Adenodermatofibroma: a rare variant

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Abstract

Adenodermatofibroma is a newly recognized variant of fibrous histiocytoma (dermatofibroma), a benign lesion frequently encountered in dermatologic practice. There are many established variants of fibrous histiocytoma but there are only eight reported cases of this specific variant in the literature. This report reviews a case of an adenodermatofibroma presenting as a large, firm, atrophic plaque on the thigh. Histopathologic findings showed dilated glandular structures with apocrine features within a fibrohistiocytic cellular infiltrate, consistent with the diagnosis. We review the characteristic findings of adenodermatofibroma, discuss the differential diagnosis, and examine current theories speculating the origin of apocrine glands present within these lesions.

Keywords: adenodermatofibroma, benign fibrous histiocytoma, cutaneous, dermatofibroma

Introduction

Adenodermatofibroma was first named in 2013 as a new variant of fibrous histiocytoma. Numerous distinct variants of dermatofibromas have been described, not limited to hemosiderotic, lipidized, clear cell, aneurysmal, palisading, keloidal, myxoid, lichenoid, and cellular [1]. Before 2013 and dating back to 2005, lesions with similar characteristics were described as "apocrine gland cysts" with "hemosiderotic dermatofibroma-like stroma" [2,3]. At present, there are only eight reports of adenodermatofibroma in the literature. These lesions are characterized by the unique presence of dilated glandular structures within the classic fibrohistocytic cellular infiltrate characteristic of fibrous histiocytomas. The glandular structures visualized in these lesions often exhibit apocrine features with decapitation secretion.

At present, there is skepticism surrounding the origin of the glandular structures within adenodermatofibromas. Proposed theories include the entrapment of apocrine glands, a collision of a dermatofibroma with a separate apocrine glandular tumor, the induction of apocrine cysts by the underlying dermatofibroma, or the entrapment of eccrine glands which later undergo apocrine metaplasia [3,4]. We present a case of adenodermatofibroma, review the differential diagnosis, and discuss current theories which speculate the origin of apocrine glands present within these lesions.

Case Synopsis

A 53-year-old healthy man presented for evaluation of a large, asymptomatic "indentation" on his left thigh that had been present for over 20 years. He did not recall any prior injury or trauma to the area. He described noticing a much smaller papule many years ago which then expanded in size and depth but remained unchanged thereafter. He had not pursued any prior treatments and his medical and family history were unremarkable. Examination revealed a 0.9×1.1cm firm, atrophic plaque of dark brown to purple color on the medial left thigh (**Figure 1**). Given the size and uncertain nature of the

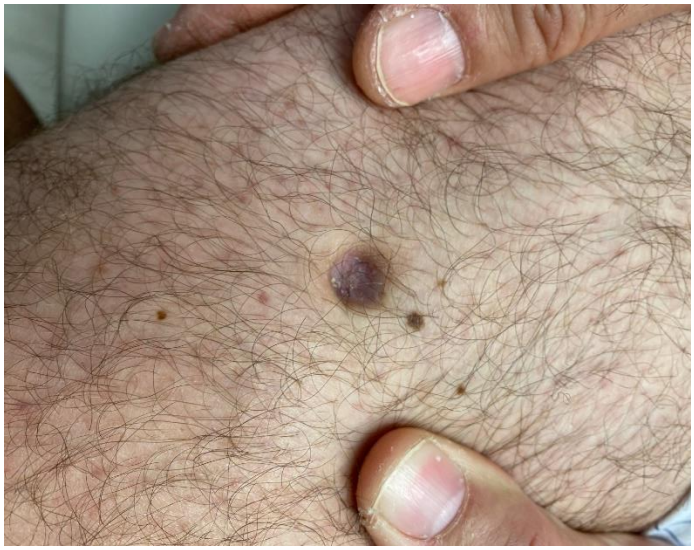

Figure 1. *A 0.9×1.1 cm firm, atrophic, dark brown to purple plaque on the left medial thigh.*

lesion, it was surgically excised for histopathologic evaluation of which representative hematoxylin and eosin-stained sections are shown (**Figure 2**).

Microscopic examination showed a dense spindle cell proliferation within which were large dilated glandular structures (**Figure 3**). Cells lining the glandular structures showed apocrine-type secretion. Epidermal acanthosis and basofollicular induction were noted overlying the proliferation. Prominent collagen trapping was visualized at the periphery of the lesion. Staining for factor XIIIa was positive and CD34 was negative. Together with the clinical and histopathologic findings, a rare variant of fibrous histiocytoma, adenodermatofibroma was diagnosed.

Case Discussion

Adenodermatofibroma is a newly established variant of fibrous histiocytoma (dermatofibroma), a benign lesion frequently encountered in dermatologic practice. There are multiple different variants of dermatofibromas and the lesions are known to induce superior epithelial, follicular, or sebaceous proliferation. This is the only variant, however, that is known to contain glandular structures as a characteristic feature. Available case reports in the literature describe these glands as dilated with apocrine-type secretion.

Current theories explaining the presence of apocrine glands within this entity include the entrapment of apocrine glands, a collision of a dermatofibroma with a separate apocrine glandular tumor, the induction of apocrine cysts by the underlying dermatofibroma, or the entrapment of eccrine glands which later undergo apocrine metaplasia [3,4]. Seven of the eight reported cases describe lesions identified on the back, forehead, calf, scapula, and thigh, areas that apocrine glands are typically not found. Thus, it has been suggested that eccrine metaplasia may best explain this phenomenon.

The differential diagnosis of adenodermatofibroma may include Kaposi sarcoma, tubular apocrine adenoma, or multinucleate cell angiohistiocytoma. Kaposi sarcoma may present as violaceous nodules on the extremities; however, this entity classically occurs in immunocompromised individuals. Histologic features vary between the patch and plaque stages of the condition but characteristic findings include slit-like vascular spaces within a

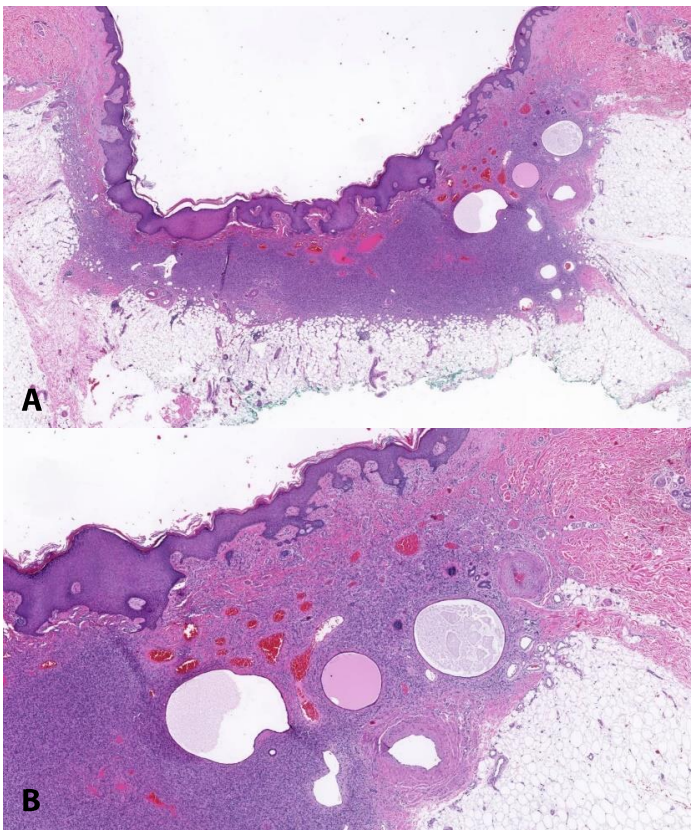

Figure 2. *H&E histopathology of the excised lesion at **A)** low power, 20×, **B)** showing dilated glandular structures, hemosiderotic cellular stroma and collagen trapping at the periphery, 100×.*

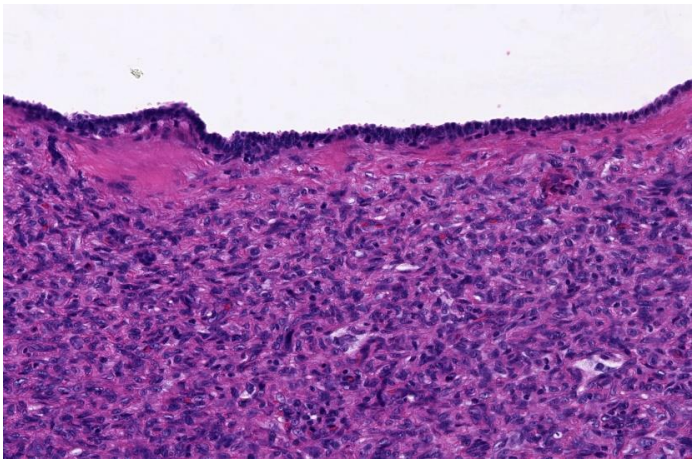

Figure 3. *Dilated glandular structures at high power. H&E, 400×.*

dermal infiltrate of eosinophilic, CD34-positive spindle cells [5]. Kaposi sarcoma can be differentiated from other vascular tumors by positive HHV-8 immunostaining. Tubular apocrine adenomas typically occur on the scalp within a nevus sebaceous but have also been reported to occur on the face, breast, axilla, or genitalia [6]. Histologically, these lesions can be described as dermal nodules containing glandular tubules of differing sizes. They lack the fibrohistiocytic infiltrate and collagen trapping observed in dermatofibromas [5,6]. Multinucleate cell angiohistiocytomas typically present as multiple asymptomatic papules on the extremities of middle-aged women. Histologic features include a proliferation of vessels, surrounded by hyalinized collagen and CD68+ multinucleate cells. Clinically, these lesions may resemble atrophic dermatofibromas but rarely present as a single lesion.

Conclusion

In conclusion, adenodermatofibroma is a new and rare entity, and further reporting and observation may provide insight into the mechanisms by which apocrine glands appear within these lesions. The most likely phenomenon is for eccrine glands to become entrapped within a fibrous histiocytoma and later undergo apocrine metaplasia. The discovery of the adenodermatofibroma adds to the many recognized variants of fibrous histiocytoma and encourages further interest in understanding the ability of these lesions to induce changes in the surrounding epithelium and adnexal structures. Our patient was instructed to follow up with our office should he have any concern for recurrence. He has had an uneventful postoperative course without signs of recurrence or further sequelae. In review of the literature, there have been no reported recurrences of these lesions after at last 6 months following excision [3].

Potential conflicts of interest

The authors declare no conflicts of interest.

References

1. Alves JV, Matos DM, Barreiros HF, Bártolo EA. Variants of dermatofibroma--a histopathological study. *An Bras Dermatol*. 2014;89:472-7. [PMID: 24937822].
2. Santos-Briz A, Llamas-Velasco M, Arango L, et al. Cutaneous adenodermatofibroma: report of two cases. *Am J Dermatopathol*. 2013;35:e103-5. [PMID: 23334518].
3. Muto I, Kuwahara F, Shintani T, et al. Adenodermatofibroma possessing dilated glandular structures with eccrine features: A case study. *J Cutan Pathol*. 2018;45:623-628. [PMID: 29752738].
4. Gonzalez S. Apocrine gland cyst with hemosiderotic dermatofibroma-like stroma: report of two cases. *Am J Dermatopathol*. 2005;27:36-8.[PMID: 15677975].
5. Calonje E, Damaskou V, Lazar A. Connective Tissue Tumors. In:McKee's pathology of the skin. Calonje E, Brenn T, Lazar A, Billings S, editors. 5th ed. Elsevier Limited; 2020. p, 1698-894.
6. Rulon DB, Helwig EB. Papillary eccrine adenoma. *Arch Dermatol*. 1977;113:596-8. [PMID: 857729].
7. Nguyen AH, Glembocki DJ, Patel NB. Multinucleate cell angiohistiocytoma. *Cutis*. 2017;100:429-31. [PMID: 29360886].