Onychocytic matricoma: Report of two cases and review of literature

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Abstract

Onychocytic matricoma (OCM) is a benign epithelial acanthoma derived from the nail matrix. It is a rare and relatively new disease of the nail first described in 2012, with only 14 cases published in English literature so far. Here, we reported the first two cases of OCM in Taiwan, presenting as longitudinal streaks of nail thickening with yellowish-white discoloration and various amount of splinter hemorrhages. A brief introduction of the pathological characteristics of various subtypes and clinical differential diagnoses were provided through literature review. OCM should be included among the differential diagnoses of longitudinal pachymelanonychia or xantholeucopachyonychia.

Keywords: Longitudinal pachyonychia, melanonychia, nail thickening, onychocytic matricoma, xantholeukonychia

Introduction

Onychocytic matricoma (OCM) is a benign but rare epithelial neoplasm arising from the nail matrix.¹ The common clinical presentations are longitudinal streaks of variegated blackish or yellowish nail thickening obscuring the lunula in a monodactylus distribution, which might be confused with other nail diseases presenting similarly as pachymelanonychia or xantholeucopachyonychia.¹²¹³ OCM has been classified into different pathological variants according to the various predominant components of matrical cells.¹³⁴ Since the first publication of this entity by Perrin et al. in 2012,¹¹ merely 14 cases have been reported in the English literature, among which only two cases are Asians; one in China and the other in Japan, respectively.¹¹³⁴ The rarity might partially be attributed to the under-recognition of this unique disease by clinicians and pathologists. We herein presented two cases of OCM with different histological subtypes from a tertiary medical center in northern Taiwan, along with a brief review of the literature.

Case Report

The first case was a 69-year-old woman, who presented with an asymptomatic, longitudinal, slightly thickened band over the radial aspect of right thumbnail for more than 2 years. Dermoscopy showed the band was 2 mm in width, extending from the proximal to the distal nail plate and obscuring the...
The lesion appeared variegated in color and comprised vague intermingled streaks of white, shades of yellow, tan, and dark brown. Splinter hemorrhages were identified under dermoscopy [Figure 1a].

A longitudinal fusiform excisional biopsy obtained from the nail matrix and proximal nail plate demonstrated the proliferation of basaloid cells over the distal nail matrix [Figure 1b]. A few round squamous eddy-like foci of keratinization composed of prekeratogenous and eosinophilic keratogenous zones were also identified [Figure 1b and c]. Human melanoma black-45 antigen (HMB-45) staining showed scarce melanocytes scattered over the nail matrix and proximal nail bed without evident proliferation. The Fontana-Masson staining revealed sparse, hardly identifiable melanin over the distal nail matrix and proximal nail plate. Based on the above-mentioned histological findings, the diagnosis of OCM, acanthotic type, is rendered.

The second case was a 63-year-old man with a yellowish-white, linear longitudinal thickened streak on the left thumbnail obscuring the lunula for more than 10 years [Figure 1d]. Subungual hyperkeratosis and transverse overcurvature at the radial side of the left thumbnail were found [Figure 1e]. The lesion had previously become inflamed but has remained asymptomatic for years thereafter. A partial nail avulsion was performed, which revealed a band-form subungual tumor adjacent to the ventral nail plate [Figure 1f], and the visible part of distal nail matrix [Figure 1g]. Excision of the remaining portion of subungual tumor down to the nail matrix was carried out subsequently.

Histopathological examination of the avulsed nail plate and attached subungual tumor revealed a digitated, papillary tumor forming a V-shaped proliferation of basaloid matrical epithelium and prominently thickened overlying layers of both prekeratogenous and keratogenous zones. Multiple cavities, onion-skin whorls of onychocytes, and melanin deposition were also identified in the overlying keratinized area and the inferior two-thirds of nail plate [Figure 1h]. The excised subungual tumor showed nail matrix with focally presence of thin papillary structures lined by acanthotic matrical epithelium [Figure 1i]. HMB-45 staining showed some scattered melanocytes over the matrical epithelium without evident proliferation. Fontana-Masson staining revealed some melanin deposition over the nail plate, the spheres of endokeratinization, and the digitated keratinized zones. Taken together, the histological findings were consistent with an acanthotic and papillomatous type of OCM.

**DISCUSSION**

OCM often presents clinically as longitudinal pachymelanonychia or xantholeucopachyonychia, namely, localized longitudinal thickening of the nail plate with...
blackish or yellowish-white discoloration. In the previous literature,[1,3,4] it mostly occurs among the middle-aged population, though the affected age group has been reported to range from 17 to 80 years. It shows a predilection for males (male:female = 10:4) and is more often seen on fingers than toes (finger:toes = 11:3), and on the right-side extremities opposed to the left (right:left = 9:5). The lesions could have persisted for 6 months to 10 years before a definite diagnosis was made [Table 1]. All the lesions were either asymptomatic or caused no specific complaint.

Several subtypes of OCM have been proposed according to both histological features (acanthotic type, acanthotic and papillomatous type, keratogenous type with retarded maturation, and germinotropic variant) and pigmentation (pigmented, melanocytic, and nonpigmented).[1,3,4] Within the literature, the acanthotic and papillomatous type is the most common subtype [Table 1]. The classification according to pigmentation, nevertheless, has rarely been adopted due to the lack of a precise definition, and the nonpigmented type has remained merely speculation by Perrin et al. up till currently, except one published case of “hypopigmented” type.[1,3]

Microscopically, OCM features acanthosis of the nail matrix, comprising basaloid compartment and various proportions of prekeratogenous and keratogenous zones according to histological subtypes. The acanthotic type is characterized by pronounced endophytic proliferation of basaloid cells, with interlaced pseudosquamous eddies, which are concentric whorls of endokeratinization composed of fairly balanced prekeratogenous, keratogenous zone, and central onychocytes, and located in the matrix epithelium or nail plate.[1] The acanthotic and papillomatous type exhibits similar pathological features as the acanthotic type at the proximal side of tumor, with additional epithelial digitations composed of predominant prekeratogenous zone projecting into the nail plate near the distal matrix. The overlying nail plate is often thickened with multiple tiny cavities, which is attributed to the loss of onychogenic capability at the tip of digitation.[1,8]

The keratogenous type is marked by retarded maturation of keratogenous zone and its consequent prominence within a large channel, starting from the proximal nail plate before its final differentiation in the distal end.[1]

| Author/published year | Case numbers | Age (years) | Gender (cases) | Clinical presentation (cases) | Lesion site (cases) | Time before diagnosis | Pathological variant |
|-----------------------|--------------|-------------|----------------|-------------------------------|---------------------|----------------------|---------------------|
| Perrin et al./2012[2][3] | 5 | 17-80 | Male=4 Female=1 | Longitudinal pachymelanonychia | Right 5th finger: 2 Right 3rd finger: 2 Right thumb: 1 | Unknown: 2 cases 6 months: 1 case 5 years: 1 case 6 years: 1 case | Acanthotic type: 2 cases Papillomatous type: 1 case Keratogenous type: 2 cases Pigmented type: 4 cases Melanocytic type: 1 case |
| Spaccarelli et al./2013[3] | 1 | 69 | Male | Longitudinal xantholeucopachyonychia and splinter hemorrhages | Right thumb | 3 years | Acanthotic and hypopigmented type |
| Wanat et al./2014[4] | 1 | 40s | Male | Longitudinal xantholeucopachyonychia | Right 2nd finger | 3 years | Keratogenous and pigmented type |
| Perrin et al./2016[5] | 4 | 36-71 | Male=2 Female=2 | Longitudinal pachymelanonychia: 2 Longitudinal xantholeucopachyonychia: 2 (Splinter hemorrhages in 1 case with xantholeucopachyonychia) | Left 1st toes: 2 Left 5th toe: 1 Left thumb: 1 | 1 year: 1 case 3 years: 1 case 5 years: 2 cases | Papillomatous type (pigmentation not specified) |
| Kusutani et al./2017[6] | 1 | 65 | Female | Irregular thickening of nail plate with diffuse nonhomogeneous melanin staining, nail scales and splinter hemorrhages | Right 4th finger | 15 years | Not specified in the article (pigmented type) |
| Song et al./2017[7] | 1 | 41 | Male | Longitudinal melanonychia, with a circular sinus beneath | Right 2nd finger | 10 years | Not specified in the article (pigmented type) |
| Perrin 2017[8] | 1 | 50 | Male | Longitudinal pachymelanonychia | Left thumb | 4 years | Germinotropic and melanocytic type |
| Present cases | 2 | 69 63 | Female Male | Longitudinal xantholeucopachyonychia with splinter hemorrhages | Right thumb: 1 Left thumb: 1 | 2+ years: 1 case 10+ years: 1 case | Acanthotic type: 1 case Papillomatous type: 1 case |
The germinotropic variant is a recently proposed histological subtype with predominant basaloid compartment constituting of two kinds of germinative cells displaying peripheral palisading resembling a basal cell carcinoma (BCC). In addition to the classical small cuboidal basaloid cells, there are another undifferentiated basaloid cells with large nuclei and small but prominent nucleoli. They do not present atypical mitoses and have aberrant expression of hair-related keratin (HK) 35, which is generally absent in the matrix of both normal nail and OCM. The characteristic spheres of prekeratogenous and keratogenous zone cells in the concentric arrangement are absent. The clinical differentiation from BCC could be achieved by the recognition of localized longitudinal pachyonychia in OCM. If the nail plate is avulsed before biopsy or excision, the lack of other typical features of BCC, including tumor-stromal clefting and staining for BerEp4 should allow for proper differentiation. Some other nail diseases presenting as longitudinal pachyonychia or longitudinal melanonychia/xantholeuconychia might pose a dilemma upon clinical diagnosis. These differential diagnoses included subungual irritated seborrhoeic keratosis, subungual Bowen’s disease, subungual melanoma in situ, onychopapilloma (OP; subungual keratosis of the nail bed), onychomatrixoma (OM; panonychoma fibropapilliferum), onychocytic carcinoma (OCC), and human papillomavirus (HPV)-induced lesions. Careful histological examination is usually sufficient to make the correct diagnosis. A brief comparison of their respective clinical and histopathological characteristics is provided in the following to aid in adequate differential diagnoses.

Acquired monodactylous longitudinal pachyonychia has been proposed as a unique and specific feature suggestive of onychogenic neoplasms, among which included OM, OCM, and OCC. While OCM presented more often as melanonychia, OM, and OCC were reported more frequently as xantholeuconychia. On histological examination, OM differs most from the other two by the fact it is a fibroepithelial tumor. OCM and OCC, in contrast, are pure epithelial neoplasms. Although OM, OCC, and acanthotic and papillomatous type OCM could all present with keratogenous digitations producing thickened nail plate and invaginations, the presence of a fibrovascular core within the villous projection, and a prominent fibrous stroma with fibroblast proliferation are consequently features distinctive of OM. The spheres with concentric arrangement of prekeratogenous and keratogenous zones within the matrical acanthoma and the nail plate are, on the other hand, a specific finding unique to mostly OCM and occasionally OCC. Reported as a malignant counterpart of OCM, OCC shares much histological similarities with OCM. The delimitation of the two relies on the presence of varying cytologic atypia, mitotic rate, and occasionally HPV-induced koilocytosis in the epithelium of OCC.[2,8,9]

Nail clipping has been proposed as a noninvasive alternative to differentiate the three types of onychogenic neoplasms.[2] While honeycombed cavities predominantly arranged in the inferior two-thirds of the thickened nail plate is a shared histological finding, the size of cavities might provide a diagnostic clue. The mean size of cavities decreased in the following sequential manner, with OM (mean size 1.13 mm × 0.49 mm) featuring large cavities, OCM comes in second (0.16 mm × 0.14 mm), and in situ OCC (0.075 mm × 0.08 mm) typically having cavities smaller than 0.1 mm.[2]

Subungual irritated seborrhoeic keratosis, though exceedingly rare, also presents as an epithelial acanthoma made up of basaloid cells in the nail matrix region. Due to irritation-induced activation, some basaloid cells mature focally into eosinophilic, flattened and apoptotic squamous cells in an onion-skin arrangement, known as squamous eddies. Similar to OCM as it may seem at first glance, true squamous eddies do not cornify with nail plate cells, and thus lack the concentric layers of prekeratogenous, keratogenous zones, and onychocytes as OCC possesses. The inflammatory cells are also more pronounced in irritated subungual seborrhoeic keratoses than OCM.[1-3,7,9,10]

OP has been reported seldom as longitudinal leukonychia (7 over 47 cases) and melanonychia (4 over 47 cases).[11] Histologically, it features subungual hyperkeratosis, and papillomatous acanthosis of the nail bed with matrix metaplasia.[1,11] The distinction from OCC could easily be drawn by the fact that OP is primarily a nail bed tumor.[1,3,7] As for subungual Bowen’s disease and subungual melanoma in situ, their recognition relies on cellular atypia of keratinocytes and melanocytes, respectively. Nail plate thickening due to active production by neoplastic matrical epithelium, and spheres of keratinization centered by onychocytes are also both not expected to be seen in either subungual Bowen’s disease or subungual melanoma.[12]

Immunohistochemical staining of OCM exhibits identical sequential expression of human keratins as the three layers in the normal matrix. The basaloid component is negative for HK, while the prekeratogenous zone presents HK85, and the keratogenous zone is positive for HK85, HK31, HK86, and HK34. As for the epithelial keratin K5 and K17, which are normally expressed in the matrical epithelium and nail bed, they are also diffusely positive in the basaloid epithelium and prekeratogenous zone of tumor. However, K6 and K16, normally limited in the nail bed, are diffusely present in the prekeratogenous zone of OCM. This has been hypothetically explained by the hyperproliferative nature of OCM.[9]

In conclusion, we reported two cases of OCM manifesting as longitudinal xantholeucopachyonychia with various amount of splinter hemorrhages. Clinicians should be aware of and raise suspicion of this rare onychogenic epithelial tumor which can be confirmed histopathologically.

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Conflicts of interest
There are no conflicts of interest.
REFERENCES

1. Perrin C, Cannata GE, Bossard C, Grill JM, Ambrossetti D, Michiels JF. Onychocytic matricoma presenting as pachymelanonychia longitudinal. A new entity (report of five cases). Am J Dermatopathol 2012;34:54-9.
2. Perrin C. Tumors of the nail unit. A review. Part II: Acquired localized longitudinal pachyonychia and masked nail tumors. Am J Dermatopathol 2013;35:693-709.
3. Spaccarelli N, Wanat KA, Miller CJ, Rubin AI. Hypopigmented onychocytic matricoma as a clinical mimic of onychomatrixoma: Clinical, intraoperative and histopathologic correlations. J Cutan Pathol 2013;40:591-4.
4. Perrin C. Germinotropic onychocytic matricoma: A new histopathologic subtype of onychocytic matricoma in the light of the microanatomy of the normal nail unit, with special reference to nail mesenchyme. Am J Dermatopathol 2017;39:e97-e101.
5. Song H, Qu F, Dang N, Sun X. Onychocytic matricoma: Report of an Asian case. Ann Dermatol 2017;29:355-7.
6. Kusutani N, Kamo R, Sowa-Osako J, Goto K, Ohsawa M, Yanagihara S, et al. Onychocytic matricoma as an underrecognized benign mimicker of subungual malignant melanoma and Bowen’s disease. J Dermatol 2017;44:e73-e74.
7. Wanat KA, Reid E, Rubin AI. Onychocytic matricoma: A new, important nail-unit tumor mistaken for a foreign body. JAMA Dermatol 2014;150:335-7.
8. Perrin C, Cannata GE, Langbein L, Ambrossetti D, Coutts M, Balaguer T, et al. Acquired localized longitudinal pachyonychia and onychomatrixomas: A comparative study to onychomatrixomas (5 cases) and onychocytic matricomas (4 cases). Am J Dermatopathol 2016;38:664-71.
9. Hancke E. Important malignant and new nail tumors. J Dtsch Dermatol Ges 2017;15:367-86.
10. Choi HJ, Yun SK, Kim HU, Ihm CW. Squamous eddies in irritated seborrheic keratosis. Am J Dermatopathol 2007;29:28-31.
11. Tosti A, Schneider SL, Ramirez-Quizon MN, Zaiac M, Miteva M. Clinical, dermoscopic, and pathologic features of onychopapilloma: A review of 47 cases. J Am Acad Dermatol 2016;74:521-6.
12. Perrin C, Cannata GE, Ambrossetti D, Patouraux S, Langbein L, Schweizer J. Acquired localized (Monodactylous) longitudinal pachyonychia and onychocytic carcinoma in situ (2 cases): Part II. Am J Dermatopathol 2017;39:40-4.