Atrophic Dermatofibroma: A Case Report and Review of the Literature

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BACKGROUND. **Atrophic dermatofibroma is an uncommon variant of dermatofibroma. It lacks the classic clinical features of a dermatofibroma and is often misdiagnosed.**

OBJECTIVE. **To describe the clinical and histologic features of atrophic dermatofibroma.**

METHODS. **Case report and review of the literature.**

RESULTS. **A 45-year-old white woman was found to have a 7 mm × 5 mm atrophic, depressed lesion in the right axilla. A clinical diagnosis of anetoderma was made and the lesion was excised with minimal margins. Histopathologic examination revealed findings consistent with dermatofibroma. However, focal CD34 staining and involvement of superficial subcutaneous tissue raised concern regarding an early dermatofibrosarcoma protuberans developing in a dermatofibroma. The patient underwent Mohs micrographic surgery (MMS) for definitive treatment. The final diagnosis is thought to be atrophic dermatofibroma. Reported cases in the English language literature are reviewed and the clinical and histopathologic findings are described.**

CONCLUSION. **Atrophic dermatofibroma is a well-described, yet uncommon, variant of dermatofibroma. It is often clinically misdiagnosed, and histopathologic evaluation can be misleading. The clinician and pathologist should consider this diagnosis in the evaluation of atrophic, depressed lesions.**

A. HENDI, MD, D. M. JUKIC, MD, D. W. KRESS, MD, AND D. G. BRODLAND, MD HAVE INDICATED NO SIGNIFICANT INTEREST WITH COMMERCIAL SUPPORTERS.

PAGE AND Assaad[1] published the first report of an atrophic dermatofibroma in 1987. Since that report there have been other published cases of atrophic dermatofibroma.[1–5] Yet this variant of dermatofibroma is uncommon and often misdiagnosed initially. The unusual appearance of atrophic dermatofibroma is characterized by inward puckering or flat lesions that on palpation become depressed,[2–4] or "sink in" during the biopsy.[1] This variant of dermatofibroma should be considered in the diagnosis of inwardly puckered, depressed lesions.

Case Report

On cutaneous examination, an incidental unusual finding was noted in a healthy 45-year-old white woman. In the right axilla the patient had a 7 mm × 5 mm asymptomatic, soft, atrophic, and depressed lesion adherent to the subcutaneous tissue (Figure 1). She denied a history of trauma or injections to that site. A clinical diagnosis of anetoderma was made and the lesion was excised with minimal margins. Histopathologic examination revealed a well-defined fibrohistiocytic dermal tumor with epidermal hyperplasia and focal basilar hyperpigmentation above the lesion. There was thinning of the involved dermis compared to adjacent normal skin, and "lacelike" involvement of the superficial subcutaneous tissue (Figure 2). Immunohistochemical staining was prominently positive for factor XIIIa (Figure 3), and focally positive for CD34 in the lower portion of the tumor that infiltrated the panniculus (Figure 4). CD34 staining and involvement of superficial subcutaneous tissue raised concern regarding an early lesion of dermatofibrosarcoma protuberans, and therefore the patient underwent Mohs micrographic surgery (MMS) for definitive treatment. During MMS, the tumor was noted at the deep, subcutaneous margin, necessitating a second stage. Of note, the patient subsequently underwent excision of three typical dermatofibromas from the lower extremities. Histologic and immunohistochemical evaluation supported the diagnosis of dermatofibroma in these lesions.

Discussion

Atrophic dermatofibroma is a variant of dermatofibroma with distinct clinical features.[1–5] It is estimated

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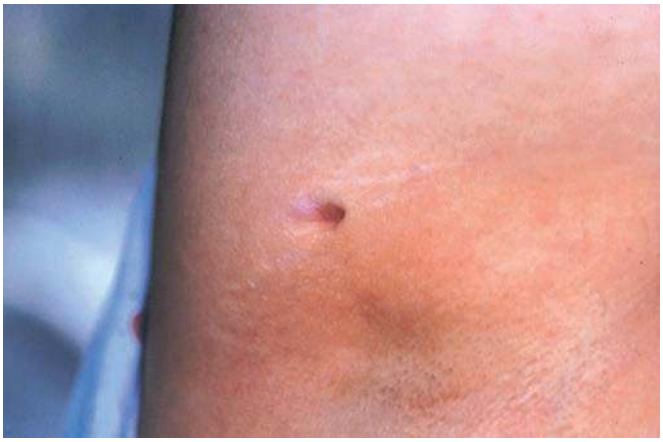

Figure 1. Depressed lesion in the right axilla.

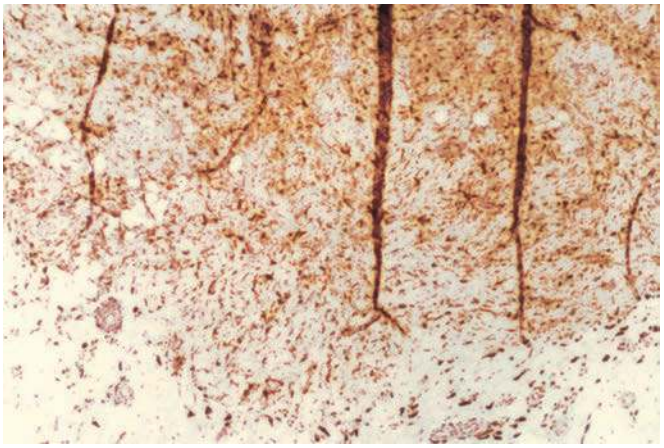

Figure 3. Prominent cellular immunostaining for factor XIIIa in the main body of the neoplasm. (Factor XIIIa immunostain; original magnification 10×.)

to account for 2% of all dermatofibromas in one study.[3] Although atrophic dermatofibroma is an accepted term in the literature, Requena and Reichel[5] have proposed changing the name to "delled dermatofibroma." In addition to atrophic dermatofibroma, there are many other clinical variants of dermatofibroma. Other reported clinical variants include atypical polypoid dermatofibroma,[6] generalized eruptive histiocytoma,[7] grouped palmoplantar histiocytoma,[8] grouped multiple histiocytomas,[9] multinodular hemosiderotic dermatofibroma,[10] giant dermatofibroma,[11] subungal dermatofibroma,[12] erosive dermatofibroma,[13] lichenoid dermatofibroma,[13] ulcerated dermatofibroma,[13] and subcutaneous fibrous histiocytoma.[14]

Whereas the typical dermatofibroma usually occurs on the lower extremities of young to middle-aged women, atrophic dermatofibroma tends to occur on the upper trunk and upper extremities of middle-aged women (40–50 years).[2,3] The typical dermatofibroma is a firm, pink-brown papule often with peripheral darker pigment. Atrophic dermatofibromas are described as being flat, umbilicated erythematous retractions of the skin"[2,3] or slightly raised oval plaques, which become depressed by finger pressure.[4] In many cases the initial clinical diagnosis is not dermatofibroma. The lesion may be diagnosed as morphea, atrophoderma, neurofibroma, localized lipoatrophy, resolving panniculitis, steroid atrophy, anetoderma, basal cell carcinoma, or nevus.[1–4] Clinical dermal atrophy is a common phenomenon in dermatofibromas as demonstrated by the buttonhole sign or dimpling with lateral pressure. However, it appears that this feature

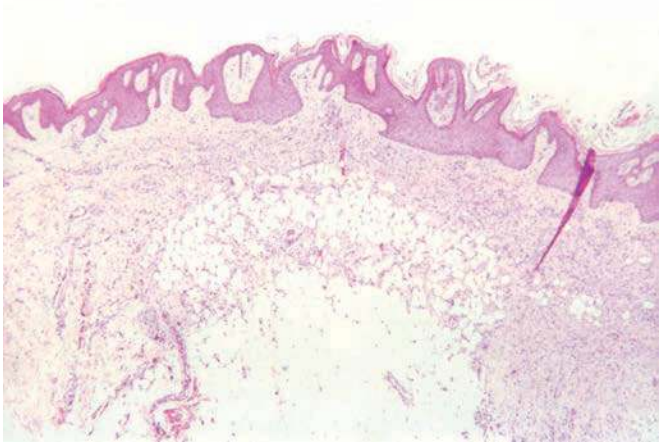

Figure 2. Epidermal hyperplasia overlying dermal neoplasm with markedly thinned dermis and involvement of the subcutaneous tissue in dermatofibrosarcoma-like fashion. (Hematoxylin and eosin; original magnification 2×.)

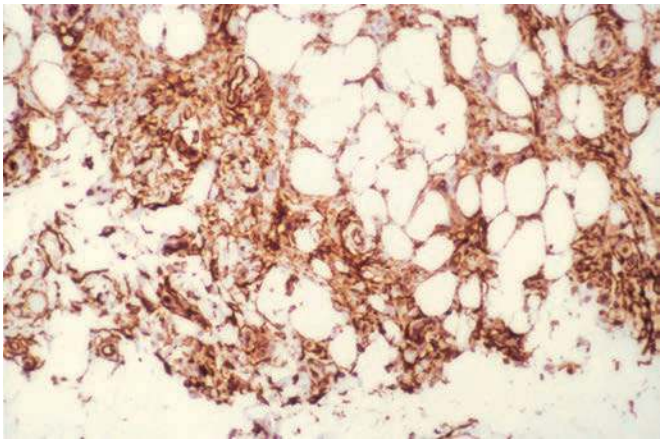

Figure 4. Focal immunostaining for CD34 in the fat-infiltrating portion of the lesion. (CD34 immunostain; original magnification 20×.)

is exaggerated in the atrophic variant of dermatofibroma;[3] hence in the resting state these lesions have a dimpled appearance or become depressed by slight finger pressure.[4] This has been attributed to focal loss of elastic fibers due to "elastophagocytosis" by the dermatofibroma cells.[4]

Histologic findings of atrophic dermatofibroma are similar to that of the "typical" dermatofibroma. A dermatofibroma is a benign, poorly demarcated dermal nodule composed of young and mature collagen, fibroblasts, capillaries, and histiocytes. Other findings include epidermal hyperplasia, basal pigmentation, sebaceous and/or hair follicle formation, peripheral sclerosis, and a smooth or scalloped lower margin.[3] In an atrophic dermatofibroma the thickness of the dermis is reported to be less that 50% of the surrounding dermis.[3] This histologic feature accounts for the dimpled appearance or depressed feel observed in atrophic dermatofibromas.

Typically, immunohistochemical labeling of a dermatofibroma reveals positive staining for factor XIIIa and negative staining for CD34. The converse is true for most dermatofibrosarcomata protuberans. In a study of 26 atrophic dermatofibromas, Zelger et al.[3] reported all but two to be positive for factor XIIIa. Twenty of these lesions were variably positive for factor XIIIa (10–50% of the tumor) and 4 had only minor reactivity (10%). Two of the 26 atrophic dermatofibromas (which stained for factor XIIIa) stained for CD34 at the periphery of the lesion. This was the case in our patient. Zelger et al.[3] attribute this to a "background/demarcation" phenomenon. Despite this aberrant CD34 staining, other clinical and histologic criteria should be used to differentiate atrophic dermatofibroma from dermatofibrosarcoma protuberans. Although atrophic dermatofibrosarcoma protuberans may have a depressed appearance, similar to that of an atrophic dermatofibroma, they typically are larger (3–6 cm versus 0.5–1 cm) with an irregular outline.[3] On histologic evaluation, dermatofibrosarcoma protuberans are ill-defined neoplasms and involve a substantial amount of the subcutaneous tissue. They also lack the epidermal hyperplasia and basal hyperpigmentation seen in dermatofibromas.[3] These features are helpful in differentiating atrophic dermatofibroma from dermatofibrosarcoma protuberans, yet equivocal cases are sure to arise. It should be noted that the center of an atrophic dermatofibroma may stain negative for factor XIIIa; loss of immunoreactivity for factor XIIIa is seen in "old" dermatofibromas and is thought to be secondary to increasing sclerosis[3] and fibroblast-like differentiation of dendrocytes.

Atrophic dermatofibroma is a well-described clinical variant of dermatofibroma. Its rarity and subtle clinical appearance often precludes accurate clinical diagnosis. Atrophic dermatofibroma should be considered in the differential diagnosis of atrophic, depressed lesions on the upper body of middle-aged women.

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