Onychomatricoma with Pterygium Aspect: Unusual Clinical Presentation

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Sir,

Onychomatricoma is a rare, benign tumour with typical clinical features, which appears to originate from cells of the nail matrix. It was first described by Baran & Kint in 1992 (1), who reported three cases, and subsequent reports have confirmed its existence. To date, about 31 cases have been reported. We report here a new clinical form of onychomatricoma arising from the matrix and the ventral part of the proximal nail fold.

CASE REPORT

A 60-year-old woman was referred to us for the evaluation of a nail lesion, which involved the whole nail plate of the fifth toenail of the right foot. She reported that the lesion had appeared progressively about 10 years before, after a violent trauma, which occurred after a bone fracture of the toe phalanx. Her past medical history was unremarkable.

Clinical examination revealed a yellow colouration along the entire length of the nail plate with splinter haemorrhages and marked thickening of the nail. The proximal nail fold was also thickened and appeared to be prolonged by a dorsal V-shaped formation "pseudo-pterygium", covering partially the nail plate and firmly attached to it (Fig. 1). An increased transverse curvature of the nail was also noted. The nails of all other toes and fingers were normal. Physical examination was otherwise normal. This onychodystrophy was painless, but caused aesthetic inconvenience. X-ray of the toe was normal.

A clinical diagnosis of onychomatricoma was made and surgical removal was decided.

Reflection of the proximal nail fold revealed fine filamentous digitations of the ventral part of the proximal nail fold, some of them were near the cuticles and seemed to be responsible of the pterygium aspect (Fig. 2). The avulsed nail plate showed the typical woodworm-like holes corresponding to the matrix digitations. Complete surgical excision was performed.

The diagnosis of onychomatricoma was confirmed by the histological study, which demonstrated a fibro-epithelial tumour encroached on the ventral face of the proximal nail fold and on the nail matrix. The thick nail plate, which was separated from the tumour, was perforated with multiple cavities filled with serous fluid. The ventral face of the proximal nail fold gave rise to a horny formation with acanthotic and papillomatous epithelium, with persistence of the granular layer. In its deep part digitations were embedded in a fibroblastic rich stroma containing fine collagen bundles with fine waxy nuclei oriented in different directions (Fig. 3). The matrix part of the tumour (distal part of the tumour on the proximal matrix) was made of glove-finger fibro-epithelial projections lined by matrix epithelium.

DISCUSSION

The case described here fulfils the criteria of onychomatricoma. However, some particularities were noted as a history of trauma 10 years ago, localization to a toenail, and bipolar tumoural proliferation from the ventral part of the proximal nail fold and the matrix.

Onychomatricoma is characterized clinically by a partial or total yellow discolouration of the nail plate with splinter haemorrhages, associated to ridging and overcuvature of the nail. Nail avulsion exposes a tumour emerging from the matrix region, and nail appears as a flat tunnel, storing filamentous digitations (1). Onychomatricoma more commonly affects fingernails of middle-aged individuals. Men and women are equally affected (2).

Perrin et al. (3) established histological criteria for the diagnosis of onychomatricoma: Onychomatricoma is a fibro-epithelial tumour consisting of two anatomic portions: a proximal zone which correspond to the base of the pedunculated fibroepithelial tumour and a distal zone which appears as multiple fibroepithelial projec-

Fig. 1. Thickened nail with pseudo-pterygium formation covering the nail plate and firmly attached to it (shown empty of blood).

Fig. 2. Reflection of the proximal nail fold revealed fine filamentous digitations of the ventral part.

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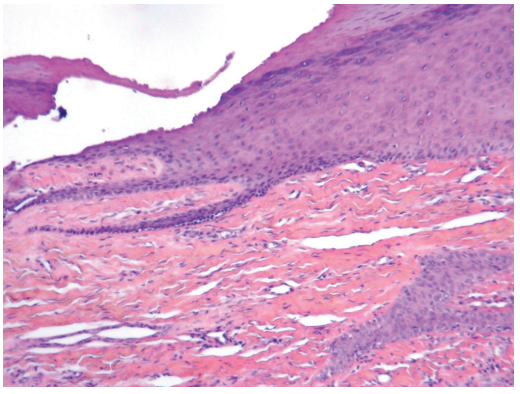

Fig. 3. Histology of the proximal nail fold with its lower part digitations in a fibrous stroma.

tions; these glove-finger-like digitations lined by matrix epithelium explain the multiple cavities observed in the thick nail plate. These authors describe a characteristic aspect of the stroma with two layers. One superficial layer highly cellular with thin wavy of fibroblasts oriented in different directions and a deep layer with less abundant fibroblasts accompanied by thicker collagen bundles oriented around the same horizontal axis. This two-layer stroma is more frequent in the proximal zone than in the distal portion.

Kint et al. (5) confirmed the nail matrix origin of the onychomatricoma by electron microscopy (5) but they showed that onychomatricoma differs from the normal matrix by the absence of a cornified envelope and a decrease in the number of tonofilaments. In contrast, Perrin et al. (4) showed by immunohistochemistry that the expression pattern of cytokeratins and integrins in onychomatricoma are identical to that observed in the normal nail matrix.

Fayol et al. (6) reported 5 cases of uncharacteristic features of onychomatricoma. Three of them presented as longitudinal melanonychia, another one had the appearance of a cutaneous horn. In 3 of the 5 cases, the tumour was associated with an onychomycosis and thus onychomatricoma may have been a predisposing factor in the secondary fungal infestation (6).

Perrin et al. (3), in their study of 12 cases of onychomatricoma, reported 2 cases of onychomatricoma that had the appearance of a cutaneous horn, situated in the lateral part of the nail apparatus. In these cases, histological examination showed an acanthotic and papillomatous epithelium, matricial in type with a thick keratogenous zone, giving rise to an orthokeratotic horny formation, similar to the nail plate, without any granular layer, whereas in our case, the acanthotic and papillomatous epithelium, showed the persistence of the granular layer.

Our case demonstrates that onychomatricoma can arise from a structure of the nail apparatus other than the nail matrix; it can occur also from the ventral part of the proximal nail fold in its distal part, near the cuticles. Onychomatricoma appears to be a tumour that actively produces the same structure of the tissue wherever it develops. In the matrix it produces nail plate substance, thus covering itself with a funnel of thickened nail plate and in the nail fold it simulates the structure of the nail fold (pseudo-pterygium).

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