Case Reports

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Giant Onychomatricoma: a case report

Onicomatricoma gigante: relato de um caso

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ABSTRACT

Introduction: Onychomatricoma is a rare benign tumor characterized by a clinical tetrad of a yellowish longitudinal band of varying thickness, splinter hemorrhages, longitudinal and transverse hypercurvature of the nail plate, and digitiform projections emerging from the nail matrix, leaving cavitations in the nail plate. We report a case of a female patient, 49 years old, with a history of a tumoral lesion in the third left toe for about four years, with a progressive increase in this period. Physical examination revealed an exuberant tumor lesion clinically compatible with onychomatricoma, confirmed by pathological examination after complete tumor excision.

Keywords: Nails; Nail diseases; Neoplasms

RESUMO

Introdução: *Onicomatricoma é um tumor benigno, raro, caracterizado pela tétrade clínica de faixa longitudinal amarelada de espessura variável, hemorragias em estilhaço, hipercurvatura longitudinal e transversa da placa ungueal e projeções digitiformes que emergem da matriz ungueal, deixando cavitações na placa ungueal. Relatamos um caso de paciente feminina, 49 anos, com queixa de lesão tumoral em terceiro quirodáctilo esquerdo há cerca de quatro anos, com aumento progressivo no período. Ao exame, apresentava lesão tumoral exuberante clinicamente compatível com onicomatricoma, confirmado pelo exame anatomopatológico após exérese completa do tumor.*

Palavras-chave: *Doenças da unha; Neoplasias; Unhas*

INTRODUCTION

Onychomatricoma is a rare benign tumor characterized by digitiform projections from the matrix, being the only tumor in which the lesion actively produces nail plate alteration.[1,2] We report a case of an onychomatricoma presenting an exuberant aspect, highlighting the clinical characteristics and the diagnosis of the lesion.

Case Report

A 49-years-old woman, who works in the shoe industry, presented at the Dermatology Department with a tumoral lesion in the third left toe for about four years, with a progressive increase in this period. The patient denied pain or other injury-related symptoms. She had undergone several treatments for onychomycosis, without improvement. Physical examination revealed nail plate thickening, transverse hypercurvature, xanthonychia, hemorrhagic splinters, and small holes in the nail's free edge (Figures 1 and 2). Dermoscopy of the blade surface allowed better visualization of the hemorrhagic splinters, and the free edge showed the presence of perforations (Figures 3 and 4).

Due to suspected onychomatricoma, an excisional surgery of the lesion was performed. Two incisions were made in the proximal nail fold, which was rebound, exposing the tumor. The nail excision allowed the visualization of digitiform projections in the matrix area (Figures 5 and 6). We performed complete tumor excision and primary closure of the incisions in the proximal nail fold (Figure 7). The histopathologic exam demonstrated squamous epithelial digitiform hyperplasia forming projections, confirming the diagnosis of onychomatricoma (Figure 8).

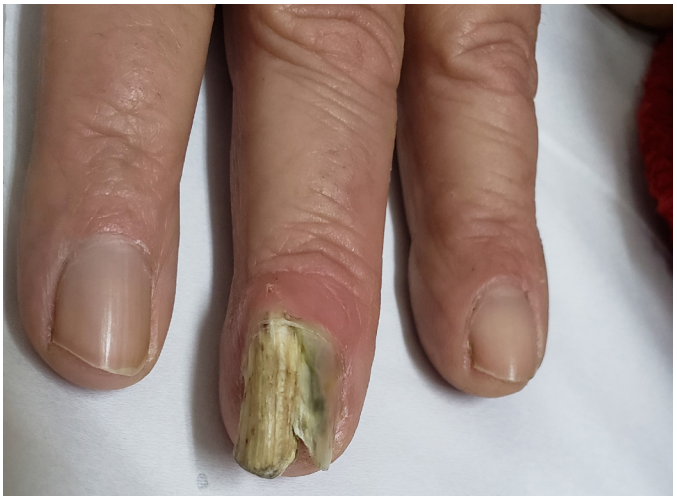

Figure 1: Nail plate thickening, transverse hypercurvature, xantonychia

Figure 3: At dermoscopy examination, hemorrhagic splinters

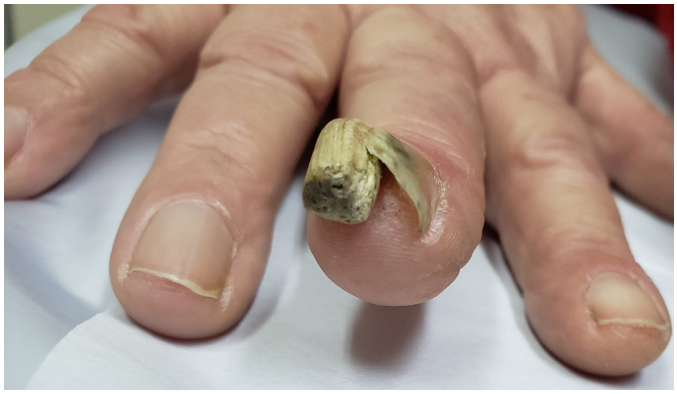

Figure 2: Nail plate thickening, transverse hypercurvature and small holes in the nail's free edge

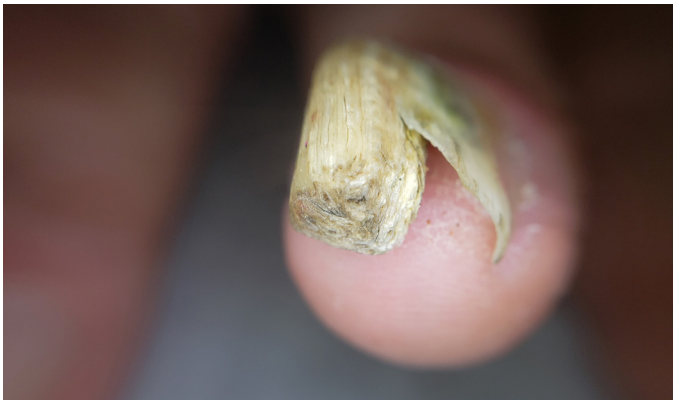

Figure 4: Dermoscopy of the nail's free edge: perforations

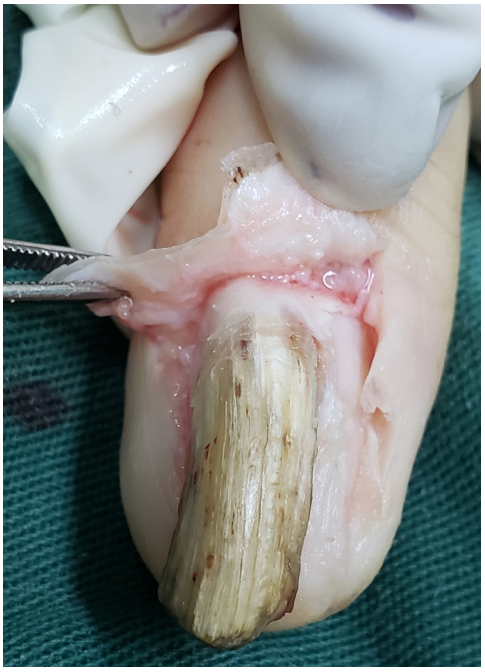

Figure 5: Transoperative: folding of the proximal nail fold, exposing the tumor

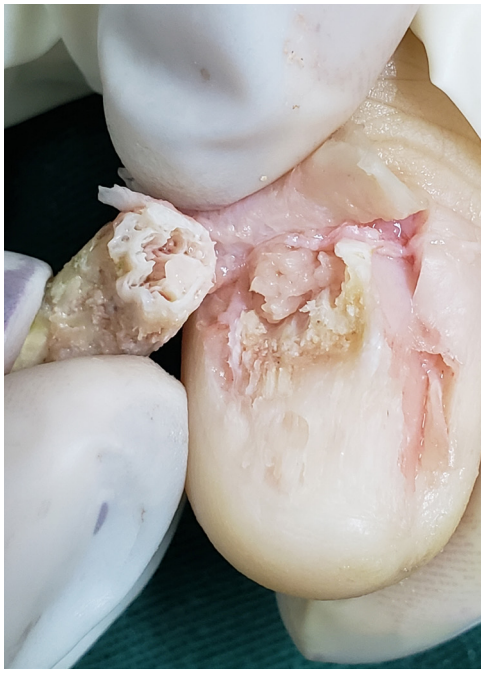

Figure 6: Transoperative: digitiform projections

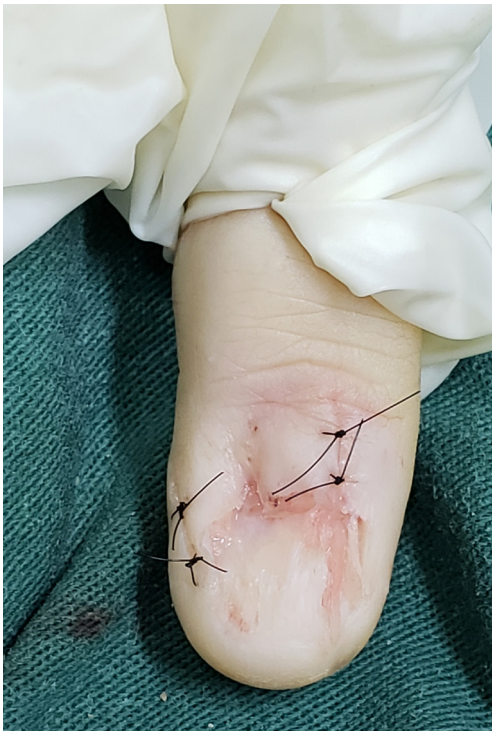

Figure 7: Primary closure of the incisions

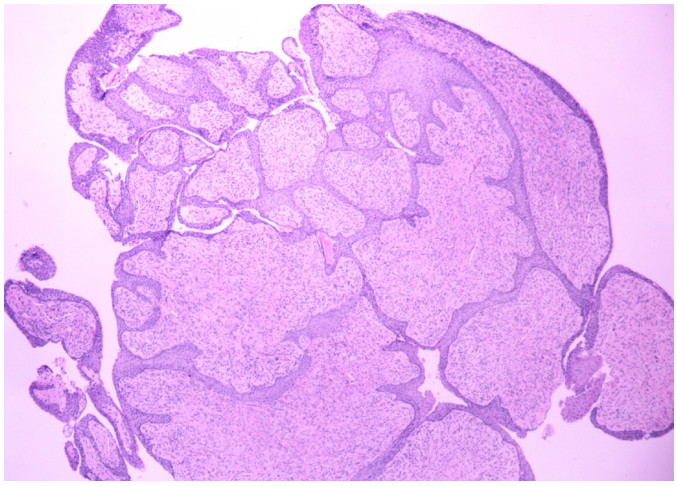

Figure 8: The histopathological examination demonstrated squamous epithelial digitiform hyperplasia forming projections (HE, 4x magnification)

DISCUSSION

A clinical tetrad characterizes the onychomatricoma, consisting of a yellowish longitudinal band of varying thickness, splinter hemorrhages, longitudinal and transverse hypercurvature of the nail plate, and digitiform projections emerging from the nail matrix, leaving cavitations in the nail plate.[3,4] It may also manifest with longitudinal melanonychia, subungual hematoma, nail dystrophy, proximal nail fold verrucosity, dorsal pterygium, giant variant, pseudofibrokeratoma type, and may have characteristics similar to onychomycosis.[4,5] Plaque dermoscopy demonstrates perforations in the distal portion of the nail plate, hemorrhagic striae, and white longitudinal grooves corresponding to the nail plate channels.[1,4,6]

Differential diagnoses include subungual exostosis, fibrokeratoma, vulgar wart, onychomycosis, squamous cell carcinoma, keratoacanthoma, superficial acral fibromyxoma, melanoma, bacterial infections, dermatofibrosarcoma protuberans, porocarcinoma, and osteochondroma. Onychomycosis is cited as a

predisposing factor for the emergence of onychomatricoma (reactive lesion theory). On the other hand, the tumor can also be considered a predisposing factor for onychomycosis.[6,7]

Imaging exams such as radiography, ultrasound, and MRI may help in the diagnosis of the lesion. Still, in most cases, the clinical and dermoscopic examination associated with the pathological examination is sufficient for the diagnosis, as in the case reported.[8,9] Anatomopathological examination demonstrates a fibroepithelial tumor composed of two portions. The first portion, proximal, is located under the posterior nail fold, characterized by epithelial invaginations filled with a thick V-shaped keratinized zone, well-defined fibrillar and fibrocystic stroma, and thickening of the nail plate. Digitiform projections, perforations in the nail plate and deep, poorly delimited penetration of the connective stroma in the dermis characterize the second portion, the distal area in the lunula.[10]

The treatment of onychomatricoma is surgical, and a complete tumor excision should be performed. The long-term prognosis is generally favorable, but nail dystrophies are common.[11]

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