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Title
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Permalink
https://escholarship.org/uc/item/07j2872w

Journal
American Journal of Dermatopathology, 40(2)

ISSN
0193-1091

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Publication Date
2018-02-01

DOI
10.1097/dad.0000000000001027

Peer reviewed

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Am J Dermatopathol. Author manuscript; available in PMC 2019 February 01.

Published in final edited form as:

Am J Dermatopathol. 2018 February ; 40(2): 139–141. doi:10.1097/DAD.0000000000001027.

Intradermal Proliferative Fasciitis Occurring With Chondrodermatitis Nodularis Helicis

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Abstract

Nodular fasciitis is a benign myofibroblastic tumor. Its uncommon variant, proliferative fasciitis (PF), can present in an even less common intradermal form. We report a case of intradermal PF of the ear in a 45-year-old man who presented with recurrent episodes of pain and swelling of the lesion. Histologic examination showed a dermal, nodular proliferation of ganglion-like basophilic fibroblasts with prominent nuclei and nucleoli, admixed with foamy histiocytes and areas of spindle cells arranged in intersecting fascicles in a fibromyxoid background. Lesional cells stained positive for smooth muscle actin and were negative for AE1/AE3, p63, and Melan-A. CD68 highlighted intervening histiocytes. We postulate that the underlying chondrodermatitis nodularis helicis was a triggering etiology, consistent with the current speculation that intradermal PF results from trauma.

Keywords

intradermal proliferative fasciitis; proliferative fasciitis; nodular fasciitis; mesenchymal tumors of the skin; soft tissue tumors

INTRODUCTION

Nodular fasciitis (NF), also known as pseudosarcomatous fasciitis, is an uncommon benign subcutaneous nodular proliferation of myofibroblasts.[1] NF usually occurs in subcutis or fascia but can occur intradermally. It has the tendency to present in young adults and children, typically on the upper extremities or the head and neck.[1] NF are composed of cytologically bland, uniform spindle cells with elongated, tapering nuclei, vesicular chromatin, small nucleoli, and palely eosinophilic cytoplasm, with no significant cytologic atypia or pleomorphism. These cells arranged in short-intersecting bundles within focally microcystic myxoid stroma, containing extravasated red blood cells and scattered lymphocytes. They are typically diffusely positive for smooth muscle actin (SMA). The morphologic features of intradermal NF are similar to NF at conventional sites and should not be confused with sarcoma.[1]

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The authors declare no conflicts of interest.

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Intradermal proliferative fasciitis (PF) is a very rare variant of NF. PF is histologically distinguished from NF by the presence of large polygonal cells with amphophilic cytoplasm resembling ganglion cells, in addition to the classic findings of NF.[2–4] Although PF and NF more commonly present as subcutaneous lesions on the extremities, their intra-dermal variants tend to arise on the head and neck.[5] A post-traumatic etiology has been proposed, although debate continues on whether the origin of PF and NF is reactive or neoplastic.[2,3,6,7,11] Detection of translocation MYH9 in some cases suggests a possible neoplastic origin to NF. Herein, we report a case of intradermal PF arising in the context of chondrodermatitis nodularis helicis.

CASE REPORT

A 45-year-old man presented to otolaryngology clinic with a 2-year history of an intermittently painful nodule on the right posterior ear (Fig. 1). The nodule had undergone recurrent episodes of marked swelling and shooting pain, worsening after lying on the ear. The patient denied any drainage from the site. There was no history of surgical intervention or other procedures to the ear. Examination showed a 7-mm mobile, firm nodule at the right posterior lobule and posterior auricle, with a focal epidermal indentation without punctum (Fig. 1). An excisional biopsy was performed. This showed a dermal, nodular proliferation of ganglion-like basophilic fibroblasts with prominent nuclei and nucleoli, admixed with foamy histiocytes blending with areas of spindle cells arranged in short intersecting fascicles in a fibromyxoid background (Fig. 2). A fragment of degenerated cartilage consistent with chondrodermatitis nodularis helicis was noted at the deep biopsy edge. Lesional cells stained positive for smooth muscle actin CD68 highlighted histiocytes. Lesional cells were negative for AE1/AE3, p63, and Melan-A. Fluorescence in situ hybridization was negative for MYH9 gene rearrangement.

DISCUSSION

Intradermal PF is a benign myofibroblastic proliferation closely related to other pseudosarcomatous lesions, such as NF and proliferative myositis.[4,5] PF usually presents as a rapidly growing, firm, often tender, solitary nodule about 1–7 cm in diameter.[3] It is regarded as a reactive process that typically involves the subcutaneous tissue and fascia of the forearm and thigh of middle-aged people, average age of 50 years and affects both sexes equally.[3] The rare intradermal variant of PF has only been reported in the literature 3 times; once in the finger and twice in the head and neck.[3,4,8] The macroscopic features are nonspecific, with nodules being poorly demarcated and composed of gray-tan tissue; microscopically, PF is characterized by a proliferation of large polygonal ganglion-like cells with abundant amphophilic, cytoplasm and eccentrically located, large nucleus with prominent nucleolus, intermixed with fascicles of spindle-shaped cells.[3] Multinucleated ganglion like cells are often present.

Standard textbooks of tumor pathology, such as the Armed Forces Institute of Pathology (AFIP) and World Health Organization (WHO) state that, by definition, PF is subcutaneous. Despite having histological features characteristic of PF occurring in the subcutis and fascia, intradermal PF can cause diagnostic confusion because of its unexpected location. The

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histologic differential diagnosis of intradermal PF includes reticulohistiocytoma, cutaneous monocytic leukemia, epithelioid or hemosiderotic fibrous histiocytoma, intradermal Spitz nevus, melanoma, pleomorphic dermal sarcoma/atypical fibroxanthoma, or squamous cell carcinoma.[3] When intradermal PF is located on the digit, as has been reported in one case, differential diagnosis includes fibromatosis, fibro-osseous pseudotumor of the digits, inflammatory myxohyaline tumor of the distal extremities (myxoinflammatory fibroblastic sarcoma), perineuroma, and epithelioid sarcoma.[4]

Reticulohistiocytoma are composed of large, mono-nucleated epithelioid macrophages with abundant pink cytoplasm, mixed with lymphocytes and sometimes granulocytes. Immunohistochemistry differentiates reticulohistiocytoma from PF—with reticulohistiocytoma expressing macrophage markers CD68, CD163, and lysozyme; and variable staining of S100, CD31, CD43, CD45, and CD45RO. The clinical picture of monocytic leukemia does not typically present like PF, with multiple plaques or nodules and often accompanying systemic symptoms. Monocytic leukemia may be epithelioid with significant nuclear atypia and mitoses that can mimic anaplastic large cell lymphoma. Immunohistochemistry will typically detect markers of monocytic lineage, making the distinction from PF. Epithelioid fibrous histiocytoma is an uncommon subtype of dermatofibroma. It is often confused with a pyogenic granuloma clinically, presenting on the extremities as a polypoid, red nodule less than 1 cm in size. Epithelioid fibrous histiocytomas are usually located in the superficial dermis and show an epidermal collarette. The epithelioid cells are typically smaller than those of PF, uniform, and usually staining diffusely with Factor XIIIa. Epithelial membrane antigen and anaplastic lymphoma kinase in approximately 64% and 88% of cases, respectively.[5,8]

Moreover, we considered a diagnosis of hemosiderotic fibrous histiocytoma but based on the focal presence of sheets of fibrohistiocytes with intra- and extra-cellular hemosiderin pigment deposition. However, the presence of ganglion-like cells blending with areas of NF favors PF. Intradermal Spitz nevi and melanoma may mimic intradermal PF, but the later lacks expression of S-100 protein and HMB45, helping to rule out a melanocytic lesion. There was no pleomorphism or atypical mitoses to suggest the possibility of atypical fibroxanthoma/pleomorphic dermal sarcoma. Negative staining for AE1a/AE3 and p63 argues against squamous cell carcinoma.

Of interest, we present the first reported case of intradermal PF arising in the context of chondrodermatitis nodularis helicis. This histologic finding, in addition to resolution of pain after excisional biopsy, absence of recurrence, and negative fluorescence in situ hybridization for MYH9 rearrangement, suggests a possible traumatic etiology rather than an underlying neoplastic process in intradermal PF. This is in contrast to the cases of NF that have been previously described in the literature,[4,9,10] raising questions about the nature of PF as a potentially reactive process that is molecularly distinct from NF. Alternatively, this may represent part of the subset of cases that are known to be negative for an MYH9 rearrangement, as seen in 10% of NF cases.[11,12]

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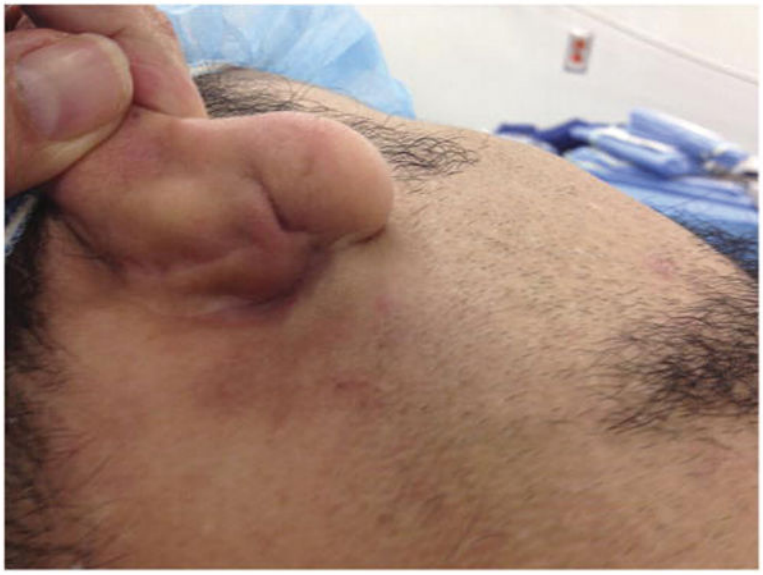

FIGURE 1.
Clinical picture of intradermal proliferative fasciitis. This was a well-defined, firm, tender 7-cm nodule on the right posterior ear.

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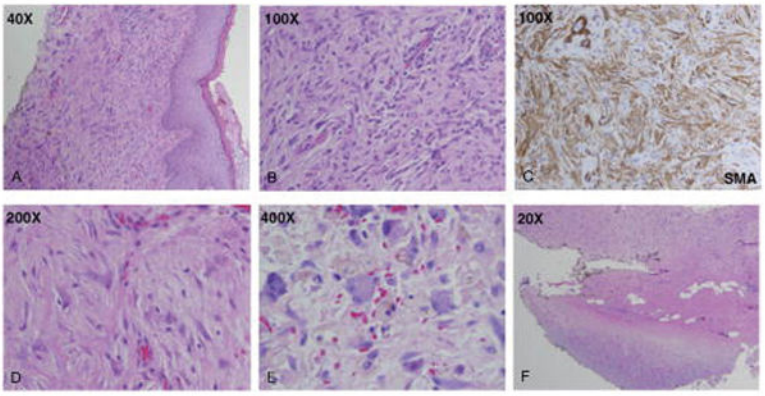

FIGURE 2.

A, B, and D, Intradermal intersecting bundles of spindled fibroblasts and ganglion-like cells disposed in a fibromyxoid background, admixed with extravasated eythrocytes, foamy histiocytes, and scattered lymphocytes (A: ×40, B: ×100, D: ×200). C, SMA immunostain highlights lesional cells (×100). E, High magnification of ganglion-like cells admixed with foamy histiocytes (×400). F, Fragment of degenerated cartilage (×20).