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Case Report

An asymptomatic plaque over right chest wall of a Chinese man

男性患者右胸壁無症狀性斑塊

AYK Chan 陳綺琪 and KK Jong 莊國坤

A 53-year-old Chinese man had an asymptomatic mass over his right chest wall. Histopathological examination confirmed dermatofibrosarcoma protuberans.

53 歲男性患者於右胸壁出現無症狀性腫塊。組織病理學檢查證實為隆突性皮膚纖維肉瘤。

Keywords: Dermatofibrosarcoma protuberans

關鍵詞：隆突性皮膚纖維肉瘤

Introduction

Dermatofibrosarcoma protuberans is a low grade sarcoma of uncommon occurrence.[1, 2] It is locally invasive but rarely metastasizes.[1-3] We report a 53-year-old Chinese man with dermatofibrosarcoma protuberans manifesting as an asymptomatic plaque over his right chest wall.

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Case report

A 53-year-old Chinese man had an asymptomatic skin lesion on his right chest wall for two years. The growth had an insidious onset and was almost unnoticed by the patient. However, a significant increase in size was found over the last six months and it became nodular. He had no systemic complaints. He recalled no past history of trauma. Concerning his past medical history, he had attended the chest clinic for shadow on chest X-ray but repeat chest radiography was found to be normal.

Physical examination revealed a 1 x 1 cm erythematous nodule with surrounding ill defined erythematous plaque measuring 4 x 2 cm on his right chest wall. It was firm in consistency with telangiectasia over surface. No ulceration was noticed (Figure 1). There was no lymphadenopathy. Incisional biopsy showed an

ill-defined tumour mass composed of densely packed, monomorphous, plump spindle cells arranged in a storiform pattern in the dermis with diffuse infiltration of the tumour cells into the subcutis (Figures 2 & 3). Immunohistochemical studies showed a positive result for CD34 (Figure 4) and negative results for LCA, desmin, S100 and actin. It was consistent with dermatofibrosarcoma protuberans. He was referred to a surgical unit for complete excision. Unfortunately, the excisional margin was not clear and further wider excision was planned.

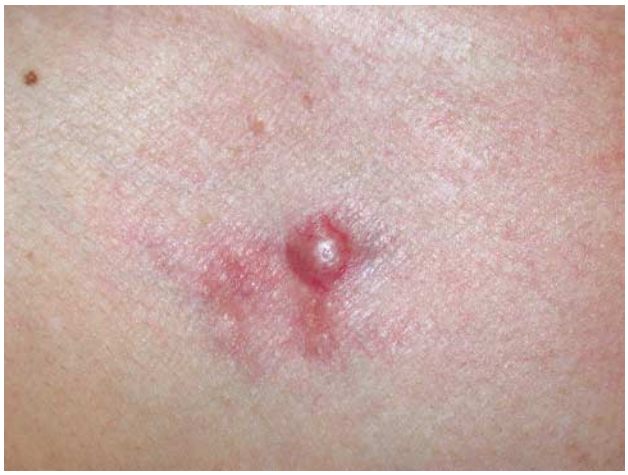

Figure 1. This clinical picture showed a 1 x 1 cm erythematous nodule with surrounding ill defined erythematous plaque measuring 4 x 2 cm on the right chest wall.

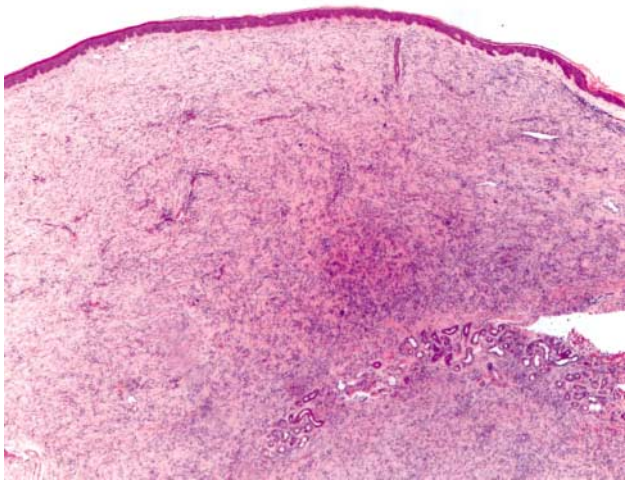

Figure 2. This incisional biopsy showed an ill-defined tumour mass arranged in storiform pattern in the dermis. (H&E, original magnification x 10)

Discussion

Dermatofibrosarcoma protuberans is an uncommon low grade sarcoma which is locally invasive but rarely metastasizes.[1-3] Its line of differentiation is still debatable, and a fibroblastic, histiocytic, neural or perineural origin have been suggested.[3] An 30-year long epidemiology study from the United States showed that black women at early adulthood to middle age may be more susceptible. It rarely occurs in childhood.[2] It was found that more than 90% of the cases showed undue chromosomal translocation between

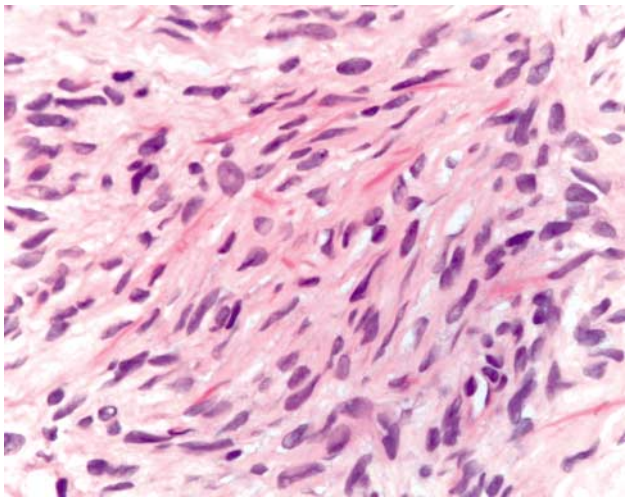

Figure 3. The proliferation was formed of densely packed, monomorphous, plump spindle cells arranged in a storiform pattern. (H&E, original magnification x 400)

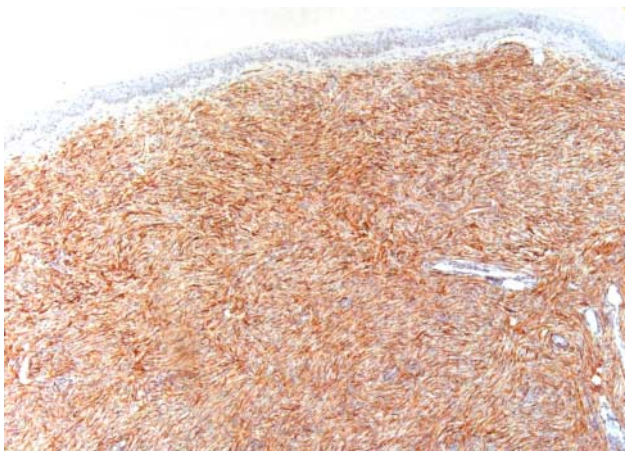

Figure 4. Immunohistochemical study showed positive result for CD34.

17 and 22 with fusion between collagen type Iα1 gene (COLIAI) and platelet derived growth factor (PDGF) β-chain gene, which led to deregulation of the wild type PDGF β-chain expression and uncontrolled receptor activation and cell growth.[1-3]

Our patient presented with an erythematous nodule with a surrounding ill defined erythematous plaque over the right chest wall. Based on the morphology of the lesion, several benign or malignant tumours and infective causes can mimic this condition. Malignant tumours include cutaneous lymphoma, nodular type of basal cell carcinoma, atypical fibroxanthoma and cutaneous metastases. Benign tumours include keloid, haemangioma, lymphocytoma cutis and dermatofibroma. Infective causes include cutaneous mycobacterial or atypical mycobacterial infection.

Dermatofibrosarcoma protuberans usually presents as an asymptomatic erythematous plaque which is initially ignored due to its insidious nature but eventually it gets noticed as it increases in size and becomes nodular. It is firm in consistency and the surrounding skin may be telangiectatic.[1] The most prevalent sites are the trunk and shoulder followed by the extremities, then the head and neck region.[1,4] The average time to diagnosis is 5 to 10 years which echoes the usual insidious and slow growing nature of the lesion. Several morphologic variants have been described such as nonprotuberant, protuberant, pigmented, myxoid and fibrosarcomatous variants.[1,4] For investigations, a skin biopsy is performed for histological confirmation. Chest radiography has been suggested as a screening test for any lung metastasis. MRI is useful in preoperative assessment to delineate the size and extent of the tumour and can be used to monitor tumour recurrence.[5]

Surgical excision is the mainstay of treatment and the key to success is clear surgical margins. Wide local excision with at least 2-3cm margin is advocated.[1,5] Where there is expertise available, Mohs' micrographic surgery may be a more supreme option. Radiotherapy has been suggested in a limited series as an adjuvant for case with positive surgical margins, or as an exclusive treatment in advanced cases with no feasible surgical approach.[1,5]

The prognosis is usually good with 5-year survival rates reaching 99% but there is a high rate of local recurrence due to its infiltrative nature.[1,5] Mohs' micrographic surgery can significantly reduce the local recurrence down to 1.6% as compared with simple excision which had a local recurrence rate of up to 60%.[6] Metastasis is uncommon and occurs mainly in those patients with risk factors including multiple lesions, recurrent tumour, advanced age, histological features of high mitotic index, increased cellularity and fibrosarcomatous change.[5] The lung is the most common site of metastasis.[1]

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