Case Report

A Case of Atrophic Dermatofibroma Overexpressing Matrix Metalloproteinase-1

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Abstract
This case report describes the importance of considering this tumor as one of the differential diagnoses when we encounter a flat and/or atrophic and depressible lesion in the upper portion of the trunk.

Introduction
Dermatofibroma is a benign fibrohistiocytic tumor with several variants classified by their clinical and histopathological features as follows: common histiocytoma (80%) and aneurysmal (5.7%), epithelioid (2.6%), cellular (2.1%), atrophic (1.0%), and clear-cell variants [1]. The rare variant atrophic dermatofibroma is characterized clinically by a flat and/or atrophic and depressible surface, a case of which is reported here.
Case Presentation

A 45-year-old male had noticed a brown spot on his right shoulder a year earlier, whose surface was atrophic and recessed. The lesion was well defined and 1 cm in diameter (Fig. 1a, b). He had neither subjective symptoms nor a history of trauma. Hematoxylin and eosin staining of the excised lesion revealed epidermal hyperplasia and remarkable dermal atrophy. Subcutaneous adipose tissue was seen immediately under the atrophic dermis (Fig. 1c). The tumor was well defined, showing spindle cell proliferation with small nuclei but no atypia (Fig. 1d). The tumor cells were factor XIIIa positive (Fig. 1e) and CD34 negative (Fig. 1f), leading to a diagnosis of atrophic dermatofibroma. Because we observed reduced volume of the extracellular matrix of the dermis in the lesional atrophic area, we performed Elastica van Gieson staining. Elastic fibers in the lesional dermis had disappeared or were greatly decreased in number (Fig. 1g) compared with the perilesional normal skin (Fig. 1h), which is consistent with a previous report [2]. We also evaluated the lesional expression of matrix metalloproteinase-1 (MMP-1), an important enzyme for degrading elastic fibers. We found a strong expression of MMP-1 in the atrophic dermis on the lower part of the dotted line compared to the perilesional dermis on the upper part (Fig. 1i). The expression of MMP-1 in the atrophic area of the lesional dermis was particularly prominent (Fig. 1j).

Discussion/Conclusion

Atrophic dermatofibroma is a rare variant of dermatofibroma, first described by Page and Assaad [3] in 1987. The entity was subsequently defined by Zelger et al. [4] as a dermatofibroma with dermal atrophy of more than 50% of the locoregional dermis. To date, 48 cases have been reported; the average age of affected patients was 49.7 years and the female-to-male ratio was 43:5. The upper trunk is the predominant site. Typical atrophic dermatofibroma is often associated with elastophagocytosis of collagen fibers [2]. In parallel, we first demonstrated the overexpression of MMP-1 by tumor cells in the present case. The elevated lysis of elastic fibers by MMP-1 may cause the atrophic dermatofibroma. There was one case of aneurysmal atrophic dermatofibroma [5]. Elastolysis may be related to the aneurysmal feature.

Because atrophic dermatofibroma is characterized by its flat and/or atrophic and depressible surface, several differential diagnoses should be ruled out: atrophic dermatofibrosarcoma protubersans, anetoderma, atrophic scarring, sclerodermiform epithelioma, morphea, atrophoderma, and localized lipoatrophy. We have to consider this tumor as one of the differential diagnoses when we encounter flat and/or atrophic and depressible lesions in the upper trunk of females.

Statement of Ethics

The authors have no ethical conflicts to disclose. Informed consent was obtained from the patient.
Disclosure Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All authors are responsible for and agree with the content and writing of the paper to which we all contributed significantly.

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Fig. 1. a, b A brown spot of 1 cm in diameter on the right shoulder, the surface of which was atrophic and recessed. c H&E staining (low magnification). Remarkable atrophy of the dermis. The tumor was well defined. Arrowheads indicate the tumor lesion. Subcutaneous adipose tissue was observed immediately under the dermis. d H&E staining (high magnification). Proliferation of spindle cells with small nuclei was observed. There was no atypia. e Immunohistochemical staining of factor XIIIa. The tumor cells were positive for factor XIIIa. f Immunohistochemical staining of CD34. The tumor cells were negative for CD34. g, h Elastica van Gieson staining. Elastic fibers in the atrophic area of the lesional dermis disappeared or greatly decreased in number (g) relative to those of the intact lesion (h). Arrowheads indicate elastic fibers. i, j Staining of matrix metalloproteinase-1 (MMP-1). i The staining of MMP-1 in the boundary area of atrophic area and perilesional area is shown. There was a strong expression of MMP-1 in the atrophic dermis on the lower part of the dotted line compared to the perilesional dermis on the upper part. j The expression of MMP-1 in the atrophic dermis was particularly prominent in comparison to that in the perilesional dermis. Brown staining indicates MMP-1 expression.