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Atrophic dermatofibrosarcoma protuberans: a clinicopathological study of 16 cases

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Summary

We present a case series of atrophic dermatofibrosarcoma protuberans (DFSP) to further characterise its clinical and pathological features. Sixteen cases were enrolled in the study. There were five males and 11 females with a median age of 28 years. The vast majority occurred in the trunk (12/16, 75%), whereas a minority involved the upper limb or limb gird (2/16, 12.5%), and head and neck region (2/16, 12.5%). The most common presentation was a depressed plaque-like lesion with a greyish-red to purplish-blue colour. Histologically, the lesion was dermal-based consisting of monomorphous spindle cells arranged in parallel fascicles with focal areas displaying storiform architecture. In addition, one case showed remarkable hyalinisation of the matrix, two cases contained scattered pigmented dendritic cells and one case had admixed giant cell fibroblastoma-like component, respectively. The diagnosis was confirmed by immunohistochemical study, and by further fluorescence *in situ* hybridisation analysis in six cases. Follow-up thus far has revealed a relatively low rate of local recurrence (1/10, 10%). Familiarity with the distinctive clinical and pathological features of atrophic DFSP helps avoid misdiagnosis. Like a classical DFSP, morphological variants can also occur in an atrophic DFSP, including pigmented, sclerosing and hybrid subtypes, albeit rare.

Key words: Atrophic dermatofibrosarcoma protuberans; fluorescence *in situ* hybridisation; PDGFB.

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a well-known fibroblastic/myofibroblastic neoplasm of intermediate malignancy.[1] Besides the classical type, several morphological variants have been described, including pigmented DFSP (also known as Bednar tumour), fibrosarcomatous DFSP (FS-DFSP), myxoid DFSP, myoid/myofibroblastic DFSP, sclerosing DFSP, hybrid DFSP/giant cell fibroblastoma (GCF) and atrophic DFSP. Among these variants, only FS-DFSP has been associated with elevated risk for recurrence or metastases.[2] The importance of recognising other variants is to familiarise the pathologists with the morphological spectrum of DFSP, in an attempt to avoid potential misdiagnosis and thus inappropriate treatment. Clinically, DFSP usually manifests as an indurated nodule that protrudes on the surface of the skin, for which it is termed 'protuberans'. Atrophic DFSP, a non-protuberant form of DFSP, can be clinically and pathologically difficult to diagnose due to its atypical presentations. Although it was first described in 1985,[3] atrophic DFSP represents a distinctly rare entity as the vast majority of the reports in the literature are individual case presentations[4–10] or small series.[11] To enhance recognition, we present here a case series to further characterise its clinical and pathological features. We also expand the morphological spectrum of atopic DFSP by describing histological subtypes.

MATERIALS AND METHODS

Sixteen cases of atrophic DFSP were retrieved from the consultation files of the Department of Pathology, Fudan University Shanghai Cancer Center, between 1 January 2012 and 31 December 2018. Except for one case (Case 15), all other cases were outside referrals. The clinical features and macroscopic findings were taken from the medical records and pathology reports. The follow-up information was obtained from the referring pathologists and clinicians, or by direct telephone communication with the patients or their relatives. Immunohistochemical staining was performed on 4-μm-thick unstained sections generated from submitted paraffin-embedded blocks or directly on submitted unstained sections, with the Ventana Benchmark immunostainer (Ventana Medical Systems, USA). Antibodies used in this study included anti-pancytokeratin (AE1/AE3, dilution 1:50; Dako, Denmark), anti-CD34 (QBEnd10, dilution 1:100; Dako), α-smooth muscle actin (SMA) (1A4, dilution 1:200; Dako), anti-desmin (D33, dilution 1:100; Dako), anti-CD68 (KP1, PGM1, dilution 1:600; Changdao, China), anti-S100 protein (polygonal, dilution 1:500; Dako), anti-HMB45 (HMB45, dilution 1:50; Dako), anti-melan-A (A103, dilution 1:100; Dako), anti-STAT6 (YE361, ready-to-use; Abcam, UK), and anti-Ki67 (MIB1, dilution 1:150; Dako). Pretreatment was carried out according to the manufacturers' recommendations. Omission of primary antibody and substitution with non-specific immunoglobins were used as negative controls. Appropriate positive controls were run concurrently for all antibodies tested. Immunoreactivity was scored by percentage of tumour cell expression: (0, none; 1+, 1–25%; 2+, 26–50%; 3+, 51–100%).

Interphase fluorescence *in situ* hybridisation (FISH) was carried out in six cases (Cases 10–15) for the presence of PDGFB gene rearrangement. Five-μm thick unstained sections were incubated in a humidified chamber (HYBriteTM system; Vysis, Abbott Molecular, USA) with the LSI PDGFRB break-apart probes (Abbott Molecular/Vysis), according to the manufacturer's protocol. The fluorescence signals were analysed with an Olympus BX51 fluorescence microscope (Olympus, Japan). A total of 200 nuclei were

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evaluated from each specimen. A positive score was interpreted as more than 20% of the nuclei showing a break-apart signal.

RESULTS

Clinical features

The clinical features of 16 patients are summarised in Table 1. There were five males and 11 females with a M:F ratio of 1:2.2. The age of patients at presentation ranged from 7 to 63 years (mean 32 years; median 28 years). The majority occurred in the trunk (12/16, 75%), including chest wall (*n*=5), abdominal wall (*n*=2), infraclavicular region (*n*=2), back (*n*=2) and groin (*n*=1). Two tumours involved the upper limb or limb gird (2/16, 12.5%), one in the forearm and the other in the shoulder. The remaining two tumours developed in the head and neck region (2/16, 12.5%), with one in the cheek and another in the lower neck. Clinical information was available in 15 cases. The most common presentation was a depressed plaque-like lesion with tan-brown, greyish-red to purplish colour (Fig. 1A,B), with a mean size of 16 mm (range 5–25 mm). Preoperative duration ranged from 1 to 15 years (median 4 years). Of note, none was considered DFSP clinically. In nine cases for which the clinical impression was documented, the lesion was considered to be hyperpigmentation (*n*=2, 22%), localised scleroderma (*n*=2, 22%), morphea, skin atrophy, panniculitis, lupus and scar-like neoplasm in one case each (11%). The submitted pathological diagnoses include DFSP (*n*=4), neurofibroma (*n*=4), spindle cell neoplasm (*n*=4), spindle cell neoplasm, suspicious of DFSP (*n*=2), atrophic pigmented DFSP (*n*=1) and possible DFSP (*n*=1).

Six patients had a biopsy as initial procedure and underwent wide excision afterwards. The remaining 10 patients had a simple excision in local hospitals and all had complementary wide excision with clear margins. No adjunctive treatment was administered. Clinical follow-up was available for 10 cases (median 51 months; range 1–80 months). One patient experienced a local recurrence at 13 months and underwent curative re-excision. At the last follow-up, all nine patients had no evidence of local recurrence or distant metastasis.

Pathological features

Histologically, all tumours were dermal-based showing a characteristic plaque-like growth pattern (Fig. 2A). There was variable infiltration into the subcutaneous adipose tissue (Fig. 2B). They were composed of monomorphous spindle cells with slender wavy nuclei, arranged in parallel or horizontally oriented fascicles (Fig. 2C). Storiform architecture was present in most cases but often focally (Fig. 2D). In one case (Case 12), there was abundant interstitial collagen fibres, giving a sclerotic appearance (Fig. 2E,F). In two cases (Cases 3 and 6), there were pigmented dendritic cells scattered among the tumour cells (Fig. 2G,H). One case (Case 14) contained hyperchromatic multinucleated giant cells admixed with the spindled tumour cells (Fig. 2I,J), consistent with a hybrid DFSP/giant cell fibroblastoma. Generally, tumour cells exhibited a mild degree of nuclear atypia with low mitotic activity (0–3/10 HPF). Necrosis was absent.

Immunohistochemically, all tumours showed diffuse and strong positivity of CD34 (Fig. 3A), including multinucleated giant cells in the hybrid lesion (Fig. 3B). They were negative for AE1/AE3, α-SMA, desmin, CD68, S-100 protein, HMB45, melan-A and STAT6. The Ki-67 index ranged from 1% to 10%, with most cases being 2–5%. FISH analysis demonstrated PDGFB-related rearrangement in all six cases tested, appearing as split signals or loss of telomeric part (green) (Fig. 3C).

DISCUSSION

DFSP is a locally aggressive neoplasm characterised by slow but persistent growth over a long period. Although most patients have a history of several years, a preoperative duration as long as 10 years or more is not uncommon. In the early stage, the lesion usually manifests as a small, firm nodule or plaque-like induration which may remain stationary for years and does not receive much attention. However, rapid growth with development of a mass eventually urges the patient to seek medical attention. On physical examination, advanced DFSP may give a typical protuberant appearance accompanied with or without satellite nodules. Although DFSP is considered a neoplasm of young to middle

Table 1 Clinicopathological features of 16 cases of atrophic dermatofibrosarcoma protuberans

Case no.	Age, years/Sex	Site	Size, mm	Duration	Submitted diagnosis	Follow-up, months
1	31/F	Left lower abdominal wall	15	2 years	DFSP	UA
2	14/M	Right upper abdominal wall	20	4 years	Spindle cell neoplasm, suspicious of DFSP	NED, 60
3	7/M	Left forearm	5	4 years	Atrophic pigmented DFSP	UA
4	45/F	Infraclavicular region	UA	UA	DFSP	UA
5	23/F	Left chest wall	15	5 years	Neurofibroma	Recurred at 13, NED, 17
6	44/M	Right back	25	3 years	Neurofibroma	NED, 44
7	27/F	Shoulder	20	10 years	Possible DFSP	NED, 26
8	19/M	Right chest wall	25	10+ years	DFSP	UA
9	28/F	Infraclavicular region	10	15 years	Neurofibroma	NED, 52
10	28/F	Chest wall	15	5 years	DFSP	NED, 52
11	24/F	Left lower neck	20	4 years	Spindle cell neoplasm	UA
12	48/F	Left cheek	14	2 years	Spindle cell neoplasm	UA
13	47/F	Left chest wall	10	4+ years	Spindle cell neoplasm	NED, 79
14	34/M	Back	12	3 years	Spindle cell neoplasm, suspicious of DFSP	NED, 80
15	36/F	Left groin	20	1 year	Spindle cell neoplasm	NED, 50
16	63/F	Chest wall	10	2+ years	Cutaneous diffuse neurofibroma	NED, 1

DFSP, dermatofibrosarcoma protuberans; NED, no evidence of disease; UA, unavailable.

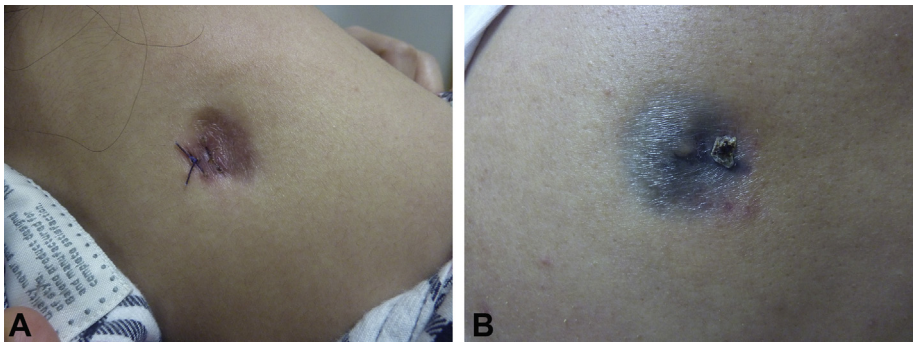

Fig. 1 Clinical features of atrophic DFSP. (A) Irregular tan brown depressed lesion (classic type, post biopsy). (B) Round bluish plaque-like lesion (pigmented variant).

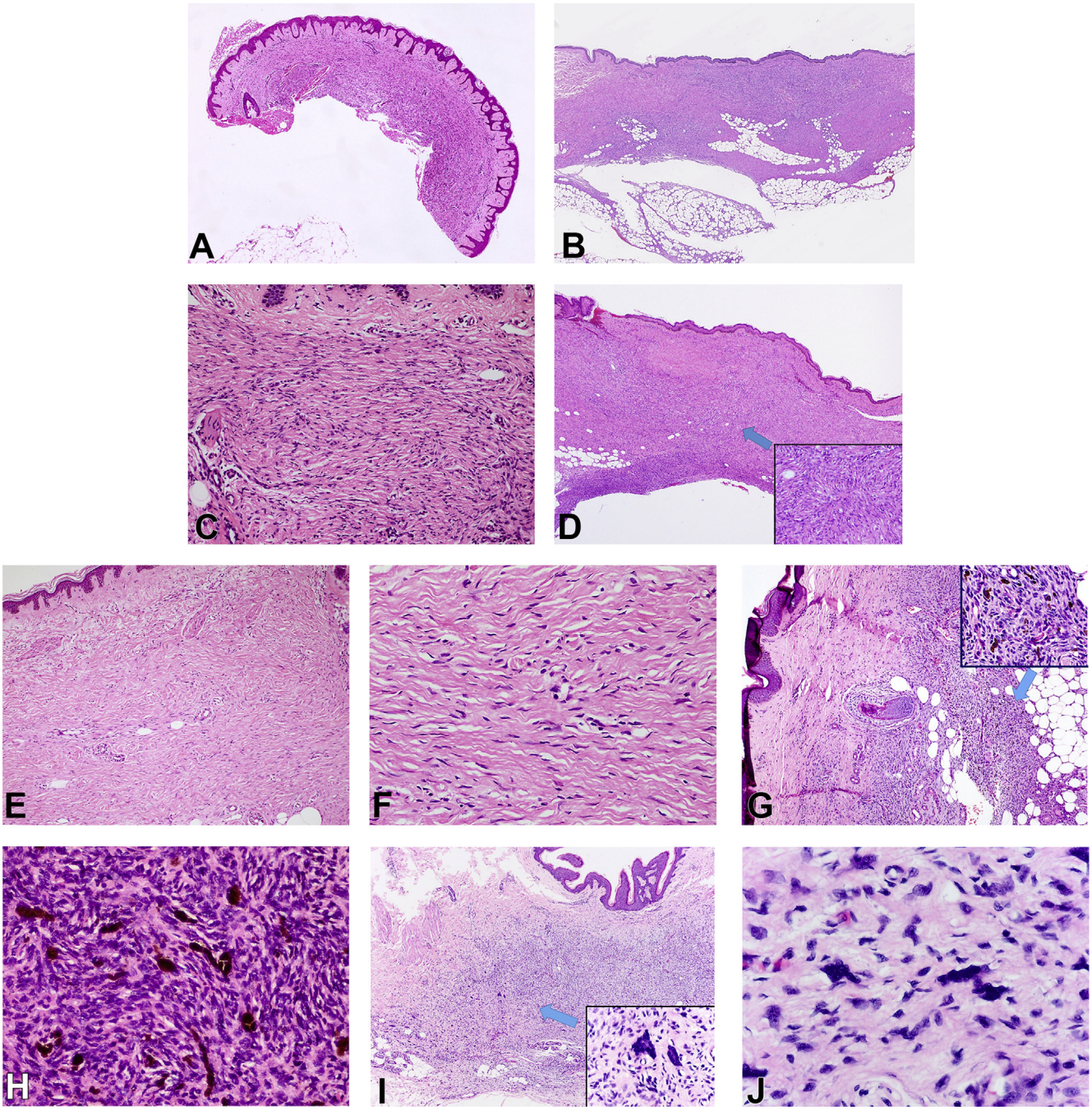

Fig. 2 Histological features of atrophic DFSP. (A) Dermal-based lesion with plaque-like growth pattern. (B) Subcutaneous infiltration. (C) Horizontally oriented fascicles. (D) Focal storiform architecture (inset). (E) Sclerotic appearance at low power. (F) Abundant collagen fibres between tumour cells. (G) Scattered pigmented dendritic cells (inset). (H) Pigmented dendritic cells at high power. (I) Hyperchromatic multinucleated giant cells (inset). (J) Giant cell fibroblastoma-like component.

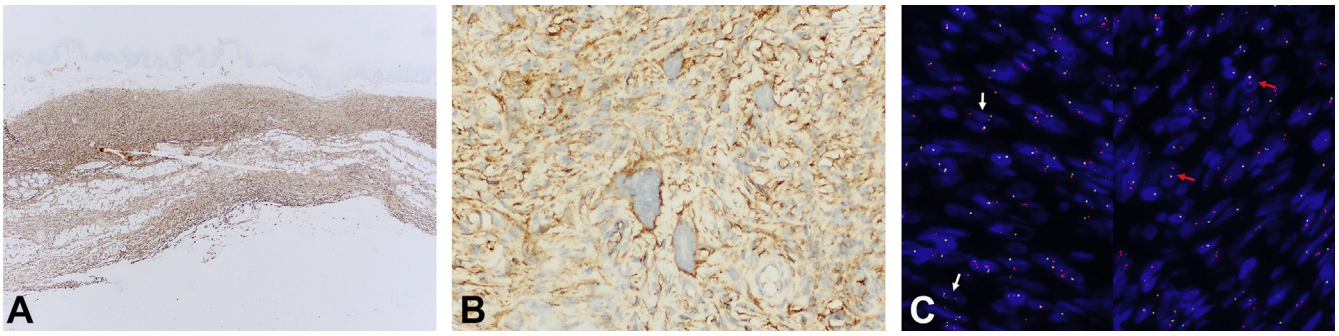

Fig. 3 Immunohistochemistry and FISH of atrophic DFSP. (A) Diffuse and strong expression of CD34. (B) Multinucleated giant cells showing immunoreactivity for CD34. (C) Split of PDGFB gene (white arrows) and loss of telomeric part (red arrows).

aged adults, occurrence of DFSP in childhood and congenital neonates has been increasingly reported,[12–14] indicating a childhood-onset in a proportion of DFSP.

Histologically, classical DFSP often appears as an ill-circumscribed lesion, composed of uniform spindled cells displaying a distinctive storiform arrangement. The tumour cells often infiltrate into the subcutaneous adipose tissue either along the fibrous septa in a lamellar fashion or between individual adipocytes creating a honeycomb pattern. Cytological atypia is minimal and mitotic activity is generally low. Immunohistochemically, DFSP is the prototype of CD34 positive cutaneous spindle cell neoplasms characterised by ubiquitous expression of CD34. The diagnosis of DFSP is usually straightforward. However, unusual clinical presentations of DFSP and morphological variants with absence of classical DFSP areas may pose diagnostic challenges.

Atrophic DFSP represents a rare variant of DFSP. In our own surgical and consultation profiles, atrophic DFSP accounts for only 1.7% (16/937) of all DFSP cases diagnosed within the same period. Including the current series, approximately 42 reports of atrophic DFSP had been documented in the English literature by 2018.[10,11] Differing from the classical DFSP which typically manifests as a protuberant nodule, atrophic DFSP usually appears as a depressed tan-brown, red to grey purplish lesion which is round, ovoid or irregularly shaped and may remain for as long as 20 years.[15] The average size is about 1.5–2.0 cm, but lesions as large as 13×12 cm have been reported.[16] Clinically, the lesions may be diagnosed as a variety of skin diseases, including morphoea, atrophic scar, idiopathic atrophoderma, localised scleroderma, lipoatrophy, atrophic dermatofibroma, resolving panniculitis, or hyperpigmentation.[3,4,7] Of note, an atrophic form of DFSP is rarely considered by clinicians.[17] Like classical DFSP, atrophic DFSP occurs predominantly in young adults with a mean and median age of around 30 years. Childhood and congenital occurrence have also been reported.[12] In contrast to classical DFSP which has a slight male predominance, atrophic DFSP shows a preference for females with a F:M ratio of 2:1. The majority of atrophic DFSP are located on the trunk, accounting for nearly 75% of all cases. Other sites such as the proximal extremities and the head and neck region can also be involved but are comparatively uncommon.

On histological examination, atrophic DFSP manifests as a dermal-based lesion showing a typical plaque-like growth pattern. In our experience, the pathognomonic storiform pattern noted in a classical DFSP is not the striking feature in an atrophic DFSP, although it could be present focally or in some cases. Instead, neoplastic spindle cells arranged in parallel or horizontally oriented fascicles are most commonly seen in atrophic DFSP. Besides the conventional morphological type, pigmented dendritic cells can also be noted in atrophic DFSP, as illustrated in the current series as well as a few cases reported in the literature.[10,18,19] Clinically, pigmented atrophic DFSP may present as a bluish plaque and be interpreted as hyperpigmentation.[18] Of interest, we describe one case of sclerosing atrophic DFSP and one case of hybrid atrophic DFSP/GCF to further expand the morphological spectrum of atrophic DFSP. The latter two variants may be occasionally seen in classical DFSP,[20,21] but are exceedingly rare in atrophic DFSP.

The differential diagnosis of atrophic DFSP embraces a wide variety of cutaneous spindle cell neoplasms. Because of parallel fascicular arrangement and elongated wavy appearance of nuclei, it is not uncommon to misdiagnose atrophic DFSP as a neurofibroma, particularly in the absence of storiform architecture. In challenging cases, immunohistochemical study with application of S-100 protein and SOX10 is helpful in the distinction. The other histological mimics of atrophic DFSP include atrophic dermatofibroma, plaque-like CD34 positive dermal fibroma (‘medallion-like dermal dendrocyte hamartoma’) and dermatomyofibroma. Atrophic dermatofibroma is a rare variant of dermatofibroma which likely represents the end stage of dermatofibroma.[22] Clinically, the lesion appears as an indented erythematous plaque. Histologically, it is characterised by proliferative fibroblast-like spindle cells and histiocytes with epidermal acanthosis and dermal sclerosis. The fibroblast-like spindle cells are generally CD34 negative. Dermatomyofibroma is also a plaque-like lesion composed of uniform, horizontally oriented, fibroblasts/myofibroblasts admixed within collagen fibres.[23] Dermatomyofibroma is variably positive for actins but negative for CD34.

Plaque-like CD34 positive dermal fibroma presents as an indurated plaque on the neck, trunk and extremities.[24] Histologically, it shows a dense, band-like proliferation of fusiform fibroblasts in the papillary and upper reticular dermis. The presence of a storiform pattern together with CD34 immunoreactivity may cause diagnostic confusion with DFSP. In this setting, molecular assays help in distinguishing atrophic DFSP from plaque-like CD34 positive dermal fibroma. Finally, the sclerosing variant of DFSP has to be distinguished from sclerosing fibroma, a benign tumour harbouring overlapping features and

immunophenotypes with sclerosing DFSP.[25] In contrast to sclerosing DFSP which is ill-demarcated, sclerosing fibroma manifests as a dome-shaped circumscribed nodule with no infiltration of subcutaneous adipose tissue. Nevertheless, in biopsy specimens an accurate diagnosis is dependent on molecular analysis.

At a molecular level, the majority of DFSP (>95%) harbour *COL1A1-PDGFB* fusion gene derived from the unbalanced t(17;22) or r(17;22). This fusion gene has been considered DFSP-specific which can be detected by reverse transcription-polymerase chain reaction (RT-PCR) or FISH using formalin fixed, paraffin embedded tissues.[26,27] In routine practice, histology and immunohistochemistry are reliable assays in the diagnosis of DFSP. Molecular analysis is not necessary in most scenarios. However, about 5% of diagnostically difficult cases need to be further confirmed by either FISH or RT-PCR analysis, including atrophic DFSP.[28,29]

Like classical DFSP, a standard wide excision with lateral safety margins is the optional treatment for patients with atrophic DFSP.[30] Mohs micrographic surgery is considered to be an alternative choice for some patients.[31] Adjunct chemotherapy and radiotherapy are not commonly applied in clinical practice. Target therapy with imatinib or sunitinib are recommended in patients with inoperable primary or recurrent tumours or metastatic disease.[32] Preoperative reduction of tumour size with targeted therapy can also be applied in accordance with specific conditions. However, it should be kept in mind that patients may develop primary imatinib resistance.

In summary, we present a series of atrophic DFSP, a rare variant of DFSP, which often poses a diagnostic challenge to both clinicians and pathologists. We also expand the morphological spectrum of atrophic DFSP by further describing a sclerosing subtype and a hybrid DFSP/GCF in addition to the pigmented variant. It needs to be emphasised that the lack of storiform architecture in some cases of atrophic DFSP (especially with sclerotic stroma) may cause diagnostic pitfalls. In challenging cases, FISH assay may help arrive at the correct diagnosis. Our study demonstrates a low rate of local recurrence if completely excised with clear margins.

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