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Case Report

Dermatofibrosarcoma protuberans: an asymptomatic multinodular plaque mimicking scar tissue

隆凸性皮膚纖維肉瘤：無症狀性疤痕樣多結節性斑塊

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A 61-year-old gentleman presented with a slow growing, nodular scar-like plaque. The diagnosis of dermatofibrosarcoma protuberans was made by histology and confirmed by immunohistochemical study with CD34. Treatment was by excision with a wide surgical margin.

一名六十一歲男士患有一緩慢生長局部擴展的腫瘤。經組織學檢查及免疫組織化學檢驗見分化群集34(CD34)淋巴細胞確診為隆凸性皮膚纖維肉瘤。病損經闊緣外科手術切除。

Keywords: Dermatofibrosarcoma protuberans

關鍵詞：隆凸性皮膚纖維肉瘤

Case report

A 61-year-old retired gentleman presented with an asymptomatic plaque on his left hip for the past four years. This was slowly increasing in size and had become mildly tender. His past health was unremarkable. He sustained an injury to his left hip as a result of a plane accident prior to the onset of this lesion.

On examination, there was a multinodular plaque of four centimetres with an irregular border on his left hip. This was violaceous in colour with bluish red surrounding skin (Figure 1). There was no regional lymphadenopathy or hepato-splenomegaly. The differential diagnosis included keloids, dermatofibrosarcoma protuberans, dermatofibroma, other dermal tumours, foreign body granuloma and mycobacterial infection.

An incisional skin biopsy was carried out and showed a non-circumscribed cellular tumour in the dermis and subcutis in a storiform pattern. The subcutis was largely replaced by the tumour with only few isolated fat cells. Tumour cells were monotonous with mild nuclear atypia. There was

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no definite mitosis or high grade sarcoma (Figure 2). An immunohistochemical study showed that the tumour cells were strongly positive for CD34 (Figure 3). The features were compatible with dermatofibrosarcoma protuberans (DFSP). The tumour was excised with a wide margin.

Discussion

Dermatofibrosarcoma protuberans was first described by Darier and Ferrand in 1924 and named by Hoffman one year later.[1] It is a locally invasive tumour arising in the dermis from

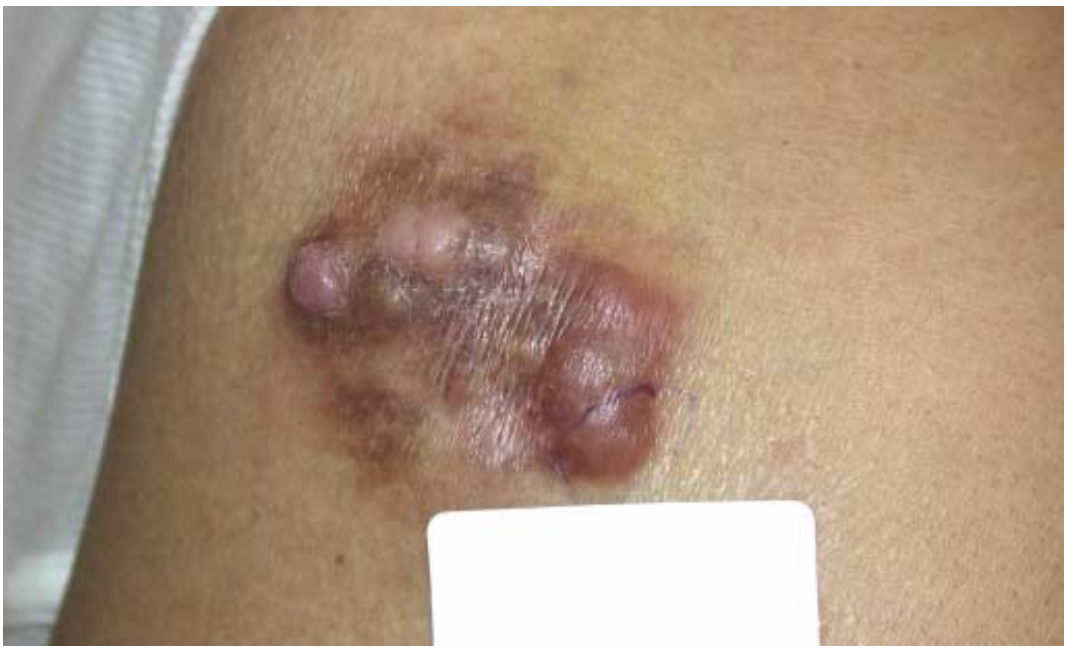

Figure 1. A multinodular scar-like plaque on the left hip.

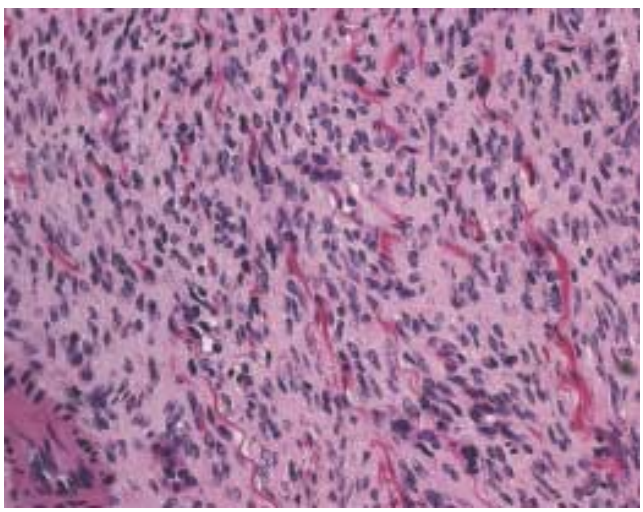

Figure 2. A storiform pattern with monotonous tumour cells and mild nuclear atypia.

Figure 3. Immunohistochemical staining strongly positive for CD34.

fibroblasts. It accounts for 0.1% of all cutaneous malignancies and 1% of all soft tissue sarcoma. The incidence ranges from 0.8-5 cases per million persons per year.[2,3] It is most commonly found in the 20-50 years age group and there is slight male predominance. The tumour is usually found on the trunk and proximal extremities. Precipitating factors including trauma, vaccination site, surgical and burnt scar, and arsenic exposure were reported. Pregnancy causes an accelerated growth of this tumour and is believed to be caused by an elevated level of platelet derived growth factor (PDGF) and the increased levels of progesterone through binding to progesterone receptors.

At the cytogenetic level, reciprocal translocation, t(17;22)(q22;q13) or supernumerary ring chromosome from translocation r(17;22) are found in 92% of all cases. As a result, there is a fusion of the two genes, collagen type 1 alpha 1 (COL1A1) and platelet derived growth factor B chain (PDGFB), related to the formation of type 1 collagen and platelet derived growth factor respectively. The breakpoint localisation in PDGFB was found to be remarkably constant, placing exon 2 under the control of the COL1A1 promotor. In contrast, the COL1A1 breakpoint was found to be variably located within exons of the alpha-helical coding region (exons 6-49). It has been proposed that PDGFB acts as a mitogen in DFSP cells by autocrine stimulation of the PDGF receptor.

Clinically, DFSP usually presents as a solitary or multiple nodules arising in an indurated plaque with an irregular outline. It ranges from 0.5 to 10 centimetres in diameter and is brown or bluish red in colour. There is a blue or reddish discoloration of the surrounding skin. The lesion is asymptomatic in the early stage but patient may experience pain or tenderness later. There is a variant known as Bednar tumour. This is usually found in Black people and is characterised by the presence of melanin. It accounts for up to 5% of all cases.

The histopathology of this tumour is divided into the plaque and nodular stage. The plaque stage has a flat surface with low cellularity and lack of storiform pattern. The slender spindle shaped neoplastic cells are lying parallel to the skin surface and is non refractile to the polarised light. The nuclei are uniform and mitotic figures are rare. In contrast, the nodular stage showed a thin epidermis with high cellularity and a storiform pattern. Hyperchromatic nuclei are seen and mitosis ranges from 3-5 per 10 high power field. It infiltrates the subcutaneous tissue in a honeycomb and later a solid pattern. There is an absence of adnexal structure. An immunohitochemical study showed strong positivity for CD34 in the plaque areas but only weak to moderately positive for the nodular areas. This is useful to differentiate DFSP from dermatofibroma which is CD34 negative but factor XIIIa positive. CD34 is also useful to evaluate the infiltrative margins for surgical excision. The histological differential diagnosis in the early stage includes scar tissue and morphoea; in the intermediate stage, dermatofibroma; and in the advanced stage, malignant fibrous histiocytoma and fibrosarcoma.

Treatment of DFSP is by surgical excision. The current recommendation is three centimetres or greater margin down to and including fascia, irrespective of the tumour size. Prophylactic lymph node dissection is not warranted. DFSP has a strong propensity to recur even after wide excision. Taylor and Helwig reported a 49% recurrence rate in 98 cases;[4] Pack and Tabah, a 24% recurrence rate in 17 patients;[5] and Hajdu, a 54% recurrence rate in 119 cases.[6] Roses reported a 41% recurrence rate with clinical margins less than two centimetres and a 20% recurrence rate with clinical margins of at least 3 centimetres.[7] McPeak and Cruz reported a 11% recurrence rate in 27 patients

treated with a 3-centimetre margin that was taken beyond clinically involved skin down to and including fascia.[8] A similar result was obtained by Haycox et al.[9] Recurrences usually occur within three years. In Taylor and Helwig's series, 40% of recurrences were found in the first year, and 75% within the first three years; in Hajdu's series, most recurrences were noted in the first year; in McPeak and Cruz's series, 80% of recurrences were noted within the first three years.

Mohs micrographic surgery (MMS) has been used to treat DFSP with great success. A review of 221 patients showed an overall recurrence rate of 2.3%.[10] Garcia et al reported no local recurrences of 16 cases.[11] Parker and Zitelli treated 20 patients with MMS and had zero local recurrences.[12] Ratner et al series had a recurrence rate of 2%.[13] Dawes and Hanke reported a recurrence rate of 8% in the 24 cases of DFSP treated with MMS.[14]

Radiotherapy had been used as an adjuvant to surgery for a big tumour, a tumour with unclear margin after excision or recurrence after surgery. It is also used for treating an unresectable tumour. There is a theoretical risk of fibrosarcomatous change. PDGF receptor tyrosine kinase inhibitor had been tried for DFSP. This drug was initially used to treat chronic myeloid leukaemia. It works by induction of apoptosis and has no effect on angiogenesis. There is an overall tissue viability. No major side effects have been reported.

The progression of DFSP to malignant histiocytoma and fibrosarcoma has been reported. Haemato-genous spread is rare and if this occurs, it is usually to the lungs and regional lymph nodes.

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