Title
Subungual pleomorphic fibroma: a case report and review of the literature

Journal
Dermatology Online Journal, 22(11)

Authors
Halteh, Pierre
Magro, Cynthia M
Lipner, Shari R

Publication Date
2016

DOI
10.5070/D32211033143

Volume 22 Number 11
November 2016

Case report

Subungual pleomorphic fibroma: a case report and review of the literature

Pierre Halteh[1], Cynthia M Magro[2], Shari R. Lipner[1]

Dermatology Online Journal 22 (11): 6

[1] Weill Cornell Medicine , Department of Dermatology, New York, NY

[2] Weill Cornell Medicine, Department of Dermatopathology, New York, NY

Correspondence:

Shari R. Lipner, MD, PhD
1305 York Avenue, 9th floor
New York, NY 10021
T 646.962.3376
Email: shl9032@med.cornell.edu

Abstract

We describe an interesting case of pleomorphic fibroma of the subungual region in a middle aged woman who presented with a recurrence of thickening, lateral ridging, and a midline split of the right third fingernail, 20 years after initial excision. Histology of the specimen demonstrated hyperchromatic stellate cells within the superficial corneum, which were CD34 positive, consistent with a pleomorphic fibroma. Pleomorphic fibroma is a rare mesenchymal neoplasm characterized by atypical spindled cells amidst a collagenous stroma. It is a benign, slow growing tumor that has only been reported in the subungual region twice previously. Although histology may show cellular atypia, the slow-growing benign nature of the tumor, reassuring features histologically i.e low cellularity, and lack of myxomatous change in the stroma, make a conservative approach to therapy reasonable.

Introduction

A 52-year-old woman with no significant past medical history presented for evaluation of changes in her right third fingernail. She stated that for the past 5 years her nail was constantly splitting and getting stuck on clothing; one part of the nail was thickeing and becoming more yellow. She was originally seen for a similar problem 23 years earlier. The patient did not present with any other skin, hair, or nail complaints. She did not have a personal or family history of nail abnormalities or skin cancer.

During the initial encounter to another dermatologist 23 years earlier, the patient stated that her right third fingernail was splitting for at least 11 years prior to her visit. She recalls injuring this fingers while opening the lid of a jar as a child, with no obvious sequelae. On examination, the right third fingernail was described as hypertrophic and yellow, with distal onycholysis and onychoschezia. She was treated topically with 3% thymol in acetone for 4 months for a presumed permanent nail deformity with secondary bacterial and fungal infection, with little improvement. Therefore, nail biopsy was subsequently performed.

The previous biopsy report was significant for a proliferation of spindled cells superficially. Although a specific diagnosis could not be established, the differential diagnosis included fibromatosis, fibrokeratoma, and subungual fibroma; a complete excision was recommended. The tumor was excised with no recurrence until 20 years later when the patient again noticed nail splitting, thickening, and yellow discoloration. An MRI performed at the time showed elliptical thickening of the radial aspect of the nail on the third digit, with mild thickening of the ulnar aspect of the nail bed, but no soft tissue mass in the subungual region. The contour change was thought to be related to the prior surgery.

She was referred to our nail clinic for evaluation. On physical examination, the right third fingernail had thickening, lateral ridging, and a midline split (Figure 1). The surrounding nail folds were normal, as were all the other nails. A full skin and hair examination revealed no abnormalities. Owing to her previous history, pathology, and symptoms a decision was made to explore the nail and re-excise the area. The nail was partially avulsed with proximal nail fold incision, the tumor was excised *en bloc*, and the resultant defect repaired with reconstruction of the lateral nail fold. The specimen was routinely processed and sent for histologic examination.

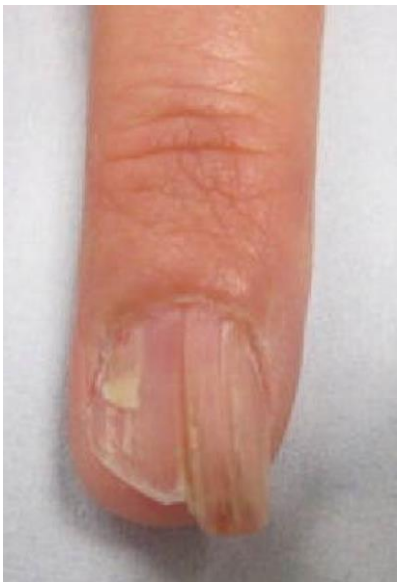

Figure 1. Right third fingernail with thickening, lateral ridging and a midline split.

The biopsy showed a moderately cellular fibrous lesion. The background stroma had a collagenized appearance and was without any discernible myxomatous alteration. The dominant cell populace was a relatively bland proliferation of spindled fibroblastic appearing cells. Also present were stellate appearing cells that exhibited marked nuclear hyperchromasia, most apparent superficially. The cells were highlighted by CD34 without any staining for S-100, EMA or Melan-A.

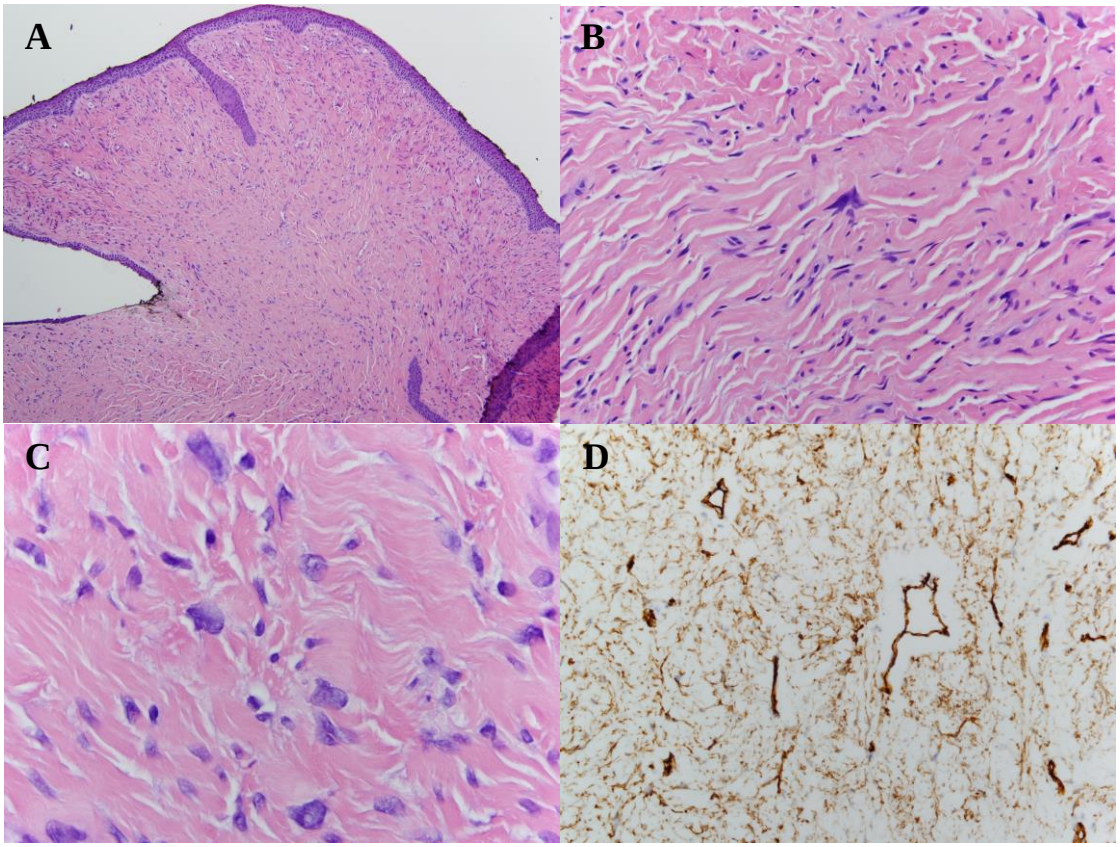

Figure 2. Histopathology. A. Low power view shows a subepithelial spindle cell lesion of moderate cellularity; the background stroma exhibits a collagenized appearance (100x, Hematoxylin and Eosin). B. The dominant cell population is a relatively bland proliferation of spindled fibroblastic appearing cells (200x, Hematoxylin and Eosin). C. Higher power view shows atypical enlarged hyperchromatic stellate appearing cells that assume a random single cell disposition amidst the monomorphic appearing spindled cells (400x, Hematoxylin and Eosin). D. There is extensive immunoreactivity of the lesion for CD34 (200x, diaminobenzidene); the S-100, EMA or Melan-A immnostains were negative (not illustrated).

Discussion

We have presented a case of a slow-growing subungual tumor that recurred 20 years following its initial excision. The recent biopsy material demonstrated a CD34 positive subungual fibrocytic lesion with features of a subungual fibroma. Owing to the hyperchromatic stellate cells within the superficial corium, the designation of pleomorphic fibroma seems reasonable.

Pleomorphic fibroma is a very rare mesenchymal neoplasm characterized by atypical spindled cells amidst a collagenous stroma. It was first reported by Kamino and co-workers in 1989. The authors presented 8 cases of polypoid and dome-shaped relatively small (i.e. 4 millimeters up to 1.6 centimeters) cutaneous fibrous lesions with sparse cellularity but striking nuclear atypia and rare mitotic figures. The mean age was 52 years of age with a slight female predominance. The lesions exhibited benign behavior with a local recurrence in only one of the 8 cases. They proposed that the atypia to that encountered in other benign tumors including pleomorphic lipoma, pleomorphic leiomyoma, ancient schwannoma, and variants of dermatofibroma with atypical cells [1].

This neoplasm typically presents in non-acral sites as a polypoid pedunculated mass, but may occur on non-acral skin [2]. The cells invariably express vimentin and are frequently positive for CD34. The combination of relatively low cellularity and lack of mitotic activity define additional critical features that enable its distinction from malignant cutaneous mesenchymal counterparts, most notably atypical fibroxanthoma, pleomorphic dermal sarcoma, and low grade fibromyxoid sarcoma. The latter is an important consideration because of its potential localization to the nail apparatus and its modest cellularity. That being said the architectural growth pattern is very distinctive, comprising whorls of spindled cells juxtaposed to spindled and myxomatous areas with arborizing blood vessels [3]. The spindled cells are only mildly atypical and the random bizarre atypia seen in pleomorphic fibroma is not a feature. Benign cutaneous mesenchymal lesions exhibiting atypical spindled cells include dermatofibroma with monster cells and sclerotic fibroma [4], the latter likely defining part of a morphologic continuum with pleomorphic fibroma [5, 6]. The classic subungual Koenen fibromas of tuberous sclerosis present with longitudinal ridges that can exhibit a reddish discoloration or a loss of color (i.e. longitudinal leukonychia [7]). Significant pleomorphism has not been described in these lesions. In this location a superficial acral fibromyxoma is an important consideration. However, this lesion was lacking in the myxomatous areas that would be typical for a superficial acral fibromyxoma. In addition, the inherent atypia manifested by this lesion would not be compatible with this diagnosis.

There are two other prior reported cases of subungual fibroma for a total of 3 cases [8, 9], Table 1. They were both slow-growing tumors. In one case the patient was a 66-year-old woman who reported a growing mass on the left middle finger for over 40 years, with distortion of the nail plate. The second case was a 54-year-old man with a one-year history of a painless slowly progressively enlarging mass on the right thumb; there was associated paronychia and subungual hyperkeratosis.

Table 1. Overview of three published cases of Pleomorphic Subungual Fibroma.

	Case 1	Case 2	Case 3
Authors of Case Report	Hassanein et. al	Hsieh et. al	Halteh et. al
Age / Sex	54 year old/ male	66 year old/ female	52 year old/ female
Affected Digit	Right 1st fingernail	Left third fingernail	Right third Fingernail
Clinical Presentation	1 year history; asymptomatic; paronychia and nail hyperkeratosis; no trauma	40 year history; asymptomatic; nail dystrophy; no trauma	28 year history; asymptomatic; splitting; preceding trauma
Treatment Method	Excisional biopsy	Nail avulsion, excision	Initial Occurence: Nail avulsion, excision. Recurrence: Partial nail avulsion, excision
Immunoreactivity of Cells	Vimentin +, CD34+, S100-	Vimentin +, CD34 -, Smooth muscle actin - , S100 -, factor XIIIa -	CD34 +, S100 -, EMA -, Melan-A -
Recurrence	No recurrence 18 months after excision	No recurrence 36 months after excision	Recurrence after 23 years after initial excision, No recurrence after 6 months after second excision

With a total of 3 cases reported to date in the literature, subungual pleomorphic fibroma should emerge as a distinct clinical pathological entity. Based on the three cases reported, the patients presented at a mean age of 57.3 years (range 42-66), but according to history they noted changes in their nails at a mean age of 34.3 (range 24-53). Two of the three cases involved the middle finger and all cases to date are associated with dystrophic alterations of the nail plate. There does not appear to be a unifying inciting event such as trauma, although trauma has been postulated in other fibromas, most notably the subungual fibroma of tuberous sclerosis [7]. Although the atypia may be alarming, the slow-growing nature of the tumor clinically, the low

cellularity, the lack of myxomatous change in the stroma, and the absence of mitotic activity should allow for the correct diagnosis to be made in all cases. Given the benign nature of the tumor, a conservative approach to therapy is reasonable in all cases.

Conclusions

Subungual pleomorphic fibroma is an uncommon mesenchymal neoplasm, characterized by spindled fibroblastic cells, with CD34 positive stellate appearing cells exhibiting marked nuclear hyperchromasia. They are benign slow-growing tumors and can be managed conservatively.

References

1. Kamino, H., J.Y. Lee, and A. Berke, Pleomorphic fibroma of the skin: a benign neoplasm with cytologic atypia. A clinicopathologic study of eight cases. Am J Surg Pathol, 1989. **13**(2): p. 107-13.[PMID: 2916726]
2. Cohen, P.R., et al., Pleomorphic fibroma of the skin. Skinmed, 2010. **8**(2): p. 113-5. [PMID: 2916726]
3. Cabibi, D., et al., Rare localization of low-grade fibromyxoid sarcoma to the nail region. Br J Dermatol, 2005. **153**(3): p. 686-8. [PMID: 16120176]
4. Tamada, S. and A.B. Ackerman, Dermatofibroma with monster cells. Am J Dermatopathol, 1987. **9**(5): p. 380-7. [PMID: 2825558]
5. Bhambri, A. and J.Q. Del Rosso, Solitary sclerotic fibroma. J Clin Aesthet Dermatol, 2009. **2**(6): p. 36-8. [PMID: 20729948]
6. Pernet, C., et al., Solitary sclerotic fibroma of the skin: a possible clue for Cowden syndrome. Eur J Dermatol, 2012. **22**(2): p. 278-9. [PMID: 22377798]
7. Aldrich, C.S., et al., Acral lesions in tuberous sclerosis complex: insights into pathogenesis. J Am Acad Dermatol, 2010. **63**(2): p. 244-51.[PMID: 20462663]
8. Hsieh, Y.J., et al., Subungual pleomorphic fibroma. J Cutan Pathol, 2003. **30**(9): p. 569-71. [PMID: 14507406]
9. Hassanein, A., et al., Subungual myxoid pleomorphic fibroma. Am J Dermatopathol, 1998. **20**(5): p. 502-5. [PMID: 9790115]