

## Case Report

# Dermatofibrosarcoma Protuberans presenting as an atrophic patch. A case report and brief review of literature

## 一宗呈現為萎縮性斑的隆突性皮膚纖維肉瘤病例報告和簡要文獻回顧

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**Background:** Dermatofibrosarcoma Protuberans is a fibroblastic skin tumour which tends to have locally aggressive behaviour. It usually presents as a slow-growing red-blue nodule. Presentation as an atrophic patch in adults is quite uncommon. **Case Presentation:** A 30-year-old female presented with a 5-month history of an atrophic patch on the right shoulder. Dermatofibrosarcoma Protuberans was confirmed with routine and immunohistochemical staining. The patient was treated with wide local excision. **Conclusion:** Although Dermatofibrosarcoma Protuberans is an indolent slow-growing tumor, considering its local subclinical deep extension, initial wide local excision with safe margin is highly recommended.

**背景：**隆突性皮膚纖維肉瘤是一種成纖維細胞皮膚腫瘤，傾向有局部侵襲性的表現。它通常表現為生長緩慢的紅藍色結節，在成人中表現為萎縮性斑塊實屬罕見。**病例報告：**一名30歲的女性患者，其右肩有著為時五個月的萎縮性斑塊，通過常規和免疫組織化學染色確診為隆突性皮膚纖維肉瘤，患者其後接受了廣泛性局部切除手術。**結論：**儘管隆突性皮膚纖維肉瘤是一種低惡性緩慢生長的腫瘤，考慮到其局部亞臨床深度延伸，故強烈建議進行包含安全邊緣的初始廣泛性局部切除。

**Keywords:** Dermatofibrosarcoma Protuberans, sarcoma, skin neoplasm

**關鍵詞：**隆突性皮膚纖維肉瘤、肉瘤、皮膚腫瘤

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## Introduction

Dermatofibrosarcoma Protuberans (DFSP) is a fibroblastic skin tumour which tends to have locally aggressive behaviour, but rarely metastasises to distant sites. It usually presents as a slow-growing red-blue nodule which ultimately enters a more rapid growth phase. It favours young to middle-aged adults and occurs more frequently on the trunk and accounts for almost 0.1% of all

malignancies with an increasing annual incidence over the past 40 years.<sup>1</sup> Herein, we report a case of DFSP with a rare presentation as an atrophic patch on the shoulder of a young adult female.

## Case report

A 30-year-old female was referred to our clinic with a 5-month history of an asymptomatic atrophic patch measuring 2x1 cm on the right supra-clavicular area (Figure 1). She had no history of previous trauma or injection at this site. Her past medical, drug and family histories were unremarkable. There was no significant clinical finding in the physical examination. An elliptical incisional biopsy was done which revealed an atrophic epidermis with diffuse spindle cell infiltrations in the dermis with nuclear pleomorphism and short fascicular growth pattern extending deep into the subcutaneous tissue. Arrector pili muscles and nerve trunks were entrapped within the lesion. On immunohistochemical (IHC) analysis, there was diffuse strong staining with CD34 in the spindle cells and very weak staining with CD68 and CD99 markers. Staining for S100, smooth muscle actin (SMA) and epithelial membrane antigen (EMA) were negative (Figure 2). After establishing the diagnosis of DFSP, the patient underwent total excision. However, in the second sample; the tumour had extended deep into the subcutaneous tissue, close to the deep margin of the excised tissue. Wide local excision with 3 cm margin was done to ensure total removal of the tumour. There was no recurrence at 6-month follow-up.

## Discussion

DFSP is a locally aggressive fibroblastic tumour which has a low risk for distant metastasis. It occurs most commonly among adults in their fourth to sixth decades of life. There is little difference in incidence rates between either of sexes. The most common site of occurrence is trunk (42%) followed

by upper (23%) and lower extremities (18%) respectively. The annual incidence of the tumour is about four per million and is relatively higher in the black population. In 99.6% of cases, the disease remains localised (57%) or invades adjacent organs (less than 43%), while the distant metastasis is seen only in 0.4% of patients. Five-year and 15-year survival rates for DFSP are 99.2% and 97.2%, respectively. Approximately 1% of tumours present as pigmented variants, namely, the Bednar tumour.<sup>1,2</sup> Molecular studies have shown chromosomal alteration in 90% of cases which involves 7q22; 22q13, with fusion of the genes collagen type 1 alpha 1 (COL1A1) and platelet-derived growth factor-B (PDGF $\beta$ ). These findings may provide additional options for the diagnosis and treatment of these patients. Rarely, fibrosarcomatous changes can occur within the tumour which results in higher rate of recurrence and distant metastasis.<sup>3</sup> Less than 40 cases of atrophic DFSP have been found in the literature including our case and it presents commonly on the trunk of young adults,<sup>4</sup> although, childhood<sup>5</sup> and congenital cases have been reported.<sup>6</sup> Despite its distinct clinical appearance, atrophic DFSP has similar epidemiology, clinical behaviour and prognosis compared to typical DFSP.<sup>4</sup>



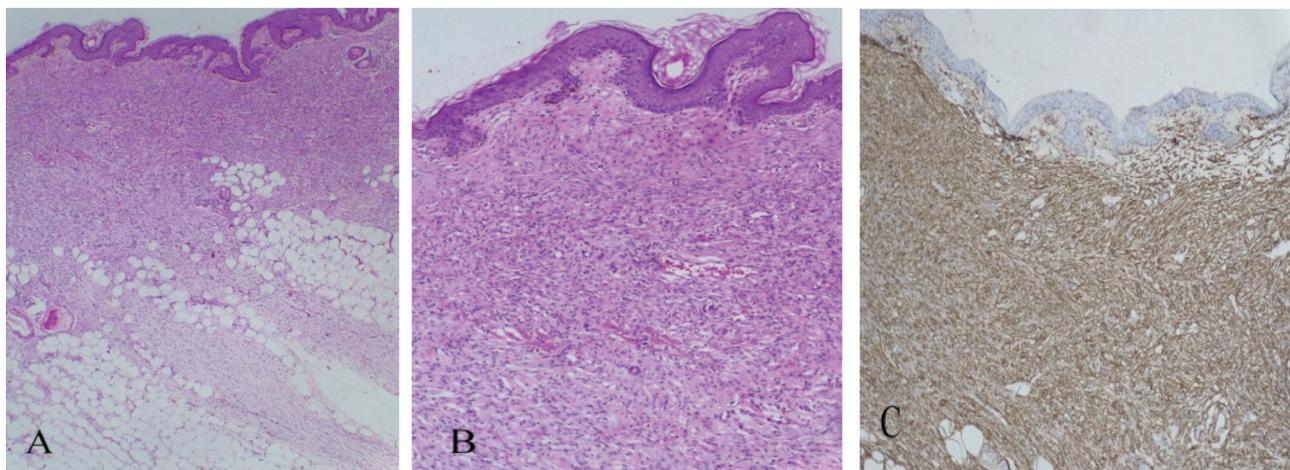
**Figure 1.** Atrophic patch on the right supra-clavicular area (2x1 cm).

DFSP usually presents as an indolent slow-growing skin-coloured asymptomatic nodule or plaque on the upper trunk of a young adult which is usually mistaken for a simple scar, keloid or cyst. Rarely, it may be present as an atrophic patch resembling atrophoderma, morphea or sclerosing basal cell carcinoma. Giant cell fibroblastoma or juvenile DFSP presents earlier than typical DFSP in the first decade and involves the trunk and inguinal areas.<sup>7</sup> A deep incisional biopsy should be taken for histopathological diagnosis. The tumour consists of fascicles of dense spindle cells in a storiform (mat-like) arrangement, extending deep into the dermis and subcutis in a honeycomb pattern. It has a limited mitotic activity (<5 mitoses per 10 high-power field). CD34 staining is usually diffusely positive which differentiates DFSP from a benign dermatofibroma. Cytogenetic studies show a t(17; 22) translocation in more than 90% of cases.<sup>8</sup>

Lymph node and distant spread and metastasis are very rare in DFSP. Therefore, surgical excision is the treatment of choice for localised tumours. Due to subclinical and deep extension of the tumour, complete removal of the tumour during the initial excision is strongly recommended.<sup>9</sup> A

standard resection margin of 3 cm is usually implemented. An additional 1 cm margin should be considered for lesions larger than 5 cm or for recurrent lesions. Narrower resection margins are employed for smaller lesions or for those located in the head and neck to prevent aesthetic and functional impairment.<sup>10</sup> In a recent study, depth of the tumour was the only factor associated with disease-free survival.<sup>11</sup> Before any reconstruction, tumour-free margins have to be confirmed. If surgical margins are not completely clear, tissue rearrangement must be avoided and split-thickness grafting should be considered to monitor the lesion for recurrence.<sup>9</sup>

Imatinib mesylate, a protein tyrosine kinase inhibitor, is clinically active against localised and metastatic DFSP containing t(17:22) translocation. Imatinib has been approved by the FDA for the treatment of unresectable, recurrent or metastatic DFSP in adults.<sup>9</sup> Other tyrosine kinase inhibitors (e.g. sorafenib) have also been reported to be efficacious in unresectable cases.<sup>12</sup> For recurrent tumours, surgical resection is recommended whenever possible. However, for unresectable recurrent tumours, radiotherapy or systemic treatment with imatinib may be considered.



**Figure 2.** (A) (H&E x 10), (B) (H&E x 40) diffuse infiltration of the dermis by spindle cells with nuclear pleomorphism and short fascicular growth pattern extending deeply into the subcutaneous tissue. (C) Strong diffuse CD34 staining of the spindle cells (IHC staining x 10).

Patients should be followed up every 6 to 12 months and re-biopsy of any suspicious lesion is recommended.<sup>9</sup>

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