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CASE REPORT

Hutchinson's sign in congenital nail matrix nevus

Sinal de Hutchinson no nevo congénito da matriz ungueal

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Abstract

Congenital nail matrix nevi (NMN) are a rare cause of melanonychia that may present with irregularity, asymmetry, and multicomponent pigmentation posing diagnostic challenges with subungual melanoma. We report a case of a 49-year-old female with longitudinal melanonychia and a 1-month recent pigmentation in the proximal nail fold. This sign is traditionally associated with malignancy, which further complicates the differentiation from acral melanoma. Therefore, a nail matrix biopsy was performed. Histopathologic examination revealed a nail matrix nevus. This procedure is crucial in suspicious cases, despite the risk of nail dystrophia. The evolution of these nevi into adulthood and their potential malignancy remains unclear, emphasizing the need for continued research and surveillance. This case highlights that congenital NMN often present with clinical and dermoscopic features of concern, mirroring those observed in subungual melanoma.

Keywords: Congenital melanocytic nevus. Congenital nail matrix nevus. Dermoscopy. Hutchinson's sign. Longitudinal melanonychia. Subungual melanoma.

Resumo

Os nevos congénitos da matriz ungueal são uma causa rara de melanoníquia que pode apresentar-se com irregularidade, assimetria e pigmentação multicomponente, o que dificulta o diagnóstico diferencial com melanoma subungueal. Descrevemos o caso de uma mulher de 49 anos com melanoníquia longitudinal que progrediu com pigmentação na prega ungueal proximal. O sinal de Hutchinson está frequentemente associado a malignidade, o que dificulta ainda mais a diferenciação com o melanoma acral. Realizou-se uma biópsia da matriz ungueal, cujo exame histopatológico foi compatível com nevo da matriz ungueal. Esta abordagem é crucial em casos suspeitos, apesar do risco de distrofia ungueal. A evolução destes nevos até à idade adulta e o seu potencial de malignidade permanecem pouco claros, enfatizando a necessidade de vigilância contínua. Este caso realça que os nevos congénitos da matriz ungueal se apresentam frequentemente com características clínicas e dermatoscópicas suspeitas, que podem mimetizar as observadas no melanoma subungueal.

Palavras-chave: Dermatoscopia. Melanoma subungueal. Melanoníquia longitudinal. Nevo melanocítico congénito. Nevo congénito da matriz da unha. Sinal de Hutchinson.

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Introduction

Nail matrix nevi (NMN) are often considered in the differential diagnosis of acral melanoma, as they both present with melanonychia as a common feature[1]. In fact, congenital NMN frequently combine semiologic features of irregularity and asymmetry with a multicomponent pigmentation of the nail plate and surrounding tissue that could suggest malignancy[1]. Despite the invasive nature and emotional distress associated with nail matrix biopsy, it is crucial in cases with a high level of suspicion[1,2]. In this context, we report a case of congenital nail matrix nevus with recent suspicious changes, exploring its clinical and dermoscopic features.

Case report

A 49-year-old female patient was referred to our dermatology department due to a 1-month evolution of pigmentation in the proximal nail fold, associated with a longstanding longitudinal melanonychia on the second toe's nail.

The patient reported having melanonychia since the 1st year of life and that it had not changed until a month before.

Physical examination showed light brown irregular longitudinal melanonychia covering over two-thirds of the nail without dystrophy, along with irregular monochromatic dark brown pigmentation in the proximal nail fold (Fig. 1). Dermoscopy revealed irregular light and dark brown longitudinal microlines in the dorsal nail plate and dark brown diffuse irregular pigmentation in the proximal nail fold (Figs. 2A and B).

Recent changes prompted a nail matrix biopsy. The nail matrix biopsy was performed under local anesthesia with a proximal digital block. Subsequently, a tourniquet was applied, the proximal nail fold was retracted, and the nail plate was avulsed (Fig. 3). A tangential incisional biopsy was performed, and finally, the retracted fold and the avulsed nail plate were sutured.

Histopathological examination revealed hyperpigmentation of the basal layer of the epidermis and coexisting melanophages in the dermis, without melanocytic junctional proliferation or dysplasia of the nail matrix, consistent with a nail matrix nevus (Fig. 4).

At the 3-month follow-up, the patient did not exhibit new changes in the nail or periungual tissue, and no dystrophy was observed.

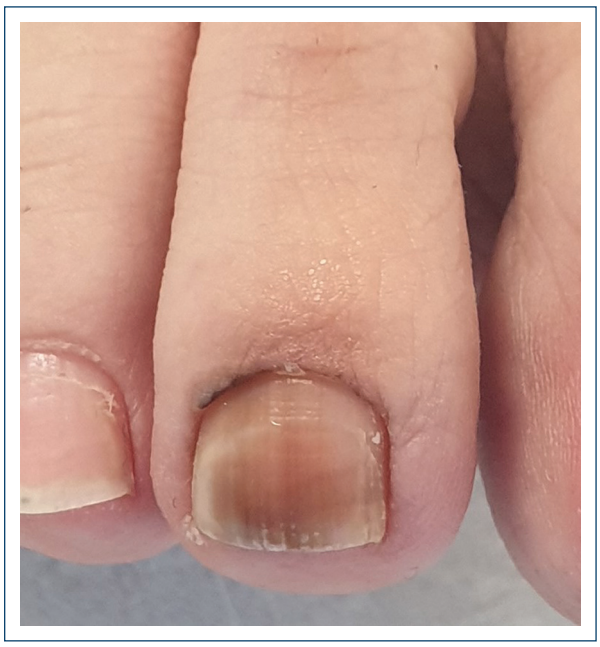

Figure 1. Clinical findings of the nail plate – light brown irregular longitudinal melanonychia covering over two-thirds of the nail without dystrophy, along with irregular dark brown pigmentation in the proximal nail fold.

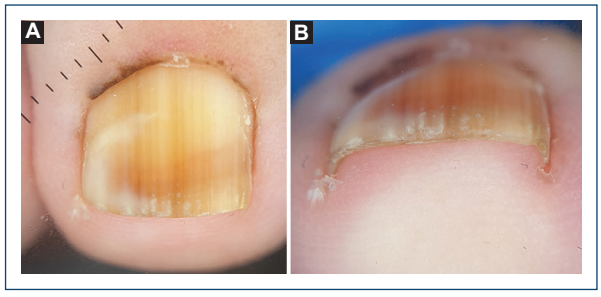

Figure 2. Dermoscopy of the nail. **A:** dermoscopy view of the nail plate: irregular light and dark brown longitudinal microlines with blurred lateral borders, covering over two-thirds of the nail plate, and dark brown diffuse irregular pigmentation in the proximal nail fold.
B: dermoscopy view of the distal edge of the nail plate: irregular light and dark brown longitudinal microlines in the dorsal nail plate without any pigmentation in the hyponychium.

Discussion

Congenital NMN are rare, with limited documented cases described in the literature[1]. They frequently exhibit clinical and dermoscopic distinctive features such as irregularity, asymmetry, and multicomponent

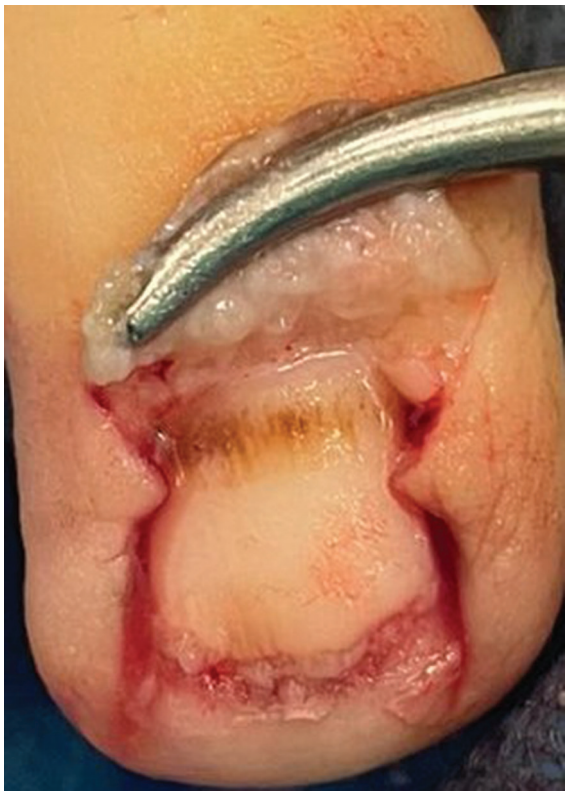

Figure 3. Intraoperative view of the nail matrix biopsy showing the light and dark brown fibrillar pattern of the nail matrix.

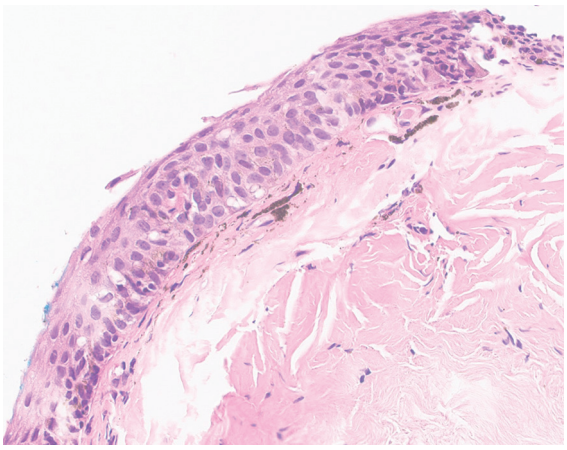

Figure 4. Histopathology of the nail matrix – hyperpigmentation of the basal layer of the epidermis and coexisting melanophages in the dermis, without melanocytic junctional proliferation or dysplasia (hematoxylin and eosin stain).

pigmentation affecting the nail plate with or without involvement of the periungual tissues, raising concerns about potential malignancy[1-3]. The main clinical and dermoscopy feature of congenital NMN is the irregular pattern, defined by longitudinal microlines, irregularity in width, space, color, triangular shape, polychromasia, and irregular periungual pigmentation[1]. Furthermore, the most prevalent dermoscopic pattern of the periungual pigmentation has been described as a distal fibrillar ("brush-like") pattern[1].

The notable characteristic of melanonychia extending into the periungual tissues, known as Hutchinson's sign, is traditionally associated with acral melanoma[2,3]. In addition to acral melanoma and congenital NMN, other differentials associated with Hutchinson's sign include ethnic variations, trauma, systemic diseases, and drug adverse effects[2,3].

Due to the rarity of congenital matrix nevi, there is a lack of consensus regarding their management. However, in the majority of congenital NMN cases, conservative management suffices, with clinical follow-up, including clinical photography and dermoscopy[2]. Performing a nail matrix biopsy is mandatory in suspicious cases, although it may have a risk of permanent scarring[1,4,5].

Histopathologic analyses of nail matrix biopsies in congenital NMN are infrequent in the literature. Nevertheless, some cases have been described as junctional NMN or as functional melanocytic activation[1]. In our case, the biopsy showed features of melanocytic activation. In addition to congenital NMN, other causes of melanocytic activation include pregnancy, chronic trauma, nail-biting, drug adverse effects, and systemic diseases[4], none of which were present in our case.

Aggressive procedures, such as complete excision involving the entire length of the nail matrix, are reserved for situations where melanoma cannot be ruled out even after an expert review of clinical and histopathologic findings[2]. However, subungual melanoma cases arising in congenital NMN are exceptionally rare, casting doubt on the accuracy of diagnoses[1,6]. Furthermore, there is currently no data in the literature regarding the evolution of congenital NMN into adulthood and their risk of malignant transformation.

Conclusion

This case highlights that congenital NMN frequently manifest clinically and dermoscopically alarming characteristics, mirroring those observed in subungual

melanoma. This resemblance is particularly evident in the presence of the Hutchinson's sign. Notably, the diagnostic criteria established for adult subungual melanoma may not be directly applicable or reliable when evaluating congenital NMN.

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript nor for the creation of images, graphics, tables, or their corresponding captions.

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