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BRIEF REPORTS

Pigmented onychomatricoma: Four cases

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ABSTRACT

Onychomatricoma (OM) presenting as a longitudinal melanonychia is a very uncommon clinical presentation with very few cases in the literature. Our aim in this article is to report four cases of pigmented OM, and describe their clinical and dermoscopic findings and the importance of the differential diagnoses, especially with melanoma.

Key words: longitudinal melanonychia, nail, nail tumour, onychomatricoma, pigmented onychomatricoma.

LEARNING POINT

1. Pigmented OM is an unusual presentation of onychomatricoma.

2. Pigmented OM should be included in the differential diagnosis of longitudinal melanonychia.

HOW THE SUBMISSION ADDS TO CURRENT RESEARCH

It details the clinical and dermoscopic features of pigmented onychomatricoma as well as the importance of the differential diagnoses, especially with melanoma.

INTRODUCTION

The more common clinical signs of onychomatricoma are localised or diffuse thickening of the nail plate, a yellow longitudinal band of variable width, increased transverse overcurvature, the presence of holes in the distal margin of the nail plate, swelling of the proximal nail fold and multiple splinter haemorrhages in the yellowish nail.[1–3] Nevertheless, different clinical characteristics of OM have been published, such as longitudinal melanonychia, nail dystrophy, subungual haematoma and dorsal pterygium.[4,5] The aim of this article is to report four cases of pigmented OM and describe their clinical and dermoscopic characteristics and their differential diagnoses.

CASE 1

A 52-year-old female patient, skin photo type IV, presented with a 3-year history of an asymptomatic pigmented band on the right great toenail, which was previously treated unsuccessfully as onychomycosis with systemic terbinafine for 8 months. Trauma was the trigger for the appearance of the lesion. A physical examination revealed a 5-mm longitudinal melanonychia from the proximal nail fold to the free margin of the nail plate (consistent with pseudo-Hutchinson's sign), associated with a thickened nail plate, a yellowish discoloration and proximal nail fold oedema (Fig. 1a). Dermoscopy showed multiple cavities at the distal nail plate margin, longitudinal parallel white lines and splinter haemorrhages in the proximal nail plate (Fig. 1b). A mycological examination was negative. After partial removal of the nail plate, under distal block anaesthesia, we observed a pigmented nail matrix tumour with a digitiform aspect, characteristic of OM. Histological examination confirmed the diagnosis of pigmented OM (Fig. 1c).

CASE 2

A 31-year-old male patient, skin photo type III, presented with a 5-year history of an asymptomatic pigmented nodule on the fifth toe of the right foot. Trauma was also the trigger for the appearance of the lesion. A physical exami-

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Abbreviations:

OM	onychomatricoma

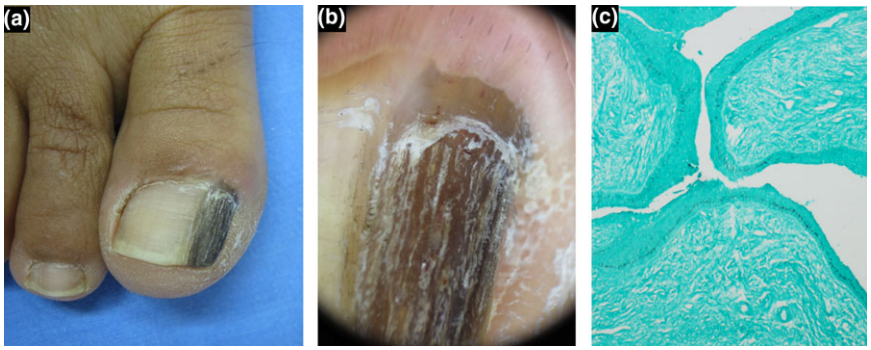

Figure 1 Right great toenail; (a) 5-mm longitudinal melanonychia; (b) dermoscopy showing longitudinal parallel white lines and splinter hemorrhages in the proximal nail plate; (c) Fontana–Masson stain of nail matrix biopsy showing an increased pigmentation of the nail unit epithelium with a normal number of melanocytes (×100).

nation revealed a total melanonychia of the fifth toe of the right foot from the proximal nail fold to the free margin of the nail plate, associated with a thickened nail plate, increased transverse overcurvature and swelling of the proximal nail fold (Fig. 2a). A mycological examination was negative. Nail surgery showed a villous tumour with hyperpigmentation of the nail matrix (Fig. 2b). Dermoscopic and histological features were compatible with the diagnosis of OM.

CASE 3

A 48-year-old male patient, skin photo type IV, presented with a 2-year history of an asymptomatic pigmented band on the third right finger. He referred to a trauma previous to the appearance of the hyperpigmentation. A physical examination revealed a 4-mm longitudinal melanonychia from the proximal nail fold to the free margin of the nail plate, associated with a thickened nail plate and swelling of the proximal nail fold (Fig. 3a). Potassium hydroxide preparation and fungal cultures were negative. After nail surgery, cavitations in the nail plate occupied by the fingerlike projections of a matrix villous tumour were seen (Fig. 3b). Dermoscopic and histological features were consistent with the diagnosis of OM.

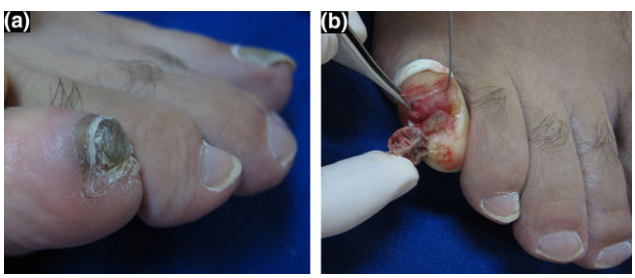

Figure 2 Fifth toe of the right foot, (a) total melanonychia, associated with a thickened nail plate, increased transverse overcurvature and swelling of the proximal nail fold; (b) after nail surgery, exposing a villous tumour and hyperpigmentation of the nail matrix.

CASE 4

A 45-year-old male patient, skin photo type IV, presented with a 3-year history of a painful pigmented band on the second toe of the right foot, with no previous history of trauma to the affected nail unit. A physical examination revealed a 5-mm longitudinal melanonychia from the proximal nail fold to the free margin of the nail plate, associated with a thickened nail (Fig. 4a,b). A mycological examination was negative. After partial removal of the nail plate, hyperpigmentation of nail matrix was observed (Fig. 4c). Dermoscopic and histological features confirmed the diagnosis of OM.

DISCUSSION

OM is a benign fibroepithelial tumour with CD34 expression in its connective compartment.[3,6] Fingernails are more often involved than the toenails; and the disease predominantly affects woman with a peak incidence in the 5th decade of life.[3,5]

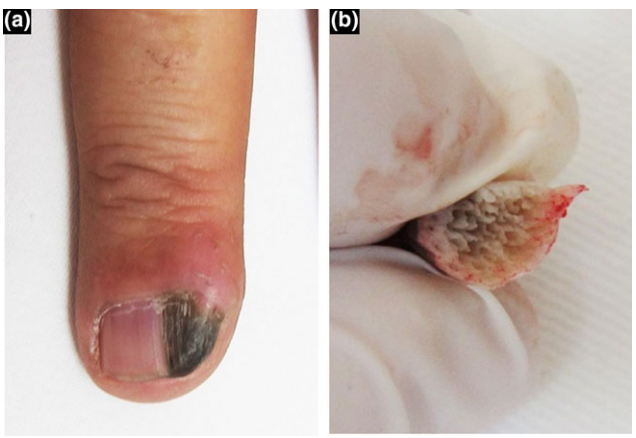

Figure 3 Third right finger, (a) 4-mm longitudinal melanonychia, associated with a thickened nail plate and swelling of the proximal nail fold; (b) after nail surgery, showing the cavitations in the nail plate occupied by the fingerlike projections of the matrix villous tumour.

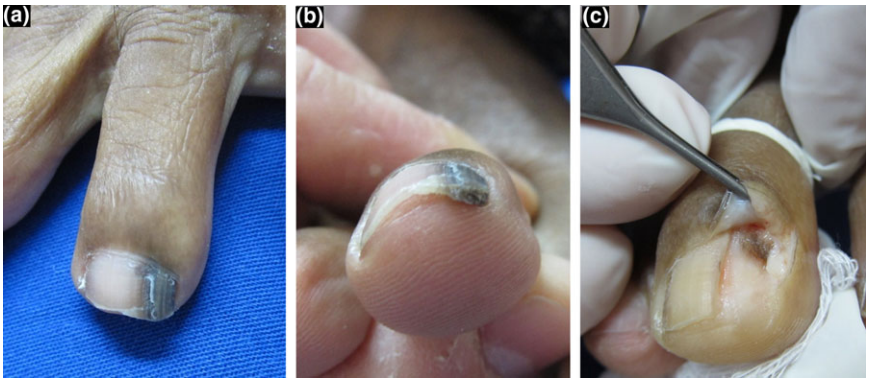

Figure 4 Second toe of the right foot, (a) 5-mm longitudinal melanonychia; (b) thickened nail plate and cavities at the distal nail plate margin; (c) after nail surgery, showing hyperpigmentation of the nail matrix.

The diagnosis of OM is made through the classical clinical signs and using complementary diagnostic methods, such as dermoscopy, ultrasound, magnetic resonance imaging, confocal microscopy, nail clipping and histopathology.[2,5]

Dermoscopy shows multiple cavities at the distal nail plate margin, longitudinal parallel white lines, parallel lesion edges, splinter haemorrhages, dark dots, nail pitting and thickening of the free edge.[6,7] High-frequency ultrasonography reveals the tumour lesion as an hypoechoic area affecting the nail matrix and an hyperechoic area corresponding to the finger-like projections.[5] With magnetic resonance imaging the portion that affects the nail matrix shows a centre with a low signal and a peripheral rim with a signal identical to that of normal epidermis; while the finger-like projections present a higher signal.[3] Electron microscopy has demonstrated basal cells in the proximal zone of the OM with a cytoplasmic rim containing mitochondria and tonofilaments.[4,8] Nail clipping histologically presents a thickened nail plate showing lacunae of different sizes and shapes filled with serous fluid and lined by a thin layer of epithelium.[9] Histopathological findings consist of two anatomical zones (proximal and distal) with three histological criteria for each zone.[3,6]

The main differential diagnoses of OM include fibrokeratoma of the nail matrix, onychomycosis, squamous cell carcinoma, Bowen disease, viral warts and nail fibroma.[2]

Only a few cases of pigmented OM have been described in the literature.[4] Pigmented OM presents with a longitudinal melanonychia associated with nail plate thickening. Some cases have also showed a yellow discoloration at the edge of the hyperpigmented band.[4] The differential diagnoses of pigmented OM is significantly different from classical OM, and includes traumatic nail changes associated with a haematoma, pigmented Bowen's disease, pigmented onychomycosis, pigmented onychopapilloma and subungual melanoma.[4,10]

Nail matrix melanocytes are usually quiescent; however, they may be activated by certain stimuli (onychomycosis, inflammation, tumours or systemic conditions).[10] In our cases, the melanonychia was confined to the tumoural lesion and the histopathology showed an increased pigmentation of the nail unit epithelium with a normal number of melanocytes.

The clinical features found in our patients were: longitudinal melanonychia with extension of the pigmentation to the cuticle and the proximal nail fold, thickening of the nail plate, a yellowish discoloration at the edge of the hyperpigmented band in all four cases, and proximal nail fold oedema in three cases. Dermoscopy showed multiple cavities at the distal nail plate margin, longitudinal parallel white lines and splinter haemorrhages in the proximal nail plate. All patients presented had a Fitzpatrick skin photo type III or IV, unlike what the literature says about the higher incidence in patients with a light skin colour.[2]

CONCLUSION

These cases describe an unusual presentation of onychomatricoma, with detailed clinical and dermoscopic description that can be used to distinguish it from other melanocytic lesions of the nail apparatus. As with classical OM, the diagnosis may be delayed because physicians are not familiar with this tumour. Pigmented OM should be included in the differential diagnosis of longitudinal melanonychia.

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