Magnetic Resonance Imaging in the Diagnosis of Onychomatricoma: A Case Report

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Established Facts

- Onychomatricoma (OM) is a rare benign nail matrix tumour with less than 200 cases reported in the literature.
- The aspect of OM on magnetic resonance imaging (MRI) scans is typical and specific but MRI is rarely performed due to this indication.

Novel Insights

- To our knowledge, only 5 reported cases had an MRI performed due to this indication.
- MRI should be considered for dystrophic nails with negative mycological tests in order to decrease the diagnosis delay and avoid useless antifungal treatments.

Keywords

Onychomatricoma · Nail matrix · Nail tumour · Magnetic resonance imaging

Abstract

Onychomatricoma (OM) is a rare benign tumour of the nail matrix characterized by specific clinical and histologic features. The main clinical signs are thickening of the nail plate, xanthonychia, overcurvature of the nail plate, and multiple splinter haemorrhages. The diagnosis is based on clinical, radiological and histopathological findings. Histologically, the tumour is characterized by filiform epithelial projections.

The objective of this study is to present the first reported Tunisian case of OM, focusing on the contribution of magnetic resonance imaging to the diagnosis of OM. A review on the subject is also presented.

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Introduction

Onychomatricoma (OM) is a rare benign fibroepithelial tumour of the nail matrix that was first described by Baran and Kint [1] in 1992 and later coined by Haneke and Fränken [2] in 1995. Since the original description,

Fig. 1. Clinical aspect: thickening, overcurvature, xanthonychia, and swelling of the proximal nail fold.

Fig. 2. MRI showing the typical finger-like projections and thickening of the periungual area.

no more than 200 cases were reported in the literature. This tumour is characterized by finger-like projections that invade the nail plate and clinically the affected digit typically shows a thickened nail plate, xanthonychia, an increased transverse overcurvature and multiple splinter haemorrhages [3]. In most cases, this lesion is asymptomatic with slow progression. Diagnosis is based on clinical, radiological and histopathological findings. The treatment is surgical, and the tumour must be removed completely [4].

In this report, we present the first reported Tunisian case of OM, focusing on the contribution of magnetic resonance imaging (MRI) to the diagnosis of this tumour. A review of the subject is also presented to provide an updated overview of this rare and often misdiagnosed pathology.

Case Report

A 46-year-old female patient had a 2-year history of nail dystrophy affecting the nail of her right great toenail. She had neither a significant medical history nor a history of trauma. Clinical examination showed swelling in the proximal nail fold and a thickened yellowish toenail with overcurvature of the nail plate (Fig. 1). The lesion was pain-free. A mycological examination was requested but was negative. Because of the clinical aspect, the patient was treated with oral antifungal agents for months without amelioration. An X-ray was performed excluding any bone involvement. A preoperative MRI was carried out and showed infiltration and thickening of the periungual area of the great toe enhanced after injection of gadolinium. It also showed finger-like projections of

the nail matrix (Fig. 2). The diagnosis of OM was suspected, and the nail was surgically removed (Fig. 3, 4). The tumour at the nail matrix was excised. Histology revealed a well-vascularized fibroepithelial lesion, with noticeable exophytic papillary structures, covered by regular squamous epithelium, discreetly keratinized in places with a sparse presence of koilocyte-like cells. With the clinicopathological correlation, a definitive diagnosis of OM was established. After 2 months of follow-up, the patient evolved with no recurrence and no secondary infection.

Discussion

OM is a benign fibroepithelial neoplasm rarely reported in the literature, but significant progress has been made over the past years in the diagnosis of this lesion.

Most presentations affect the fingernails of middle-aged Caucasian women and involve the third or second finger of the dominant hand [5]. However, recurrence of OM in toenails could be underreported due to the clinical similarity with onychomycosis. When it reaches toenails, most cases were found in the big toe. It is rare in non-Caucasian descendants. There has been only one presentation in a child [6].

The long latency of OM is explained by its slow growth, the absence of symptoms and its misdiagnosis (often confused with onychomycosis).

The typical clinical tetrad of OM can be associated with wood-worm like (or honeycomb-like) cavities in the distal margin of the nail plate. A nodular tumefaction at the nail base can be associated as well. Many variants of

Fig. 3. Digitations emerging from the matrix after nail avulsion.

Fig. 4. Characteristic cavities of the OM in the proximal portion of the nail plate after nail avulsion.

OM are described; especially pigmented forms creating concern for melanoma of the nail matrix and prompting a biopsy [7], but also giant forms and forms associated with pterygium or with a cutaneous horn [8–10].

Diagnosis is made clinically in most cases where the typical nail modifications are present, but in unusual presentations, additional diagnostic methods may be helpful and include dermoscopy, radiography, ultrasonography, confocal microscopy, MRI, nail clipping, and histopathologic examination.

Dermoscopy typically shows perforations in the distal portion of the nail plate, haemorrhagic striae, and white longitudinal grooves corresponding to the nail plate channels [11, 12].

High-resolution ultrasound reveals a hypoechogenic tumour in the nail matrix with hyperechogenic finger-like projections [13]. Colour Doppler sonography can show a hypovascular pattern within the lesions [14].

MRI is typical but is rarely performed; it shows a tumour emerging from the nail matrix: the proximal portion of the lesion shows low-signal intensity and the finger-like projections distally show high-signal intensity. The digitations into the nail plate appears with a Y-shape and with a high-intensity signal, probably due to a concentration of water higher than that surrounding nail,

and axial images can show the holes in the nail plate and the tumoural digitations [3, 13, 15]. To the best of our knowledge, there are no other lesions reported in the literature, neither benign nor malignant, that have the same presentation as OM on MRI. However, in atypical presentations or clinical signs of malignancy, the diagnosis has to be confirmed by histopathology [3, 15, 16].

Confocal microscopy was used in 4 cases [17] and seemed useful in the preoperative diagnosis of OM. It showed longitudinal dark areas and bright/grey lines, forming channel structures within the distal intermediate nail plate. It also showed the splinter haemorrhages.

More recently, nail clipping was used as an easy and minimally invasive method to help with the diagnosis and to exclude fungal infections by a PAS stain. It showed a thickened nail plate with lacunae of different sizes and shapes filled with serous fluid and lined by a thin layer of epithelium [18].

The treatment consists of a complete surgical excision of the tumour. Nail avulsion exposes a villous tumour emerging from the matrix that is evocative of a sea anemone with its characteristic digitations in a thickened bored nail with multiple cavities and extending into the proximal nail. The tumour should only be shaved from the underlying matrix [19, 20].

The histopathological examination is unique and describes the OM: the fibroepithelial lesion has two zones. The proximal zone is characterized by deep vertical epithelial invaginations centred on empty V-shaped cavities. It is dome-shaped in transverse sections and is lined with a papillomatous matrix-type epithelium. The distal zone is characterized by multiple "glove finger" papillary projections covered by a matrix-type epithelium that is devoid of a stratum granulosum and that keratinizes through an eosinophilic keratogenous zone [3, 19, 21]. In our case, histopathology was typical but also showed a sparse presence of koilocyte-like cells, which has not been reported for OM in the literature.

Immunohistochemical analysis is rarely performed but it can help distinguish histologic variants of OM from other lesions. Indeed, the tumour differs from other lesions by expressing CD34 but not CD99 [22, 23].

In conclusion, OM remains a rare tumour but should be considered as a differential diagnosis for dystrophic nails that do not respond to antifungal treatment. Clinical suspicion must be confirmed by histopathology, but this report also draws attention to diagnostic imaging examinations as an ally for diagnosis, especially MRI, which can reveal the typical finger-like projections.

Statement of Ethics

The authors have no ethical conflicts to disclose. The patient has given her consent to publish photos and details of the case.

Disclosure Statement

The authors have no conflicts of interest to declare.

Author Contributions

Dr. Olfa Charfi wrote the first draft of the manuscript. Dr. Kahena Jaber contributed to conception and design, analysis and interpretation of data, and revision of the article. Dr. Ferdaous Khammouma contributed to the interpretation of the data and the revision of the article. Dr. Faten Rabhi contributed to the interpretation of the data and the revision of the article. Dr. Soumaya Youssef contributed to the interpretation of the data and the revision of the article. Dr. Raouf Dhaoui contributed to the interpretation of the data and the revision of the article. Dr. Nejib Doss contributed to conception and design, analysis and interpretation of data, and revision of the article.

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