Painful Pseudoclubbing of a Single Nail Unit

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Question

A 38-year-old female was referred for dermatologic evaluation for gradual growth and thickening of her left second fingernail. The deformity had been present for the past 10 years and was occasionally painful. On examination, the distal portion of the left index finger was at least 1.5 times bigger than that on the contralateral hand. The

fingernail demonstrated overcurvature in both the transverse and longitudinal axes causing pseudoclubbing, and the nail plate surface was distorted, with discrete transverse ridging (Fig. 1a, b). All other fingernails and toenails were normal, and other bone lesions were not identified. The patient denied history of trauma to the digit or family history of bone or nail lesions.

What is your diagnosis?

Fig. 1. Clinical presentation. **a** The distal portion of the left second finger is enlarged when compared to the rest of the digits. **b** Lateral view of the index finger. There is pronounced longitudinal and transverse overcurvature, and Lovibond's angle is $>180^\circ$.

Color version available online

Fig. 2. X-rays. **a** Posteroanterior view of left hand. A markedly expansile, largely lytic, osseous lesion in the distal phalanx of the second finger is seen. **b** Oblique view. There is chondroid matrix throughout the lesion measuring up to $1.6 \times 1.4 \times 1.2$ cm and ex-

tending to the articular surface without disrupting its surface. No involvement of the interphalangeal joint is detected. **c** Magnified posteroanterior view. The appearance of the lesion is typical of an enchondroma.

Answer

Enchondroma

X-ray imaging of the left hand showed a $1.6 \times 1.4 \times 1.2$ cm expansile, lytic, osseous lesion of the distal phalanx of the left index finger. The lesion extended to the articular surface, without evidence of articular surface disruption or distal interphalangeal joint involvement. Chondroid matrix was seen throughout the lesion (Fig. 2a–c). Given these findings, a diagnosis of enchondroma was rendered.

Since the patient complained of painful throbbing of the affected nail, the cartilaginous tumor was thoroughly removed by curettage, and demineralized bone matrix (Grafton®) was used as scaffolding for bone ingrowth.

After fixation in formalin, the specimen was decalcified and embedded in paraffin for microscopic analysis. Histologic sections revealed a well-differentiated chondroid neoplasm, composed of mature cartilage with focal osteoid formation consistent with enchondroma (Fig. 3a–d).

Discussion

An enchondroma is a benign cartilaginous neoplasm that occurs in intramedullary bone. The precise etiology of enchondromas is not entirely known, but they are thought to be growth plate remnants, which are not resorbed and either persist unchanged or grow [1]. Enchondromas most often affect the cartilage that lines the inside of bones, and 50% of the cases are located on small tubular bones of the hands and feet. They are considered the most common primary bone tumor arising in the hands, occurring mainly in the proximal phalanx [2]. Lesions of the distal phalanx are uncommon, and this location has been shown to have a female predominance in small series [3, 4].

A single lesion is the most common presentation; however, multiple enchondromas should prompt the clinician to consider the possibility of Ollier disease (multiple enchondromatosis) or Maffucci syndrome (enchondromas and angiomatous skin lesions) [5]. Metachondromatosis should be suspected in patients with multiple

Fig. 3. Histopathologic findings. **a** Fragments obtained from surgical curettage showing a chondroid neoplasm. H&E. $\times 5$. **b** The neoplasm is composed of regular lobules of cartilage separated by fibrous tissue. H&E. $\times 20$. **c** The lobules are formed by hyaline carti-

lage with small single-nucleated chondrocytes. No mitoses are identified. H&E. $\times 50$. **d** Minimal ossification is present with focal increased cellularity, but no permeation of the normal bone trabeculae is seen. H&E. $\times 100$.

enchondromas and a family history of osteochondromas and/or enchondromas. These syndromes are associated with a higher risk of malignant transformation [6].

Enchondromas are usually asymptomatic, and these lesions can be followed clinically [4]. Conversely, if lesions affect the nail bed, pain can be present, and therefore, surgical excision is indicated. The aim of the surgical procedure should be complete excision to prevent recurrence. Presence of pain should also raise suspicion of either a pathologic fracture or malignant transformation into a chondrosarcoma. In this scenario, radiologic evaluation must be performed immediately [4]. The histologic

distinction between enchondroma and low-grade chondrosarcoma can be challenging for the pathologist, as the latter may lack definitive diagnostic atypical features. Because lesions replace normal bone with mineralized or unmineralized hyaline cartilage, enchondromas have a lytic appearance on radiographs. The presence of deep and extensive endosteal scalloping with involvement of soft tissue and permeation of surrounding bone marrow spaces favor the diagnosis of chondrosarcoma. Therefore, clinicopathologic and radiologic correlation are critical for the definitive diagnosis [7].

Statement of Ethics

The patient's consent was obtained.

Keywords

Enchondroma · Nail tumors · Pseudoclubbing

Disclosure Statement

None of the authors declare any financial support or relationships that may pose a conflict of interest regarding this paper.

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