lack of continuity between the lesion and the medullary canal of the affected bone; the cortex was normal (Figure 3).

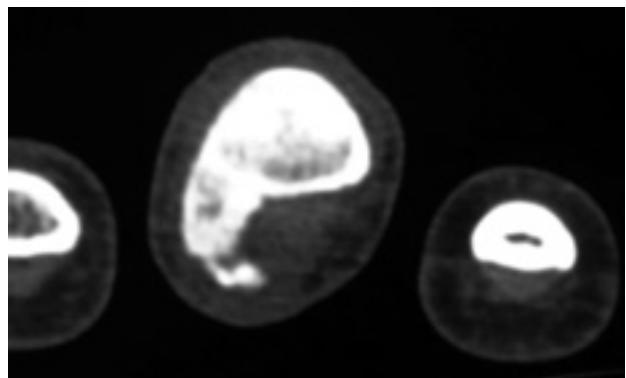
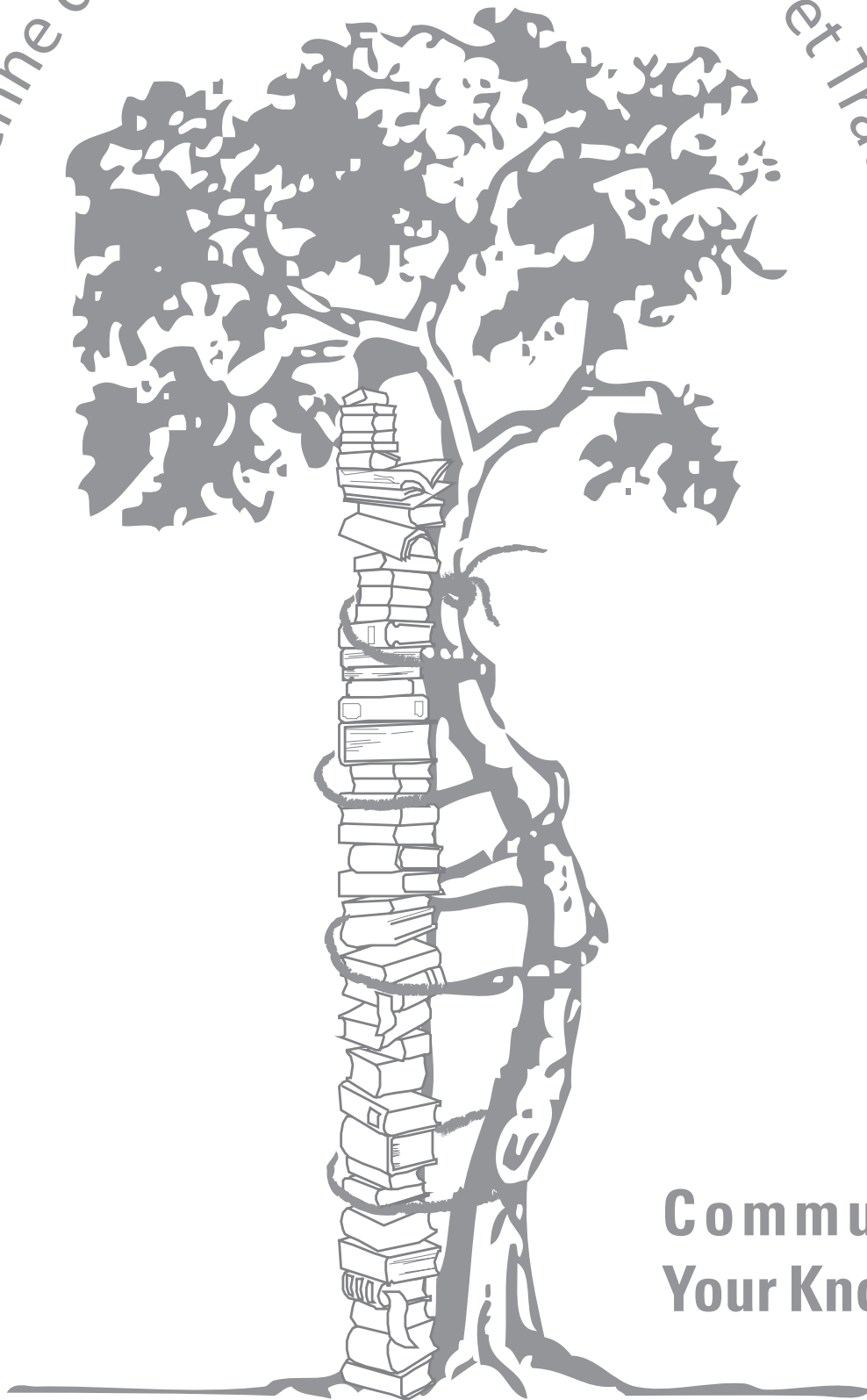


Fig. 3: Transversal CT view (bone algorithm): Surface bone lesion with large cortical base. There is no continuity between the lesion and the underlying bone cortex

What is your diagnosis ???

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Answer:**Bizarre parosteal osteochondromatous proliferation (BPOP) or Nora's lesion**

The patient underwent surgical excision of this lesion. Gross pathology examination showed a well circumscribed pediculated mass, of hard consistence and white greyish color (Figure 4).



Fig. 4: Gross pathology: Well circumscribed pediculated mass, of hard consistence and white greyish color

Microscopically, the lesion appeared as a tumoral proliferation with osseous, cartilaginous and fibrous components (Figure 5).

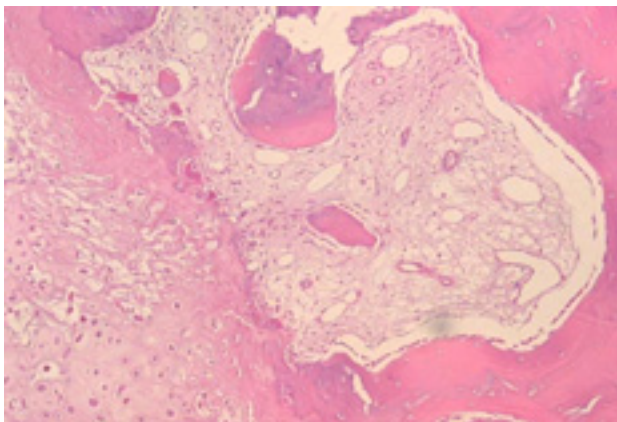


Fig. 5: Tumoral proliferation with osseous, cartilaginous and fibrous components (HEEx200)

Cartilaginous proliferation was made of chondrocytes with irregular morphology (bizarre cells) (Figure 6).

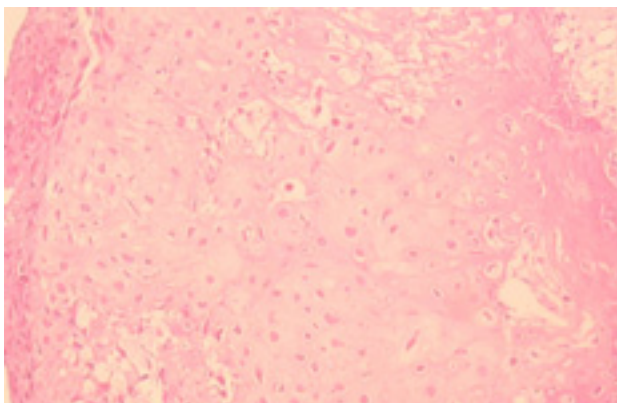


Fig. 6: Cartilaginous proliferation made of chondrocytes with irregular morphology (bizarre cells) (HEEx400)

The follow up showed no clinical or radiological signs of recurrence at more than one year.

II. LITERATURE

Bizarre parosteal osteochondromatous proliferation (BPOP) or NORA's lesion was named by the pathologist who first described it in 1983 at the Mayo Clinic [1]. He reported 35 cases, all involving the small bones of the hands and feet. MENESES et al. [2] expanded on Nora's work in 1993, when 65 cases of BPOP were presented, in which various other sites were outlined.

A- Epidemiology:

BPOP is a benign and rare surface lesion of bone that usually involves the proximal and middle phalanges, as well as metacarpal or metatarsal bones [1, 3]. It affects males and females equally [1, 4] and is most commonly encountered in the third and fourth decade [4, 5].

Hands are four times more commonly affected than feet [4], and the lesion does not tend to involve the distal phalanx in distinction from subungual exostosis [5, 6]. BPOP have been reported in unusual locations such as humerus [7] and clavicle [8].

B- Clinical presentation:

The typical clinical presentation is a painless swelling that grows over a period of months to years [9]. Any pain or skin erythema occasionally experienced by the patient is likely to be secondary to mass effect or mechanical problems [10, 11]. On examination, BPOP is a firm mass, usually small, ranging from 0.4 to 3cm in diameter and does not involve the overlying skin [1]. Joint motion may be limited when the lesion is located at the end of a bone [12].

C- Imaging findings:

On plain radiographs, BPOP is a well-marginated, calcified or ossified mass arising directly from the cortical surface of the underlying bone [5, 12]. It is generally attached by a broad base and the underlying cortex is intact [2, 12]. There is no periosteal new bone formation. However, cortical erosion has been reported [12-14].

In BPOP, fine cut Computed tomography scan shows a mass with well defined margins, intensely calcified or ossified, arising from the cortex of the affected bone. CT is, with evidence, better than plain radiography in showing the absence of continuity between the cortex and medullary cavity of the bone and the absence of cortical flaring in this affected bone [11, 15].

On MRI images, BPOP displays homogenous low signal intensity on T1 weighted sequences with uniform enhancement following Gadolinium administration. On T2 weighted images, the lesion is of high signal intensity, slightly increased signal centrally with its periphery being of higher signal intensity. In addition, BPOP exhibits neither periosteal reaction nor medullary involvement. It has normal underlying bone and adjacent soft tissues [5, 9, 12, 15].

D- Histology:

BPOP has an atypical histological appearance with highly cellular, disorganized and irregular cartilage, proliferation of bizarre fibroblasts and disorganized bone with spindle shaped fibroblasts [6, 9].

E- Differential diagnosis:

BPOP can appear similar to osteochondroma but the absence of continuity between the lesion and medullary cavity of the bone has been singled out as a key radiographic finding that differentiated BPOP from osteochondroma [5, 15]. In osteochondroma, chondrocytes lack atypia and often are arranged in parallel lacunar spaces [2, 12].

BPOP needs also to be distinguished from parosteal osteosarcoma, and a grade I or II chondrosarcoma, florid periostitis, myositis ossificans, stress fracture with extensive callous formation and Turret exostosis.

F- Pathogenesis:

HORIGUSHI [6] considered BPOP as a reparative process after periosteal injury. In addition to the frequent reported history of trauma, this author also demonstrates the expression of basic fibroblast and vascular endothelial growth factors, and chondromodulin I (ChMI) in the cartilaginous cap of BPOP, all involved in osteocartilaginous formation.

YUEN [16] suggested the existence of a continuum among florid reactive periostitis, BPOP, and turret exostosis and that each is a different stage of proliferative periosteal process.

LY [14] suggests a relationship between myositis ossificans and BPOP.

These theories are based on a history of trauma initial to BPOP, whereas many patients do not complain about previous injury or trauma.

For ORUI [3], systemic or focal inflammation may be a possible aetiology.

Recently, NILSSON [17] found a translocation between chromosome 1 and 17 t(1;17) (q32;q21) and identified a number of genes involved in other neoplasm (BRCA1 associated with breast cancer, and COL1A1 involved in dermatofibrosarcoma protuberans and giant cell fibroblastoma).

These recent findings lead to the suggestion that BPOP represents a neoplastic rather than a reactive lesion.

G- Treatment:

No treatment is required if a BPOP is asymptomatic, as the lesion is benign [9]. If the patient is symptomatic (pain or compromised function), definitive treatment is by surgical excision with wide margins [9, 17]. BPOP has a remarkable tendency to recur [7, 15], en bloc negative margin excision by the excision of the pseudocapsule over the lesion and any periosteal tissue beneath the lesion and decortication of any areas in the underlying host bone that appear abnormal has been

shown to be beneficial in preventing local recurrence [12]. Nora's lesion requires then long-term follow up.

No malignant transformation, metastases or deaths have been described so far in patients with BPOP [1, 7].

III. REFERENCES

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