From Morphea to Dermatofibrosarcoma Protuberans

Dear Editor,

Morphea profunda (MP) is a chronic autoimmune disease, a subtype of localized scleroderma that presents clinically as local discomfort due to the impairment of skin motility (1). Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue neoplasm that not only infiltrates the dermis and subcutaneous tissue, but can also affect the muscles and bones with finger-like extensions, usually present on the trunk and the proximal extremities (2). DFSP is known for its indolent clinical course, locally aggressive behavior, and high local recurrence rates, but relatively low risk of metastatic spread (2). DFSP frequently arises in middle-aged adults, affecting both sexes equally with an incidence of 4 per 1,000,000 people (3).

We report the case of a 39-year-old female patient who first presented to our clinic at the age of 20 years due to a brownish atrophic coin-sized lesion appearing on the left side of the abdomen. Medical reports indicated that biopsies had been performed previously on 3 occasions, and histopathologic findings confirmed the diagnosis of MP. The aforementioned lesion on the abdomen had been growing slowly over the years, and the patient finally visited our clinic 15 years later after noticing two palpable nodules developing within the affected skin (Figure 1, A, B). Clinical examination revealed an indurated ill-defined plaque measuring 10 cm with partially atrophic surface and 2 centrally located palpable nodules measuring between 3 and 5 mm. A deep biopsy of the lesion was performed, and histopathology and immunohistochemical analysis of CD34 expression confirmed the diagnosis of dermatofibrosarcoma protuberans (Figure 1, C, D).

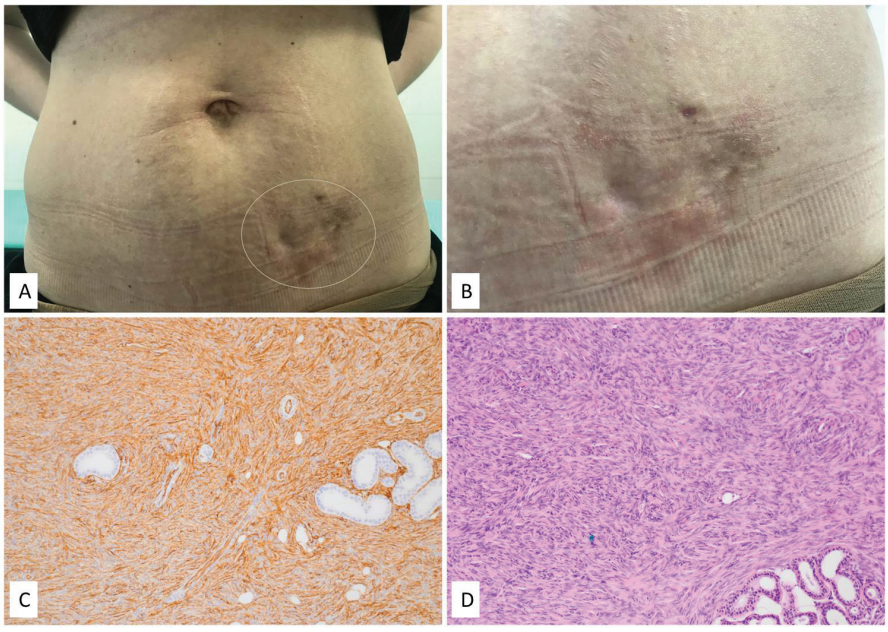

Figure 1. (A, B) Clinical presentation of morpheaform plaque on the left side of the abdomen with newly developed nodules; (C) The dermis is replaced by bundles of uniform small spindle cells, typically with spared adnexal structures (hematoxylin and eosin ×200); (D) The tumor cells characteristically express immunohistochemical positivity for CD34 (CD34 ×200).

Computed tomography scans of the thorax, abdomen, and pelvic region were subsequently performed, revealing no further disease progression. Complete excision of the tumor was performed and followed by wide scar re-excision due to narrow surgical margins of only 1 mm. No further disease progression or recurrences have been noted during the follow-up, and the patient has been disease-free for one year postoperatively.

Although the etiology of DFSP is unknown, trauma has been hypothesized as a predisposing factor. It usually presents on the trunk and the proximal extremities (4). Patients usually report disease progression over a long period of time, ranging from several months to years. The tumor is associated with variable color changes, even proximal skin discoloration, and often presents with a slowly growing indurated dermal plaque or firm nodule attached to the skin (4).

Clinically, it can be difficult to distinguish DFSP from a wide number of diagnoses, including morphea, idiopathic atrophoderma, atrophic scar, anetoderma, lipoatrophy, cellular dermatofibroma, fibrosarcoma, malignant fibrous histiocytoma, atypical fibroxanthoma, desmoplastic melanoma, Kaposi sarcoma, and solitary fibrous tumors (5). Immunohistochemistry staining for CD34 cells can be helpful in differentiation, since spindle cells stain positively in DFSP (6). Due to alteration of dermal collagen, histopathological differential diagnoses of DFSP includes lichen sclerosus, atrophic scars and keloids, as well as morphea (7), atrophic dermatofibroma, and undifferentiated pleomorphic sarcoma (6).

The mainstay of DFSP treatment is tumor excision performed either by wide local excision or Mohs surgery and having surgical margins between 1 and 5 cm. Several studies have confirmed that patients treated with the Mohs technique have significantly lower recurrence rates (8). Due to the high number of unsatisfactory primary excisions, wide free surgical margins are important for disease control (3).

Radiotherapy might be considered as a therapeutic option for inoperable tumors or relapses, as well as an adjuvant therapy after primary excision or re-excision with positive margins (8). Furthermore, recent findings indicate positive therapeutic efficacy after administration of imatinib mesilat – a tyrosine kinase inhibitor due to over expression of PDGFβ (9). Clinical follow-up of patients with DFSP after tumor excision should be performed every six months for the first five years, followed by yearly intervals thereafter for up to 10 years (3).

Previous case reports have claimed that the diagnosis of DSFP is commonly delayed as a result of slow tumor growth and nonspecific initial clinical findings (10). To the best of our knowledge, our case is the first description in the literature of DFSP developed within a MP plaque. We speculate that trauma from repeated punch biopsies taken from the sclerotic morpheaform plaque may represent the trigger for the development of the DFSP. Another notable clinical challenge was the surgical excision itself, since the majority of cases presented in literature mentioned unsatisfactory resection margins and a high risk of local disease recurrence. Although complete excision of the neoplasm was performed, re-excision was performed in order to provide wider resection margins. Surgical resection remains the main treatment for dermatofibrosarcoma protuberans, with the main challenge being the achievement of clean excision margins. Proper management of the disease and continuous follow-up are important in order to prevent local recurrence of dermatofibrosarcoma protuberans or its potential metastases.

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