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A pediatric case of unusual melanocytic proliferation of the nail

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Abstract

Pigmentation of the nail plate, or melanonychia, is typically a benign condition caused by melanocyte activation. Although rare, melanonychia may be the initial presentation of melanoma, thus all cases require an in-depth examination. Evaluation in pediatric patients can prove especially difficult as benign cases have a higher prevalence of atypia compared to adults. Lack of specific treatment guidelines in the pediatric population can make diagnosis and treatment challenging. We report a pediatric patient with melanonychia with atypical features that required significant evaluations and collaboration to ultimately reach a treatment plan.

Keywords: melanonychia, pediatric, subungual, melanoma, nail, pigmentation

Introduction

Melanonychia is defined as brown or black pigmentation involving the nail plate owing to the presence of melanin [1]. The pigmentation may relate to several causes including activation of melanocytes in the nail matrix or melanocytic hyperplasia [2]. The etiology of melanonychia is variable; it ranges from a benign nevus to subungual malignant melanoma [1]. Although the diagnosis of melanoma of the nail is rare, it is important to exclude this possibility. We report a pediatric case of melanonychia exhibiting atypical features that was determined to be benign despite concerning features on histopathology. The difficult decision was made to excise the affected nail plate and nail bed.

Case Synopsis

A 9-year-old African American boy presented with diffuse blue-black pigmentation under the right fifth digit fingernail. At the age of 18 months, the patient experienced trauma to the fingernail. Initially, the lesion was a single, dark-brown pigmented streak under the nail plate. The patient was followed every two years with close observation following the incident. The pigmentation continued to expand and ultimately covered the patient's entire nail by the age of five. During the next four years, the diffuse pigmentation remained stable. The patient had no other significant medical history or associated symptoms.

On physical examination, the patient was a well-appearing young boy with diffuse, uniform blue-black discoloration on the right fifth fingernail and dark brown pigmentation of the hyponychium. He exhibited a positive Hutchinson sign with discoloration extending onto the proximal nail fold (Figure 1A). The remaining nails were clear.

On dermoscopy, there was a lentiginous, but mostly nested proliferation of melanocytes involving the nail plate, nail matrix, and nail folds. A 4-mm punch biopsy of the proximal nail fold and matrix was performed under local anesthesia. Histopathologic examination exhibited residual atypical intraepidermal melanocytic proliferation involving the nail matrix. A diagnosis of congenital nevus was made. Although the risk of future malignant transformation is rare, this possibility as well as the cosmetic state of the nail greatly concerned the family. After much review and collaborative discussions on the topic, surgical excision of the entire nail plate, nail matrix, and nail folds was performed.

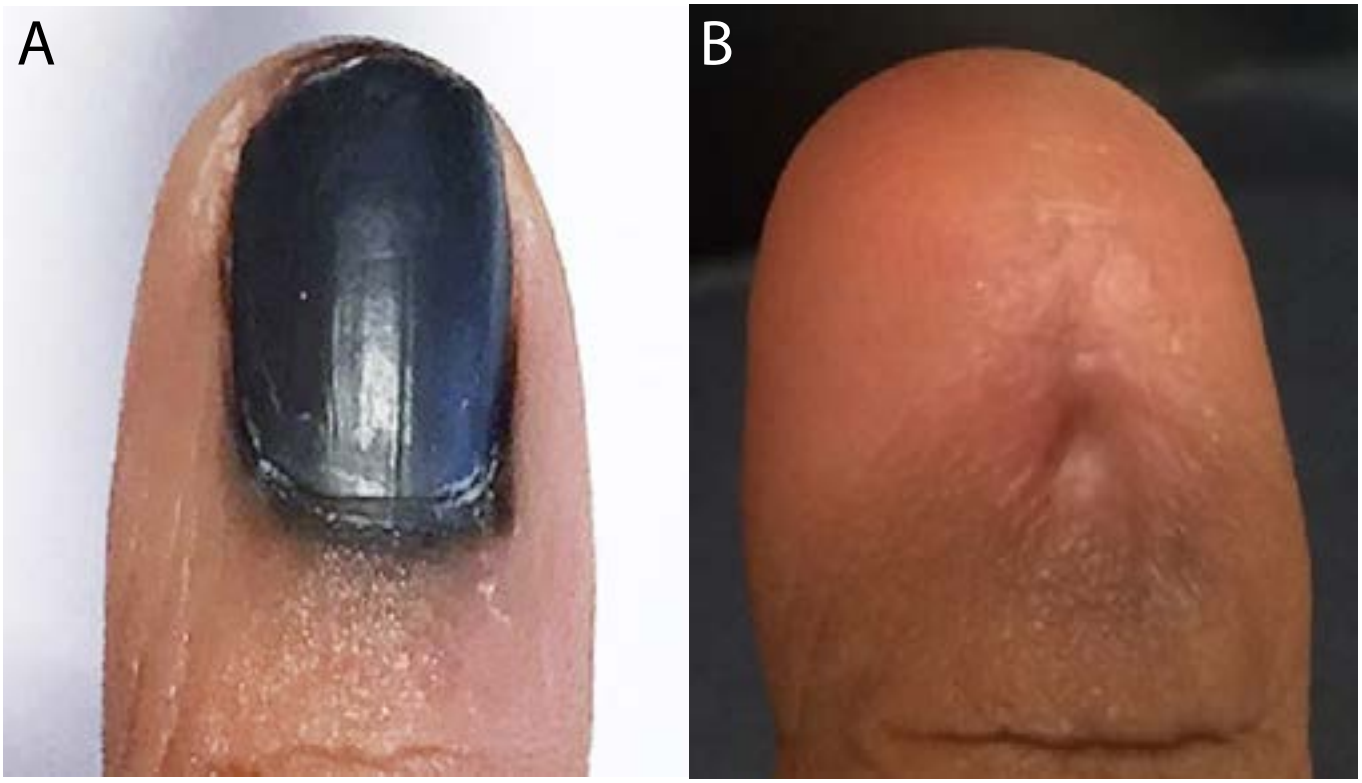

Figure 1. *Right fifth digit fingernail. A) Prior to excisional biopsy, the fingernail displays diffuse, uniform blue-black discoloration extending onto the proximal nailfold and hyponychium. B) After excisional biopsy, the fingertip healed with minimal hypertrophic and hyperpigmented scarring.*

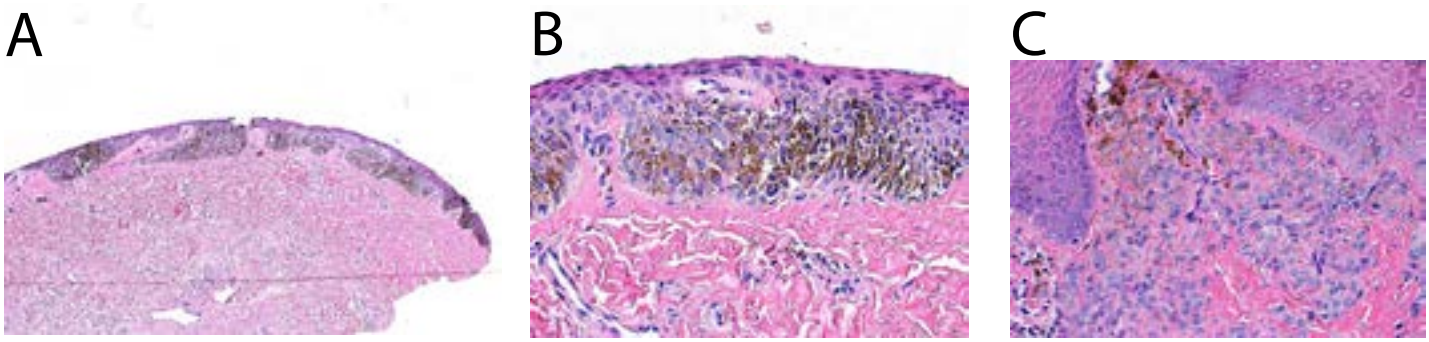

Figure 2. *Excisional histopathology. A) Low power view displays a broad proliferation of melanocytes throughout the nail matrix epithelium arranged as confluent solitary units and small nests (H&E, 40x). B) Higher magnification exhibits confluent solitary melanocytes within the lower matrical epithelium and focally extending to higher levels (H&E, 400x). C) Collections of banal nevoid melanocytes within the nail bed stroma (H&E, 400x).*

Histopathology following surgical excision exhibited melanocytes that were small to intermediate in size and epithelioid-to-dendritic in shape, clustered mainly at the rete ridges. Melanocytes were arranged as confluent solitary units and small nests within the lower matrical epithelium and focally extended to higher levels (**Figure 2A, 2B**). Collections of banal nevoid melanocytes were noted within the nail bed stroma (**Figure 2C**). A small focus of residual neoplasm was identified within the deep surgical site. FISH testing was negative for melanoma.

The patient's fingertip healed very well with only minimal hypertrophic and hyperpigmented scarring (**Figure 1B**). The residual neoplasm seen on pathology was not pursued surgically. The patient is monitored closely with examination every six months. There is a low threshold for re-excision with any pigment transformation at the surgical site over concern for malignancy.

Case Discussion

Melanonychia typically presents as longitudinal melanonychia or melanonychia striata. This is

identified as a longitudinal streak of pigmentation on the nail plate. It is more commonly seen in individuals of African or Asian descent. When determining the etiology of melanonychia, the ABCDEF rule has been useful for differentiating between benign and malignant lesions. This criteria includes negative prognostic factors such as age between 50-60 years old, African or Asian ethnicity, presence of a band over 3mm, brown or black pigmentation, any change in pigmentation, digit most commonly involved (thumb/hallux), extension of pigment onto proximal or lateral nail fold, and a positive family history [3]. In adults, the use of dermoscopy has proved useful for diagnosis. Concerning features include a positive Hutchinson sign or a melanocytic proliferation with an irregular pattern. If the lesion is suspicious for melanoma, then a biopsy is indicated. If the lesion is not suspicious for malignancy, then close observation is preferred [4-6]. This criterion is not valid in children because benign lesions in children exhibit features diagnostic of malignant lesions in adults [1].

In a cohort study of 40 pediatric patients with longitudinal or total melanonychia, 19 patients were diagnosed with melanocytic nevus, 12 patients with lentigo, and 9 patients with functional longitudinal melanonychia. None of these patients fulfilled the criteria for diagnosis of melanoma [7]. Although pediatric subungual melanoma is rare, there have been case reports of melanoma-in-situ that started with a longitudinal melanonychia, which expanded to encompass the entire nail plate. The histopathologic examination of two of these cases found atypical proliferation of melanocytes arranged in single "pagetoid" unit and nest distribution. Upon re-examination in subsequent years following excision, the patients showed no sign of recurrence [8, 9].

In our case, the patient exhibited several criteria concerning for the diagnosis of subungual melanoma according to the ABCDEF rule used in adults [3]. However, the lesion was ultimately determined to be a benign congenital nevus with atypical features as is commonly seen in the pediatric population. Multiple dermatologists were consulted including specialists from pediatric dermatology, dermatopathology, and MOHs surgery prior to reaching a consensus on surgical excision. The family was involved in each step of the decision process and all the pathology specimens were reviewed by a second dermatopathologist. Ultimately, the parents and patient were in favor of excision for fear of malignancy in the future. As a result, the entire nail plate, nail matrix, and nail folds were excised.

Conclusion

Diagnosis and treatment of pediatric melanonychia remains difficult and controversial. Treatment plans are made on a case-by-case basis. Although the risk for melanoma is exceedingly low in the pediatric population, it is important to exclude melanoma. This case demonstrates that more research is needed in order to develop diagnostic criteria that can guide clinicians in the assessment and treatment of pediatric melanonychia.

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