Asymptomatic Longitudinal Pachyxanthonychia of the Fingernail

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Question

A 62-year-old white female without prior history of skin cancer presented with an asymptomatic white streak on her right fourth fingernail of 3 months' duration. On examination, a 3-mm white to yellow longitudinal nail plate thickening with ridging and transverse overcurvature was noted (Fig. 1). Dermoscopy revealed longitudinal white streaks and several proximal and distal splinter hemorrhages (Fig. 2). Proximal nail matrix biopsy revealed a fibroepithelial tumor consisting of digitate projections of typical but reduplicated nail matrix epithelium with fibrovascular cores (Fig. 3). The fibrovascular stroma was superficially cellular but relatively hypocellular and fibrotic deeply. Immunohistochemistry showed strong CD34 staining of the fibrovascular component (Fig. 4).

What is your diagnosis?

Fig. 1. Right fourth fingernail with localized nail thickening, xanthonychia, and longitudinal overcurvature.

Fig. 2. Splinter hemorrhages and longitudinal white lines seen on dermoscopy.

Fig. 3. Biphasic fibroepithelial tumor consisting of epithelial papillomatosis and multiple deep and thick epithelial ridges with superficially cellular and collagenized fibrovascular cores (hematoxylin-eosin, original magnification $\times 40$).

Fig. 4. Stromal spindle cells of the fibrovascular cores stain strongly positively for CD34 (original magnification $\times 40$).

Answer

Onychomatricoma

First described in 1992, onychomatricoma (OM) is a rare benign neoplasm that predominantly affects females in their late 40s to early 50s and presents twice as often on the fingernails than toenails [1, 2]. Its four cardinal clinical signs are nail plate thickening with ridging and xanthonychia, proximal splinter hemorrhages, longitudinal and transverse nail plate overcurvature, and, upon nail avulsion, a villous tumor originating in the nail matrix and perforating into the nail plate. In addition to longitudinal parallel white lines and parallel lesion edges, dermoscopic evaluation of OM shows specific findings on the free edge of the distal nail plate including plate thickening, dark dots, and pitting [2].

A practical approach to the diagnosis of OM involves consideration of tumors that clinically present as localized longitudinal thickening of the nail plate. Formally referred to as localized longitudinal pachyonychia (LLP), this nail band pattern may present as LLP melanonychia or, as depicted in the present case, LLP xantholeuconychia [3]. LLP is a distinct clinical finding that should prompt consideration of onychogenic neoplasms in the differential diagnoses. In addition to OM, these include onychocytic matricoma (OCM) and onychocytic carcinoma. Both OM and onychocytic carcinoma have been reported to present as LLP xantholeuconychia, in contrast with OCM, which more often presents as pachymelanonychia [4].

OM is a fibroepithelial tumor that recapitulates normal matrix epithelial histology [5]. Based on the tumor's anatomic localization and the evaluated planes of section, OM may have varying histomorphology that may present diagnostic challenges [4, 6]. When the tumor primarily involves the short apical matrix and the ventral surface of the proximal nail fold, the tumor is characterized on longitudinal sections by thick V-shaped keratogenous zones arising from deep epithelial invaginations. This epithelial architecture is associated with a thickened nail plate with an inferior border that terminates into unequal spurs [2, 6]. Transverse sections may show pseudocanals due to the breaking up of the roof of the OM cavities. Hence, apical matrix OM may clinically mimic a wart, nail unit squamous cell carcinoma in situ, and fibrokeratoma [6].

OM that primarily involves the ventral matrix is more common and has a characteristic plane-of-section-dependent histomorphology. Longitudinal sections reveal a single broad pedunculated fibroepithelial tumor with

papillomatosis of the matrical epithelium and multiple deep undulating ridges. This “gloved-finger” monodigitate pattern is in stark contrast to the morphology seen on transverse sections, which is characterized by deep epithelial ridges composed of basal and prekeratogenous cells that are centered on small empty cavities which correspond to areas of avulsed nail plate and keratogenous zones. Transverse sections of nail clippings show similar histologic findings, thus, providing a minimally invasive alternative to a nail matrix biopsy [7]. OM involving the ventral matrix may clinically mimic subungual fibromas and onychopapillomas.

Despite the architectural complexity of OM, which is a consequence of the bipartite organization of the nail matrix, the presence of a prominent fibrous stroma and keratogenous zone reduplication in OM permits its reliable distinction from OCM. OM's fibroblastic stroma stains positively for CD34 and may show wreath-like multinucleated cells (pleomorphic OM) and mucinous changes (myxoid OM) [8]. Aside from lacking a stromal component, OCM has a relatively flattened and relatively thicker basal epithelium in contrast to the undulating thin basal compartment seen in the acanthothic epithelium of OM [3]. Moreover, endokeratinization presents in OCM as whorls of prekeratogenous and keratogenous cells, which cornify with onychocytes. Onychocytic carcinoma, a rarely reported and perhaps controversial entity, and nail unit squamous cell carcinoma in situ are characterized by the presence of keratinocytic atypia, features that are not observed in OM [3, 4]. Complete excision, to include normal nail matrix proximal to the lesion, is generally recommended for the treatment of OM and prevention of its recurrence [9]. However, it is a benign entity and may be observed clinically.

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Statement of Ethics

The authors have no ethical conflicts to disclose. The patient's consent has been obtained.

Disclosure Statement

The authors have no relevant conflicts of interest to declare.

Keywords

Onychomatricoma · Nail pathology · Nail disorder

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