Hand Surgery and Rehabilitation 38 (2019) 386–389

Available online at
ScienceDirect
www.sciencedirect.com

Elsevier Masson France
EM|consulte
www.em-consulte.com

Case report

# Pigmented onychomatricoma may originate from a childhood trauma: A case report

## *L'onychomatricome pigmenté peut provenir d'un traumatisme durant l'enfance : à propos d'un cas*

S. Madi [a], Z. Meguerian [b], V. Kanj [c], J. Bakhach [d,*]

[a] *Department of plastic & reconstructive surgery, St. George Hospital University Medical Center, P.O. Box 166 378, Achrafieh, Beirut, Lebanon*
[b] *Department of anatomic pathology laboratory, St. George Hospital University Medical Center, P.O. Box 166 378, Achrafieh, Beirut, Lebanon*
[c] *Department of diagnostic radiology, St. George Hospital University Medical Center, P.O. Box 166 378, Achrafieh, Beirut, Lebanon*
[d] *Department of plastic & reconstructive surgery, hand & microsurgery, American University of Beirut Medical Center, Cairo street, Beirut, Lebanon*

ARTICLE INFO

*Article history:*
Received 4 April 2019
Received in revised form 18 August 2019
Accepted 10 September 2019
Available online 21 September 2019

*Keywords:*
Pigmented onychomatricoma
Nail disease
Benign nail tumor
Crush injury
Childhood

*Mots clés :*
Onychomatricome pigmenté
Pathologie unguéale
Tumeur bénigne de l'ongle
Traumatisme
Enfance

ABSTRACT

Pigmented onychomatricoma (OM) is a very rare benign fibroepithelial tumor of the nail matrix. We report the case of a 23-year-old Lebanese man with 15-year history of nail plate dystrophy with longitudinal ridging, yellowish discoloration, excessive transverse curvature and late-onset melanonychia along the medial third of the right thumb nail. Excisional biopsy was performed and confirmed OM. We outline the clinical history, radiological and histopathological findings as well as the surgical and reconstructive technique of this unusual case of OM. The age group, history of crush injury, and pigmentation of the nail plate make of this rare form of ungual tumor an interesting case report.


RÉSUMÉ

L'onychomatricome pigmenté (OMP) est une tumeur bénigne très rare de la matrice fibroépithéliale de l'ongle. Nous rapportons le cas d'un Libanais de 23 ans ayant depuis 15 ans une dystrophie unguéale avec des crêtes longitudinales, une décoloration jaunâtre, une courbure transversale plus prononcée et l'apparition tardive d'une bande mélanique le long du tiers médial de l'ongle droit du pouce. La biopsie-exérèse a montré un OMP. Nous décrivons l'histoire clinique, radiologique, histopathologique et les techniques chirurgicales et reconstructives de ce cas d'OMP. L'âge, l'anamnèse et la pigmentation de la tablette unguéale font de cette forme rare de tumeur de la tablette unguéale un cas intéressant.


## 1. Introduction

Tumors of the nail unit are often misdiagnosed clinically. Onychomatricoma (OM) is a rare benign neoplasm that can present with misleading clinical and histological features. OM is less commonly association with both trauma to the nail apparatus and onychomycosis. This case illustrates a condition in which both of these findings are present.

## 2. Case report

A 23-year-old man with no past medical history was concerned about the appearance of his right thumb nail as it had recently gained dark pigmentation. Fifteen years prior to consulting with us, his thumb nail had sustained a severe injury when it was crushed by a closing door. At that time, the nail was completely avulsed, and the wound was sutured by a primary care physician. Twelve months later, the regrowing nail became dystrophic, thickened and had a yellowish discoloration. This dystrophy and discoloration persisted over time. A few years prior to our consultation, the nail developed a melanonychia band that was

* Corresponding author at: Department of plastic & reconstructive surgery, hand & microsurgery, American University of Beirut Medical Center, Cairo street, Beirut, Lebanon.

https://doi.org/10.1016/j.hansur.2019.09.002
2468-1229/

centered around the dystrophic region. Secondary infection by Pseudomonas was suspected and the patient was treated empirically with oral ciprofloxacin for 6 weeks with no improvement.

Upon clinical examination, there was a rough 3 mm ridge along the medial aspect of the right thumb's nail plate. In the frontal plane, the nail had an excessive transverse curvature; this was comprised of woodworm-like cavities of the medial free edge and yellowish, discrete, longitudinal streaks extending along the medial side of the nail plate, with well-demarcated borders measuring 5 mm in width. The overlying nail plate also had a melanonychia band that was condensed at the far distal nail bed and reached the proximal nail fold. Splinter hemorrhage could not be seen due to the dominating melanonychia. There was mild-to-moderate tenderness over the medial side of the hyponychium (Fig. 1). Clinical diagnosis of OM was suspected. An MRI was carried out, followed by an excisional biopsy of the nail bed lesion for pathological confirmation.

Multiplanar, multisequence MRI exam with gadolinium administration showed a 7.7 × 6.9 × 4.1-mm lesion in the matrix on the ulnar side, proximal to the nailfold. The nail was thickened with fingerlike projections but without erosive bone changes (Fig. 2). Enhancement of the tumor was seen on T2-weighted sequence after gadolinium injection. A radiological diagnosis of OM was made, and the patient was referred for surgical treatment.

The surgery was performed under digital block with a tourniquet applied at the base of the right thumb. The eponychial fold was retracted proximally from the nail plate after performing a full thickness skin incision of its medial border. The nail plate was totally excised, exposing the fibrous projections beneath the proximal nail fold and the filamentous tumor that was emerging at the medial border of the nail matrix. The tumor was resected

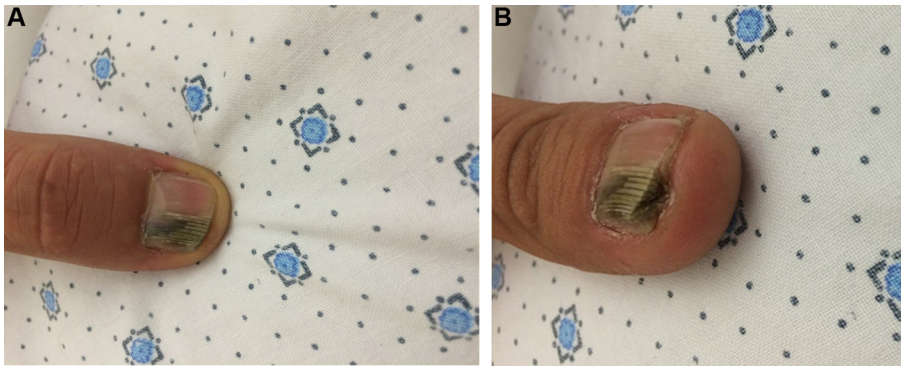


**Fig. 1.** Preoperative top (A) and distal (B) views. Note the thickened yellow longitudinal bands with transverse curvature and melanonychia.

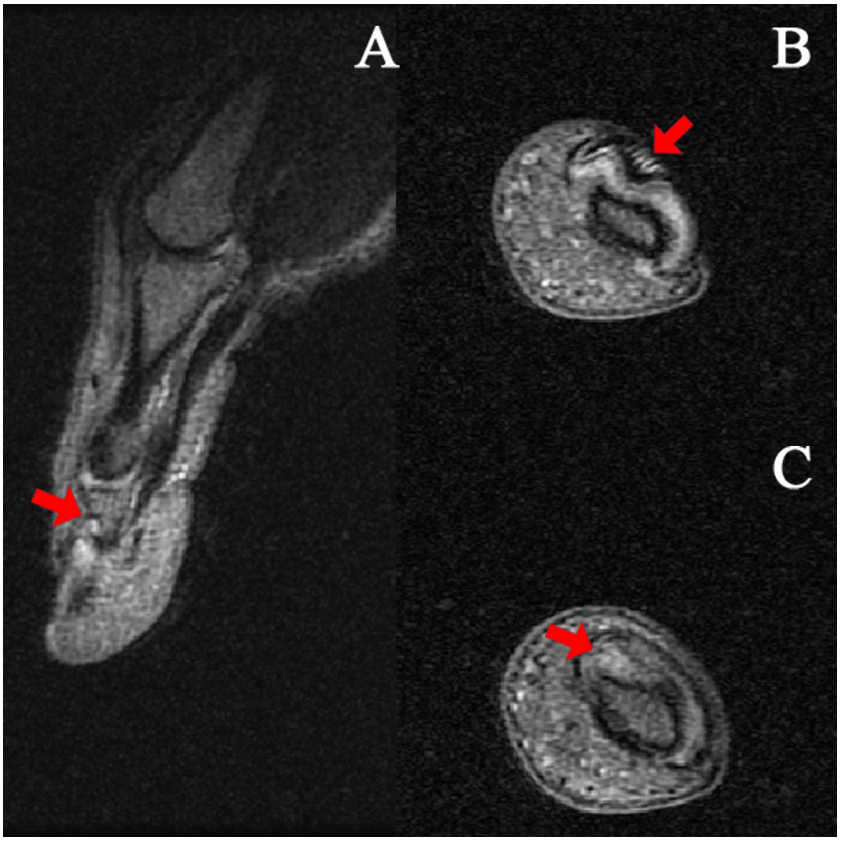


**Fig. 2.** Fat-saturated proton density-weighted MRI of the thumb showing the 7.7 × 6.9 × 4.1-mm lesion. Longitudinal (A) and transverse (B and C) slices of the tumor arising from the ulnar side of the thumb's nail. It has a heterogeneous hypersignal on proton density fat-saturated images with fingerlike projections within its thickness.

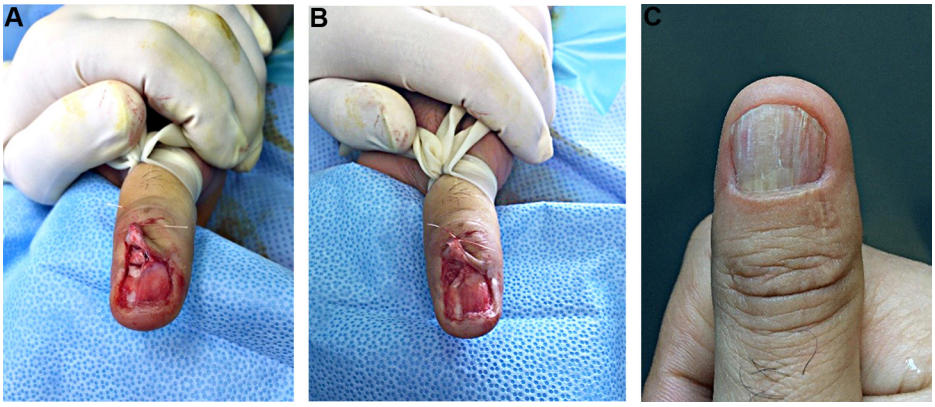


**Fig. 3.** Photographs showing the nail bed before (A) and after (B) removal of the tumor. Postoperative photo 15 months later showing the normal nail plate growth following complete tumor excision (C).

completely, while preserving 1 mm of normal matrix all around, resulting in a matrix defect of 3 × 3 mm (Fig. 3A and B). Two matrix flaps were designed. The medial flap was distally based while the lateral flap was proximally based. These flaps were elevated on the subperiosteal layer, and then glided centrally to reconstruct the defect and to cover the underlying bone structures. Flaps were fixed with Monocryl 6/0 absorbable sutures. The eponychial flap was replaced and the skin was approximated with simple interrupted VICRYL RAPIDE™ 6/0 sutures.

Histological examination of the surgical specimen found a polypoid fibroepithelial lesion originating from the ungual matrix, likely corresponding to the distal zone of the lesion. The tumor had fingerlike projections, covered by a regular, stratified, squamous epithelium lacking stratum granulosum with epithelial-lined invaginations. The superficial stroma was dense and poorly cellular. Sections also revealed the proximal root of the nail had early epithelial invaginations. The avulsed nail plate had an undulating inferior border, epithelial invaginations and V-shaped keratogenous zone, likely corresponding to the proximal zone of the lesion. Sections of the nail plate had PAS-positive fungal spores in the crevices of the nail plate and cavities filled with serous fluid.

The patient was reviewed 15 months post-surgery and had no signs of tumor recurrence with normal growth of the nail plate (Fig. 3C).

## 3. Discussion

OM is a rare benign tumor of the nail matrix. It was first described in 1992 by Baran and Kint [1]. To date, only about 100 cases have been reported. OM is considered as the only tumor to actively produce a nail plate substance, thus causing ungual changes. It is more common in light-skinned individuals, with predominance in females and peaking in the 5th decade of life [2]. The tumor is unusual in children with only one case reported in the pediatric population in a 4-year-old female, which was thought to be a reactive hyperplastic process caused by onychomycosis rather than a primary benign tumor [3,4]. In the current case, our patient reported having a normal fingernail prior to the crush injury, making it a true case of OM in a children's age group. The painless slow growth of the tumor explains the characteristic delay in seeking medical care that can last up to an average of 8 years [5]. Since our patient was a dark-skinned individual, this is the first case report to be documented in the Middle East region.

The predisposing factors are onychomycosis and trauma. The latter has only been identified in three cases [3]. Clinically, the tumor is characterized by four cardinal features: thick yellow longitudinal discoloration, splinter hemorrhage, excessive transverse curvature, and fingerlike fibrokeratogenous villous projections upon nail avulsion.

Pigmented OM is a variant that is extremely rare with only four cases previously described in the literature [6]. In our case, the pigmentation was concentrated underneath the distal thickened longitudinal bands that fade as they migrate proximally. It seems that the dark pigmentation usually follows the trajectory of the nail plate thickening and condenses in close proximity to the transverse curvatures.

Complete surgical excision of the tumor is the treatment of choice. The proximal part of the resection should always include the distal matrix to avoid recurrence [7]. The benign, slow growing course of OM makes the tumor a bone-sparing entity. As in our case, the tumor could be easily delineated from the bony elements underneath it. Defects larger than 4 mm should normally be closed by two flaps whereas defects less than 4 mm should be carefully sutured to avoid onycholysis [7]. Although the defect in our case was 3 × 3 mm, healing by secondary intention or regular suturing would have resulted in noticeable decrease in the nail plate width; thus for cosmetic purposes, we chose to use two matrix flaps to close the defect. The regrowing nail had a growth delay and mild nail dystrophy compared to the normal part; however, the nail had fully regrown with minimal dystrophy after 15 months.

The definitive diagnosis of OM is made by histological examination. The main differential diagnoses are fibrokeratoma and ungual fibroma. In the longitudinal sections of the OM, the lesion resembles a fibrokeratoma; but the presence of multiple fibroepithelial fingerlike projections and the presence of cavities filled with serous fluid in the distal portion of the nail plate contributed to ruling out fibrokeratoma. The stroma of OM located in the lunula may also suggest the diagnosis of ungual fibroma; however the presence of the glove-like filiform projections as well as the absence of longitudinal depression in the nail plate, due to the compression in the nail matrix, contributed to ruling out ungual fibroma.

OM has been suggested to be a predisposing factor in the development of secondary fungal infection. To date, few cases of OM have been associated with onychomycosis and most of them affect toenails, where fungal infections are very frequent [4,8–10]. In our case, the PAS stain revealed incidental fungal spores in the nail plate, hence strengthening the idea that onychomycosis can be a consequence of the unorganized spacial orientation of the growing nail bed as a result of tumor invagination, which creates tunnels favoring fungal growth.

## 4. Conclusion

OM is a rare, benign, fibroepithelial neoplasm of the nail matrix that has specific clinical characteristics. Although this lesion peaks

in the 5th decade of life, we have reported an unusual presentation of pigmented onychomatricoma that arose in a Middle-Eastern dark-skinned child with a history of severe nail trauma. Surgical resection of the tumor is the treatment of choice. Reconstructive flaps are recommended even with small sized tumors, especially in the fingernails as the hands are more visible and important in interpersonal relationships. Tumor invagination in the nail plate creates a favorable environment for fungal growth. Further studies should be conducted to elucidate the significance of trauma as a predisposing factor for this rare type of tumor.

### Disclosure of interest

The authors declare that they have no competing interest.

### Funding

The authors received no specific funding for this work.

## Acknowledgement

We would like to thank Dr. Laurie Abi Habib (Associate Professor, Faculty of Health Sciences, University of Balamand) for providing valuable comments.

## References

[1] Baran R, Kint A. Onychomatrixoma. Filamentous tufted tumour in the matrix of a funnel-shaped nail: a new entity (report of three cases). Br J Dermatol 1992;126:510–5.
[2] Prevezas C, Triantafyllopoulou I, Belyayeva H, Sgouros D, Konstantoudakis S, et al. Giant onychomatricoma of the great toenail: case report and review focusing on less common variants. Skin Appendage Disord 2016;1:202–8.
[3] Cañueto J, Santos-Briz Á, García JL, Robledo C, Unamuno P. Onychomatricoma: genome-wide analyses of a rare nail matrix tumor. J Am Acad Dermatol 2011;64:573–8.
[4] Piraccini BM, Antonucci A, Rech G, Starace M, Misciali C, Tosti A. Onychomatricoma: first description in a child. Pediatr Dermatol 2007;24:46–8.
[5] Di Chiacchio N, Tassara Tavares G, Padoveze EH, Bet DL, Di Chiacchio NG. Onicomatricoma. Surg Cosmetic Dermatol 2013;5:10–4 [quiz 15].
[6] Wynes J, Wanat KA, Huen A, Mlodzienski AJ, Rubin AI. Pigmented onychomatricoma: a rare pigmented nail unit tumor presenting as longitudinal melanonychia that has potential for misdiagnosis as melanoma. J Foot Ankle Surg 2015;54:723–5.
[7] Tambe SA, Ansari SMM, Nayak CS, Chokkar R, Patil PD. Surgical management of onychopapilloma, onychomatricoma, and subungual osteochondroma: case series. J Cutan Aesthet Surg 2018;11:143–7.
[8] Wang L, Gao T, Wang G. Nail bed onychomatricoma. J Cutan Pathol 2014;41:783–6.
[9] Reserva JL, Ruben BS, Venna SS. Asymptomatic longitudinal pachyxanthonychia of the fingernail. Skin Appendage Disord 2017;3:32.
[10] Kallis P, Tosti A. Onychomycosis and onychomatricoma. Skin Appendage Disord 2016;1:209–12.