

Continuing Medical Education



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Onychomatricoma

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ABSTRACT

Onychomatricoma is a benign neoplasia that occurs specifically in the nail apparatus. It was described in 1992, and is the only tumor in which the change of the nail plate is actively caused by the lesion. It lies in the nail matrix, and has fingerlike projections that are embedded in the nail plate, resulting in thickening, longitudinal grooves, yellow discoloration and splinter hemorrhages. Although described as a rare tumor, it is believed to be underdiagnosed. The present study is aimed at reviewing onychomatricoma's clinical features as well as the complementary examinations that are used to recognize and diagnose this tumor.

Keywords: nail diseases; nails/pathology; neoplasms.

RESUMO

Onicomatricoma é neoplasia benigna específica do complexo ungueal descrita em 1992, sendo o único tumor em que a alteração da placa ungueal é produzida ativamente pela lesão. Encontra-se na matriz ungueal apresentando projeções digitiformes que ficam impressas na placa ungueal, resultando em espessamento, estrias longitudinais, coloração amarelada e hemorragias em estilhaço. Apesar de descrito como tumor raro acredita-se que seja subdiagnosticado. O objetivo deste artigo é revisar as características clínicas e os exames complementares subsidiários para o reconhecimento e diagnóstico desse tumor.

Palavras-chave: doença das unhas; unhas/patologia; neoplasias.

INTRODUCTION

Onychomatricoma (OM) is a benign tumor arising from the nail matrix. It was first described by Baran and Kint in 1992¹ as the first onychomatricoma. In 1995, Hanekee and Fränken, based on histological aspects, proposed the term onychomatricoma, which has been used since then.² Also, on a histological basis, other quotes and nomenclature, such as onychoblastoma, onychoblastic fibroma and atypical onychoblastic fibroma are currently found, however the term onychomatricoma remains the most frequently used and cited in the current literature.^{3,4}

It is considered a rare and specific tumor of the nail apparatus, characterized by the presentation of fingerlike projections from the matrix, and is the only tumor in which alterations of the nail plate are actively produced by the lesion.^{2,5}

Since its first description, just over 40 cases have been reported. Although considered a rare condition, its clinical, radiological, dermoscopic, histological, and electron microscopy-based aspects have been well documented. Recent studies consider the tumor's

genetic alterations, such as losses on chromosome 11.⁴ Its slow growth and the absence of pain in most cases explain patients' typical delay in seeking medical attention.

ETIOPATHOGENESIS

Although onychomatricoma's etiology is still not fully understood, trauma is considered the main predisposing factor. Another hypothesis is that it corresponds to a reactive picture and not to a matrix tumor.² Some authors suggest that onychomatricoma is an epithelial and conjunctive tissue hamartoma that mimics the nail matrix's structures.^{1,6,7}

CLINICAL PICTURE

Onychomatricoma affects mainly females (2,16:1), with the peak of incidence around the age of 51.⁸ It rarely affects children, with only one case described in the literature.⁹ Despite its prevalence in Caucasians, there are reports of involvement of other ethnicities 10 and one case described in a patient of African heritage.¹¹ It rarely causes pain and fingers are more affected than toes—this information is biased however, since there is greater patient concern and demand for a doctor's intervention for lesions that affect fingers.⁴

Onychomatricoma classically manifests with the clinical tetrad: (1) yellowish longitudinal band of variable thickness, (2) splinter hemorrhages preferentially affecting the proximal portion of the nail plate, (3) longitudinal and transverse hypercurvature of the nail plate, and (4) fingerlike projections that emerge from the nail matrix, leaving cavities in the nail plate.¹² (Figure 1)

Due to the fact that it is a matrix tumor, a nodule can be clinically observed in the proximal nail fold. Besides the clinical tetrad, onychomatricoma can present as longitudinal melanonychia (hypermelanosis), nail dystrophy, subungual hematoma, verrucous aspect in the proximal nail fold, dorsal pterygium, giant variant and normal type of pseudo-fibrokeratoma, in addition to the resemblance to 11 onychomycosis.^{13,14} (Figure 2) The main differential

diagnoses include subungual exostosis, fibrokeratoma, fibroma, onychomycosis, epidermoid carcinoma, Bowen's disease, keratoacanthoma, verruca vulgaris, acral superficial fibromyxoma, melanoma, bacterial infections, dermatofibrosarcoma protuberans, porocarcinoma, and osteochondroma.⁴

Onychomycosis has been implicated as a predisposing factor for the emergence of onychomatricoma (reactive theory of the lesion). On the other hand, onychomatricoma can also be considered a predisposing factor for onychomycosis.^{9,15}

DIAGNOSIS

In addition to the classic tetrad of signs, other methods such as dermoscopy of the nail plate,¹⁶ ultrasound^{17,18} MRI¹⁹ unguial clipping,²⁰ and histologic study,^{7,13,21} can be employed to aid in the diagnosis of onychomatricoma. (Figures 1 and 2)

The dermoscopy of the nail plate evidences perforations in the distal portion of the unguial plate,¹⁶ hemorrhagic striae, and white longitudinal grooves corresponding to the nail plate channels.⁵ (Figure 3)

Radiologically, there is no bone involvement linked to onychomatricoma.²⁰

Ultrasound has been shown useful in the tumor's detection, delimitation, and topography. For a good view of the lesion at this body site, the device's frequency should be set at seven to 15 MHz. The tumor is seen as a hypoechogenic area affecting the nail matrix and a hyperechogenic area corresponding to the fingerlike projections, in addition to having a low blood flow.¹⁸

With MRI, the tumor is easily seen in the sagittal cuts, affecting the nail matrix, with low signal capture, and resembling normal epidermis. At the distal section, the digitations are observed with high signal capture, as the mucoïd stroma present in the area of the tumor has a high concentration of water (T2). The axial cuts allow the viewing of perforations in the nail plate.¹⁹

Nail clipping is the histologic study of the nail plate in which the analyzed specimen is removed by cutting the distal part of the

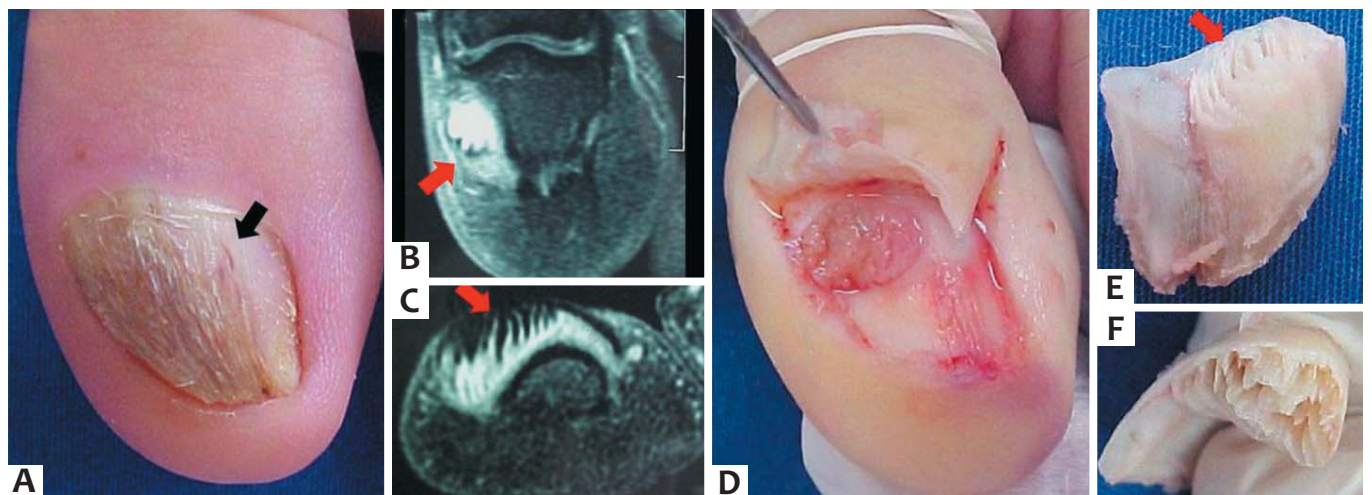


FIGURE 1: A) Thickening, yellowish color, longitudinal striation and small splinter hemorrhages in the nail plate (black arrow); b) longitudinal section; c) MRI cross-section showing a lesion with fingerlike projections (red arrows); d) tumor seen intraoperatively; e) bottom view; f) posterior view of the onychomatricoma's pathognomonic digitations impressions of the nail plate.

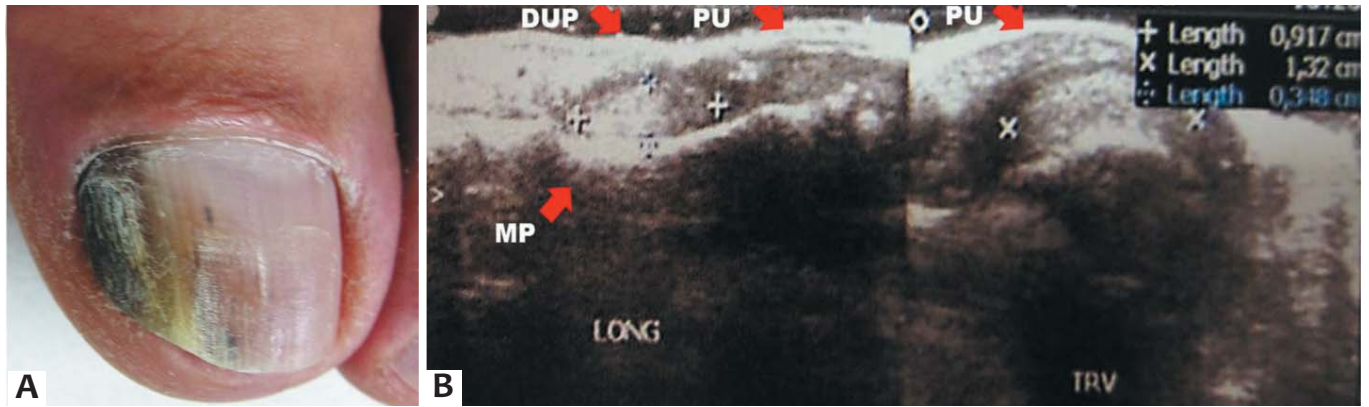


FIGURE 2: A) Nail plate thickened with yellowish color emulating onychomycosis. It is also possible to observe chromonychia suggestive of secondary infection by *Pseudomonas*; B) ultrasound showing tumor under the proximal nail fold, DUP longitudinal and transversal cuts: proximal nail fold; MP: proximal matrix; PU: nail plate; LONG: longitudinal view; TRV: transversal view

nail plate. This technique allows the visualization of the gaps (perforations) of different shapes and sizes, suggestive of onychomatricoma. The technique is deemed straightforward, fast, cost effective, and minimally invasive. In addition to allowing the diagnosis of the tumor, it provides assistance in the differential diagnosis of onychomycosis using PAS staining, and enables the immunohistochemical study of the nail plate.²¹

Diagnosis is confirmed histologically. It is a fibroepithelial tumor composed of two distinct areas: the proximal and distal zones. The first is located below the posterior nail fold and is characterized by deep epithelial invaginations filled by a thick keratin zone in the shape of a "V", well-defined fibrillar and fibrotic stroma, in addition to the thickening of the nail plate without cavities. The distal zone, which corresponds to the lunula region, is characterized by finger-like projections, perforations in the nail plate, and deep and poorly delimited penetration of the connective stroma in the dermis.⁴⁷

The fingerlike projections are formed by matrix epithelium, located around the connective tissue, in the antero-oblique axis, that proliferate and cause perforations in the nail plate, generating cavi-

ties, which in the distal part of the lunula lose their epithelium and are filled with serous fluid.⁴⁷

Some authors mention the presence of mast cells in the stroma of onychomatricomas.²² The keratinization zone can be adhered to the nail plate. Thus, both the tumor and the removed plate must be sent for histological examination.⁴ The splinter hemorrhages seen in the nail plate's proximal region, correspond to the fingerlike projections' loose vascular stroma.¹²

The yellowish color occurs due to the thickening of the nail plate, resulting from the layers of keratinization that involve the fingerlike projections. Its intensity is proportional to the degree of thickening.¹²

The main histological differential diagnoses are the fibrokeratoma and the unguis fibroma. In the longitudinal cuts of the onychomatricoma, the structure is reminiscent of a fibrokeratoma; nevertheless that diagnosis may be excluded due to the presence of multiple fingerlike projections, the absence of a cutaneous horn, and the presence of cavities filled with serous fluid in the distal portion of the nail plate. The onychomatricoma's stroma, located in the lunula, may suggest the diagnosis of fibroma, which in turn may be discarded by the hyperplastic and onychogenic nature of the epithelium. Moreover, unguis fibroma generates a longitudinal depression in the nail plate, in the shape of a channel, due to the compression in the nail matrix.¹²

The immunohistochemical study using the cell proliferation marker Ki67 (MIB-1) demonstrates a low rate of cell proliferation in the onychomatricoma.² The observed expression pattern of the cytokeratin and integrins is identical to those of the normal matricial epithelium, in spite of the fact that the antibody AE13—specific for the trichocystic keratin Ha 1-4—can be potentially useful as an onychomatricoma marker.²³

A study with adhesion proteins demonstrated the absence of beta-catenin in comparison with other unguis tumors.²⁴ The involucrin is expressed from the basal layer up to the upper part of the epithelium, where it is more pronounced and the transglutaminase-1 is limited.⁵ The immunophenotyping expresses CD34, but not CD99 or epithelial membrane antigen, S100 protein, actin, and desmin.⁵

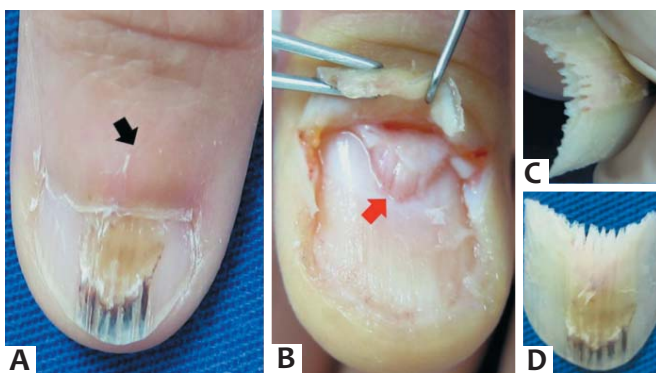


FIGURE 3: A) Bulging of the proximal fold (black arrow) and thickened nail plate with yellowish color and distally striated hemorrhages; B) intraoperative aspect of the tumor with fibrous digitations (red arrow); C) posterior view; D) ventral view of the nail plate with digitation impressions in the proximal region, thickening and central yellowish color, and striated hemorrhages close to the free border.

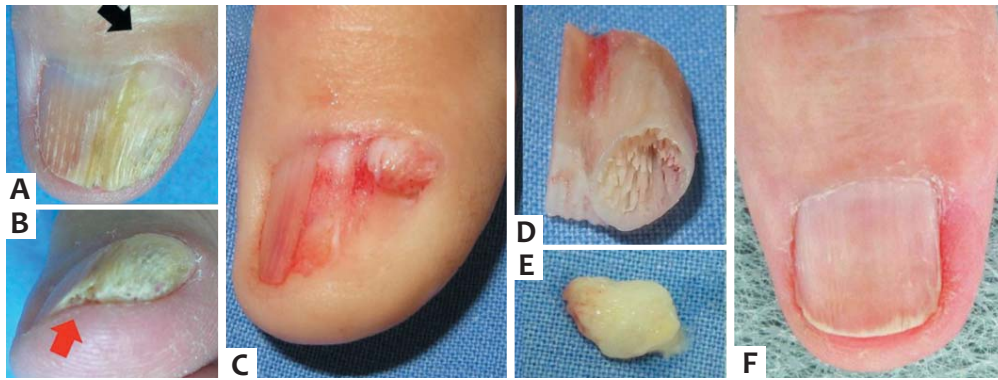


Figure 4: a) Bulging of the proximal fold (black arrow), thickened nail plate with yellowish color; b) thickening of the nail plate with longitudinal striae and visualization of cylindrical tunnels in the distal section (red arrow); c) Intraoperative aspect of the tumor with fibrous digitations; d) posterior view of the nail plate with fibrous digitation impressions; e) completely excised tumor; f) results after seven months

In electron microscopy, the basal cells apparently contain a reduced number of tonofilaments and desmosomes, which have a non-uniform development.⁸

TREATMENT

The treatment for onychomatricoma is surgical (Figure 4). After anesthesia (proximal or distal locking) the proximal nail fold is bent and the nail plate is gently removed in order to prevent the villi from being torn. The tumor must be removed completely. Nail dystrophy after surgical removal can occur depending on the preservation of the nail matrix during the removal of the tumor.²⁵ ●

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Questions for continuing medical education – CME

1) About onychomatricoma it is correct to state, except for:

- a) the avulsion of the nail plate is diagnostic
- b) it is a benign tumor arising from the nail matrix
- c) it is common in people of African origin and in children
- d) it is usually painless and predominantly affects fingers
- e) it can manifest with nodules in the proximal nail fold

2) The below are considered clinical signs of the onychomatricoma's tetrad, except for:

- a) yellowish longitudinal strip of varying thickness
- b) splinter hemorrhages
- c) longitudinal and transverse hypercurvature of the nail plate
- d) blackened periungual pigmentation
- e) fibrous digitations that emerge from the nail matrix

3) Select the correct choice regarding the clinical signs that may arise in onychomatricoma:

- a) nail dystrophy
- b) melanonychia
- c) nail bleeding
- d) verrucous aspect in the nail fold
- e) all of the above are correct

4) About the use of nail clipping for the diagnosis of onychomatricoma, we can say:

- a) it is a fast, straightforward, cost effective and minimally invasive examination
- b) it is an old examination, which is not carried out anymore
- c) it does not allow the visibility of the nail plate's perforations due to the tumor
- d) it allows for ruling out other tumors of the nail matrix
- e) it allows the histologic study of the nail plate, however it does not allow the performing of an immunohistochemistry analysis

5) Among the onychomatricoma's differential diagnoses is:

- a) Bowen's disease
- b) Subungual exostosis
- c) Melanoma
- d) Onychomycosis
- e) all of the above are correct

6) Select the incorrect answer:

- a) the visualization of fibrous digitations emerging from the nail matrix and the cavities produced by them in the nail plate are features of onychomatricoma.
- b) ultrasonography is not an auxiliary method in the diagnosis of onychomatricoma
- c) it is the only tumor in which alterations in the nail plate are actively produced by the lesion
- d) the main histological differential diagnoses are the fibrokeratoma and the nail fibroma
- e) the treatment for onychomatricoma is surgical

7) Regarding the examinations used in the diagnosis of onychomatricoma, select the incorrect item:

- a) ultrasonography can delineate the tumor
- b) it is possible to see the splinter hemorrhages in the proximal portion of the nail through dermoscopy
- c) digital projections can be visualized in MRI
- d) X-Ray is of utmost importance in the diagnosis of onychomatricoma
- e) Nail clipping allows performing PAS

8) Which of the following findings is most suggestive of onychomatricoma?

- a) white longitudinal striations
 - b) longitudinal depression in the shape of channels
 - c) pyogenic granuloma in the nail edge
 - d) longitudinal perforations of the nail plate
 - e) Hutchinson sign
- 9) The onychomatricoma's histology evidences:
- a) basaloid cells with myxoid stroma in the dermis
 - b) matrix proliferative epithelium that invades the nail plate
 - c) hyperkeratosis with parakeratosis and presence of koilocytes
 - d) atypical keratinocytes in the nail bed
 - e) atypical melanocytes with increased mitotic activity and dermal invasion

10) Select the right option regarding the treatment of onychomatricoma:

- a) surgery is the treatment of choice
- b) the proximal matrix must be removed in order to prevent recurrence
- c) nail dystrophy occurs invariably in the postoperative
- d) the complete removal of the nail apparatus is performed during the surgery
- e) the treatment may be clinical

Key:

Hormone in the rejuvenation: a review of its real effectiveness. *Cosmet Dermatol Surg* 2012; 4(4):322-30.

1 d 2 b 3 c 4 d 5 b 6 a 7 c 8 a 9 d 10 c

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