

## A Case of Atrophic Dermatofibrosarcoma Protuberans

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Atrophic dermatofibrosarcoma protuberans (DFSP) is a rare variant of DFSP, which presents as a depressed scar-like lesion with atrophic patch. Although atrophic DFSP is identical to classical DFSP in biologic and histologic features, such atrophic lesions can be difficult to diagnose clinically. Better knowledge of atrophic DFSP is important for making an early diagnosis and for proper surgical treatment. We report the case of a 33-year-old female presented with a gray depressed atrophic patch with a skin-colored, pea-sized nodule on the left lateral rim, and an erythematous indurated plaque on her upper back. Histopathologically, the atrophic patch showed densely packed, monomorphous, plump spindle cells arranged in a storiform pattern, infiltrating the dermis and subcutaneous tissue, producing a characteristic honeycomb pattern. The tumor cells exhibited strong and diffuse positivity with CD34, establishing the diagnosis of DFSP.

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### INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a slow-growing, locally aggressive tumor marked by its high rate of local recurrence<sup>1</sup>. It usually begins as a red-brown indurated plaque, and develops nodules slowly over many years<sup>2</sup>. Rarely, the initial skin lesion may be atrophic or depressed, and may persist despite the advance stage of tumor. Atrophic DFSP is a rare variant of DFSP, which presents as a depressed violaceous scar-like lesion with atrophic patch. The biologic behavior and histologic characteristics of atrophic DFSP is identical to classical DFSP. However, clinical diagnosis is sometimes difficult for the atrophic lesion<sup>1</sup>. We report an unusual case of atrophic DFSP in a 33-year-old female.

### CASE REPORT

A 33-year-old female presented with gray depressed atrophic lesion accompanying a skin-colored pea-sized nodule arising from it, and erythematous indurated plaque with intermittent itching and pricking on her upper back. She had first noticed an asymptomatic erythematous plaque 7 years ago. Later, a skin-colored nodule had developed on the left lateral rim of that lesion and other part became depressed with grayish discoloration. And then an erythematous indurated plaque newly developed below the preexisting lesion (Fig. 1). There was no history of trauma, operation, or biopsy in the vicinity of the lesion. Laboratory tests including complete blood count, blood chemistry analysis, and urinalysis were normal. An incisional biopsy was taken from depressed atrophic lesion including skin colored nodule. The biopsy specimen showed densely-packed, monomorphous, plump spindle cells arranged in a storiform pattern, infiltrating the dermis (Fig. 2A, C) and subcutaneous tissue, producing a characteristic honeycomb pattern (Fig. 2B). Using immuno-histochemical stain, the biopsy specimen exhibited strong and diffuse positivity with CD34 (Fig. 2D). The diagnosis of DFSP was made based

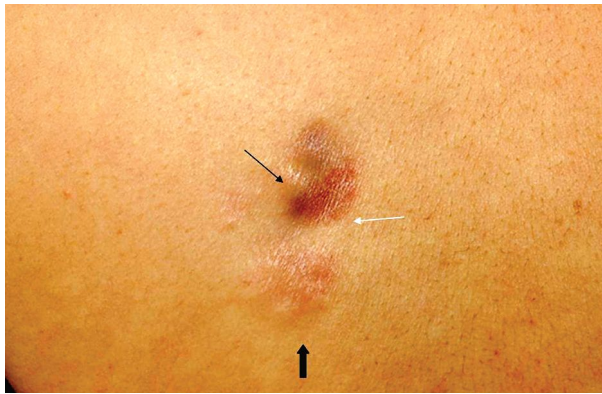
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on clinical, histological features and immunohistochemical stain. The patient was referred to the

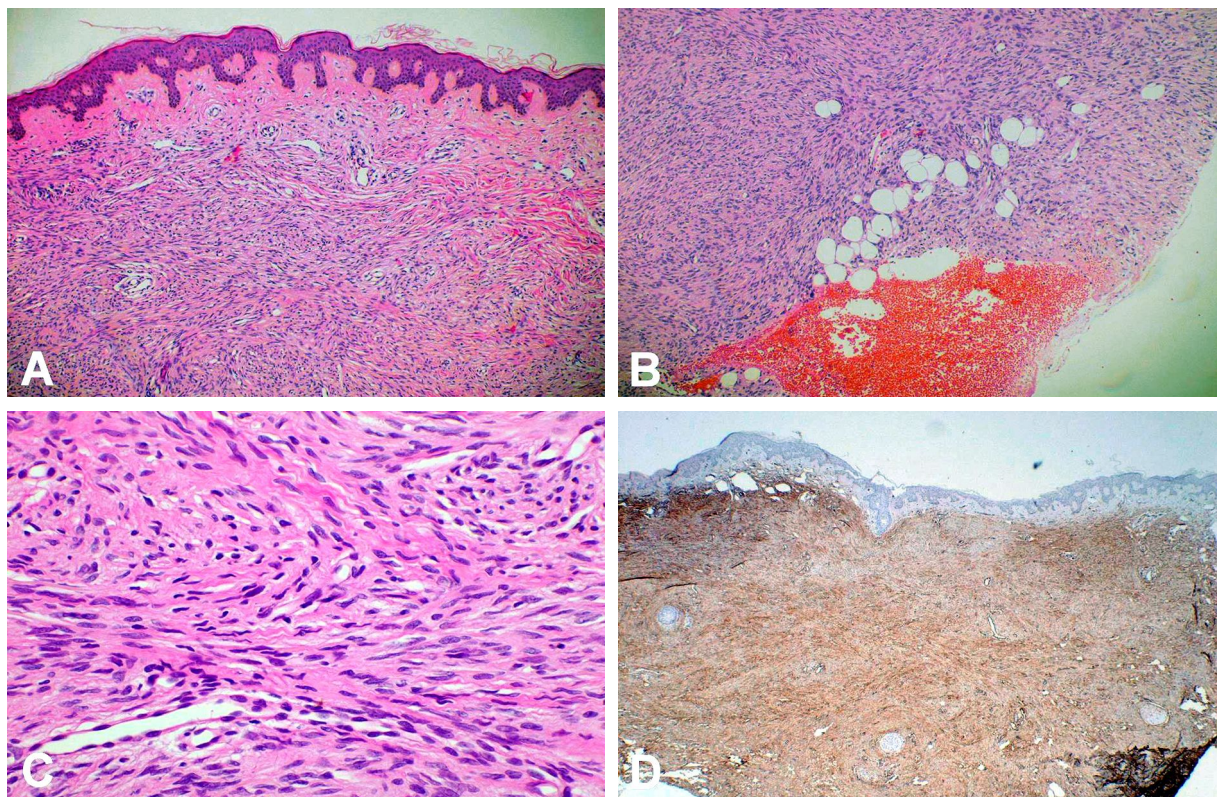


**Fig. 1.** Gray depressed atrophic lesion (white arrow) with skin colored pea-sized nodule (black arrow) and erythematous indurated plaque (bold arrow) are seen on the upper back.

department of plastic surgery for wide excision. However, follow-up failed because the patient transferred to a neighboring hospital of her own accord.

## DISCUSSION

DFSP is an uncommon mesenchymal neoplasm originating in the dermis. About 1,000 cases have been reported since DFSP was first described<sup>3</sup>. It usually occurs as an indurated plaque that slowly increases in size and develops multiple firm nodules. Rarely, the initial plaque may be atrophic or depressed, and this atrophic appearance may persist. Lambert et al<sup>4</sup> first described 5 cases that resembled morphea or morpheaform BCC but showed typical DFSP on biopsy. They suggested that the term *protuberans* should be discarded from the name *dermatofibrosarcoma protuberans*.



**Fig. 2.** Dense spindle cell proliferation with a storiform arrangement infiltrating the dermis (A) and subcutaneous tissue, producing honeycomb pattern (B). Higher magnification shows densely packed, monomorphous, plump spindle cells with some atypical mitotic figures (C). Marked positivity with immunohistochemical marker CD34 (D) (H & E; A,B,D;  $\times 100$ , C;  $\times 200$ )

Atrophic DFSP is an uncommon clinical variant of DFSP. To our knowledge, there have been 31 reported cases of atrophic DFSP to date<sup>1,3-8</sup>. While atrophic DFSP has a distinct clinical appearance, its epidemiology, histology and clinical behavior appear to be similar to the common protuberant type. Truncal involvement is seen in 79% of cases, and there is a slight female predominance (62%), with the average age of onset being 30 years<sup>5</sup>. It presents as a large, irregularly outlined, tan to brownish depressed scar-like lesion with atrophic patch. It is generally asymptomatic, slow growing, and often benign appearance, thus diagnosis is frequently delayed (median delay 6 years)<sup>6</sup>. It may be clinically confused with other atrophic or sclerotic dermatologic conditions, such as morphea, anetoderma, morpheaform basal cell carcinoma, scar, and lymphocytoma<sup>1,5,6</sup>.

Histopathologically, atrophic DFSP is characterized by a dense spindle cell proliferation arranged in a cartwheel appearance infiltrating the surrounding dermis and subcutaneous tissue. The tumor shows a deep/irregular infiltration of fatty tissue in a lacelike/honeycomb and multilayered pattern. Tumor cells are uniform spindle cells with little cytoplasm and elongated hyperchromatic, but not pleomorphic, nuclei. Usually there is little mitotic activity. The epidermis is normal or flat. The thickness of the dermis is reduced to < 50% of surrounding dermis, placing the subcutis close to the epidermis<sup>3</sup>. The use of immunohistochemistry is a helpful adjunct to identify the tumor. The atrophic DFSP are consistently CD34 positive, factor XIIIa and metallothionein-negative<sup>9</sup>.

Mohs' micrographic surgery using immunohistochemical staining with CD34 on frozen section, is widely regarded as the treatment of choice for DFSP<sup>2</sup>. Because lateral extensions of tumor cells into surrounding dermis are common, wide surgical excision might not be effective for these tumors. The rate of local recurrence is estimated at 70% with surgical margins of 1 cm, 40% with 2 cm, 15% with 3 cm, 5% with margins of 5 cm, compared with less than the 2% quoted in Mohs' micrographic surgery<sup>10</sup>. Recently, imatinib mesylate, which inhibits PDGF receptor tyrosine kinase, has been shown to have some effect on metastatic DFSP, and other compounds acting as platelet-derived growth factor receptor inhibitors are currently in clinical development such as SU11248 or SU9518<sup>11</sup>.

The atypical appearance of atrophic DFSP may be misdiagnosed as other sclerotic and atrophic dermatologic conditions including morphea and anetoderma. Therefore it is important to recognize that DFSP begins and may persist as a non-protuberant plaque, often with an atrophic component. Greater clinical awareness of atrophic DFSP will allow earlier diagnosis and proper management. So we highlight this atrophic variant of DFSP.

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