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A Case of Atrophic Dermatofibrosarcoma Protuberans

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Dear Editor:

Dermatofibrosarcoma protuberans (DFSP) is a rare infiltrative skin tumor with low malignancy, slow growth, but easy to relapse. Atrophic DFSP is a rare clinical variant[1].

A 39-year-old female presented with an asymptomatic, gradually enlarging atrophic reddish-brown plaque on the right lower leg for 7 years. The initial skin lesion appeared as a mung bean-sized light red plaque. Four years ago, the patient did skin biopsy in local hospital, but misdiagnosed as dermatofibroma and without any treatment. Recently, atrophy of the lesion aggravated with mild pain occasionally.

Cutaneous examination revealed an infiltrating, atrophic, morphea-like and dark reddish plaque with a size of about 2 mm in depth and 15 mm in diameter on the right tibial anterior skin (Fig. 1). Histopathology examination of the lesion revealed it was a cutaneous spindle cells derived tumor. Further immunohistochemistry demonstrated the tumor with positivity of CD34, Vimentin and factor XIII (Fig. 2). According to the manifestation, a diagnosis of atrophic DFSP was made.

DFSP is locally invasive, grows slowly and has a high recurrence rate. Lesions are usually reddish-brown plaques or nodules, which can progress to multiple nodular lumps locally. However, atrophic DFSP are usually presented as atrophic or sclerotic plaques, which are more easily misdiagnosed as scleroderma, panniculitis and dermatofibroma. In this case, the patient was misdiagnosed as dermatofibroma early, which delayed the optimal timing of treatment. Histologically, besides atrophic dermis, compared with typical DFSP, the atrophic variant shows non-specific manifestations. In addition, in immunohistochemistry examination, atrophic DFSP also presented as diffuse expression of CD34, positivity of vimentin and factor XIII[1].

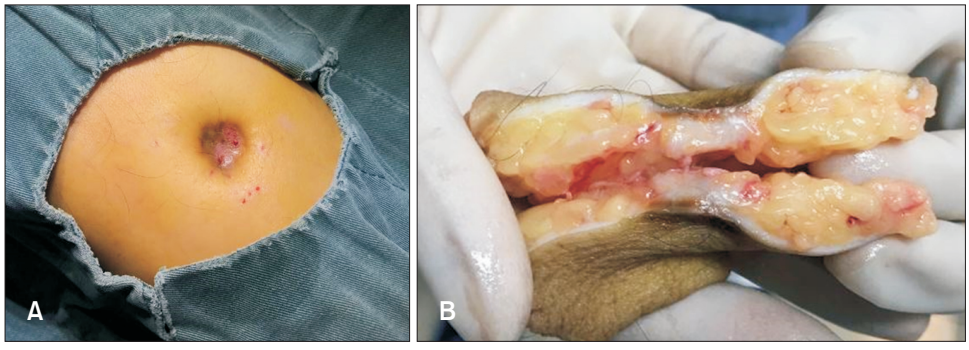

Fig. 1. Clinical features. (A) Preoperative appearance. (B) Atrophy of the lesion. We received the patient's consent form about publishing all photographic materials.

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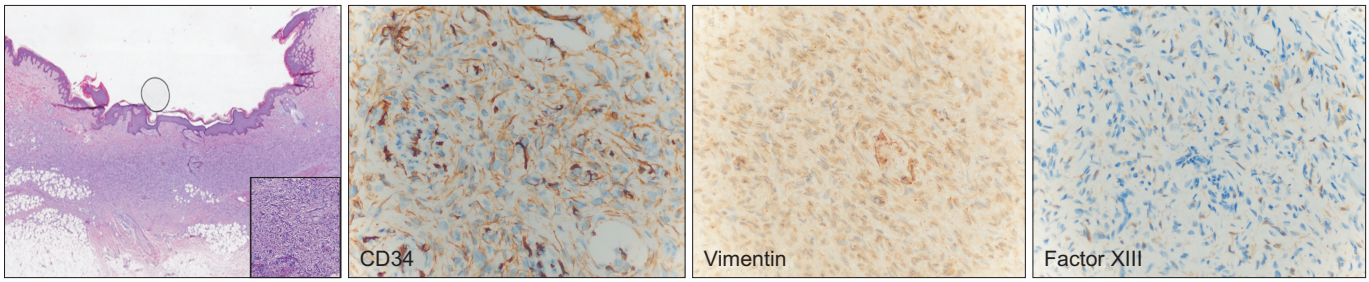

Fig. 2. Histopathologic section. H&E staining results suggested atrophic spindle cell tumor (10×). Immunohistochemistry results suggested dermatofibrosarcoma protuberans: CD34 (+), Vementin (+), Factor XIII (+) (Immunohistochemistry, 40×).

Mohs surgery is the first-line treatment of DFSP and can reduce the risk of recurrence to 2.72%, which is significantly lower than traditional surgery with a surgical margin of at least 2 to 3 cm (9.1%). Former study revealed that frozen Mohs surgery has the advantages of shorter surgery time and immediate closure, which is as effective as paraffin Mohs surgery in the treatment of DFSP[2,3]. As the pathogenesis of DFSP, the fusion of chromosomal platelet-derived growth factor β-chain (*PDGFB*) genes may be one of the mechanisms leading to tumorigenesis. It indicates that imatinib, an inhibitor of PDGFB-receptor can be used in patients with advanced stages[2,4]. In this case, local enlarged resection with a surgical margin about 3 cm was used and a follow-up for 4 months showed no local recurrence and distant metastasis.

Generally, due to the atypical clinical manifestations of atrophic DFSP, it is frequently neglected by the patients and easily misdiagnosed as benign lesions by dermatologists. As with this case, for atrophic lesions with no apparent cause and no symptoms, should be aware of atrophic DFSP in early stage.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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REFERENCES

1. Bichakjian CK, Olencki T, Alam M, Andersen JS, Berg D, Bowen GM, et al. Dermatofibrosarcoma protuberans, version 1.2014. J Natl Compr Canc Netw 2014;12:863-868.
2. Rutkowski P, Dębiec-Rychter M, Nowecki Z, Michej W, Symonides M, Ptaszynski K, et al. Treatment of advanced dermatofibrosarcoma protuberans with imatinib mesylate with or without surgical resection. J Eur Acad Dermatol Venereol 2011;25:264-270.
3. Lee SH, Oh Y, Nam KA, Oh B, Roh MR, Chung KY. Mohs micrographic surgery for dermatofibrosarcoma protuberans: comparison of frozen and paraffin techniques. J Eur Acad Dermatol Venereol 2018;32:2171-2177.
4. McGee MW, Boukhar SA, Monga V, Weigel R, Phadke SD. Dermatofibrosarcoma protuberans - the use of neoadjuvant imatinib for treatment of an uncommon breast malignancy: a case report. J Med Case Rep 2019;13:374.