

## Case Report

# A young lady with facial asymmetry: a case of lupus profundus

## 一名年輕女士的面部不對稱：深在性紅斑狼瘡一例

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A 26-year-old lady presented with nine years' history of facial indurated nodule and subsequent facial tenderness, swelling and asymmetry. The histology revealed dense lymphoid aggregates with germinal centre formation in the subcutis associated with septal fibrosis, and hyaline type fat necrosis characteristic of lupus profundus.

一名二十六歲女士因九年前開始發覺面部有皮下硬結，其患處繼而出現腫脹，觸痛感及不對稱現象而求診。皮膚切片顯示病灶皮下組織有著密集的淋巴細胞聚集並形成生長中心，更兼有間隔纖維化和透明式脂肪壞死等深在性紅斑狼瘡的相關特徵。

**Keywords:** Lupus panniculitis, lupus profundus

關鍵詞：狼瘡性脂膜炎、深在性紅斑狼瘡

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## Introduction

Lupus profundus, also known as lupus panniculitis, is a subtype of chronic cutaneous lupus erythematosus. It is characterised by a female

predominance and the median age of onset is 30-40 years. Patients present with tender, subcutaneous nodules and plaques especially over the face, upper arms, upper trunk, breasts, buttocks and thighs. We report a young Chinese lady with lupus profundus who presented with facial asymmetry.

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## Case history

A 26-year-old lady was referred to the Social Hygiene Service, Centre for Health Protection, HKSAR in May 2011 for suspected lipoatrophy of the left cheek. The history could be dated back to 2002 when she presented to our service with a tender indurated nodule on the left cheek. However, she defaulted follow-up afterwards and

no formal diagnosis was made. She was seen by a private surgeon in 2010 and an incisional biopsy on the nodule was performed. She was told by the surgeon that she was suffering from panniculitis and lipoatrophy. In the following year, she noted that there was continuous "shrinkage" of the left face and she also developed pain, tenderness and swelling over the right face.

Physical examination showed atrophy on the left side and a tender swelling on the right side of the face (Figure 1). There were no epidermal skin changes, follicular plugging, oral ulcer or photosensitivity. The differential diagnosis included lupus profundus, acquired partial lipodystrophy, familial partial lipodystrophy, HIV/HAART-associated lipodystrophy syndrome and deep morphea.

Initial investigations which included blood tests for haemoglobin, white cell count, platelet count, liver and renal function, thyroid function, C3, C4, rheumatoid factor were all normal. HIV-antibody was negative. The titre of ANA was 1:80. However, anti-DNA and anti-ENA were negative.

The histological slides of a skin biopsy performed by a private surgeon over the patient's left face were retrieved for review. Direct immunofluorescence was not performed at that time. The skin biopsy revealed an unremarkable epidermis and dense lymphoid aggregates with germinal centre formation in the subcutis associated with septal fibrosis (Figure 2). Hyaline type fat necrosis characteristic of lupus profundus was demonstrated but features of lipoatrophy (involutional or inflammatory type) were absent (Figure 3).

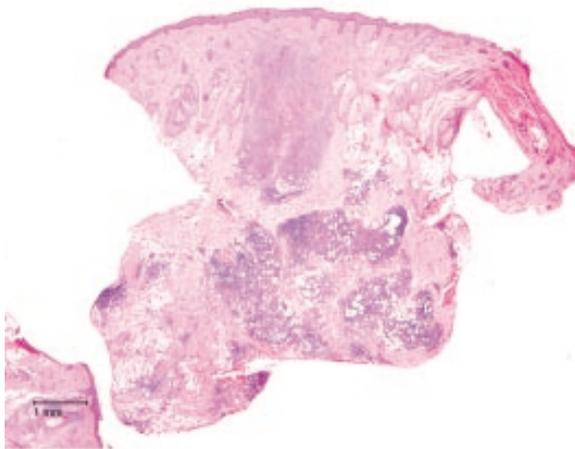
The clinical and histological findings were compatible with the diagnosis of lupus profundus. The patient was started on hydroxychloroquine 400 mg daily and naproxen 250 mg three times daily. She was also referred to the plastic surgeon for consideration of autologous fat transplantation.

## Discussion

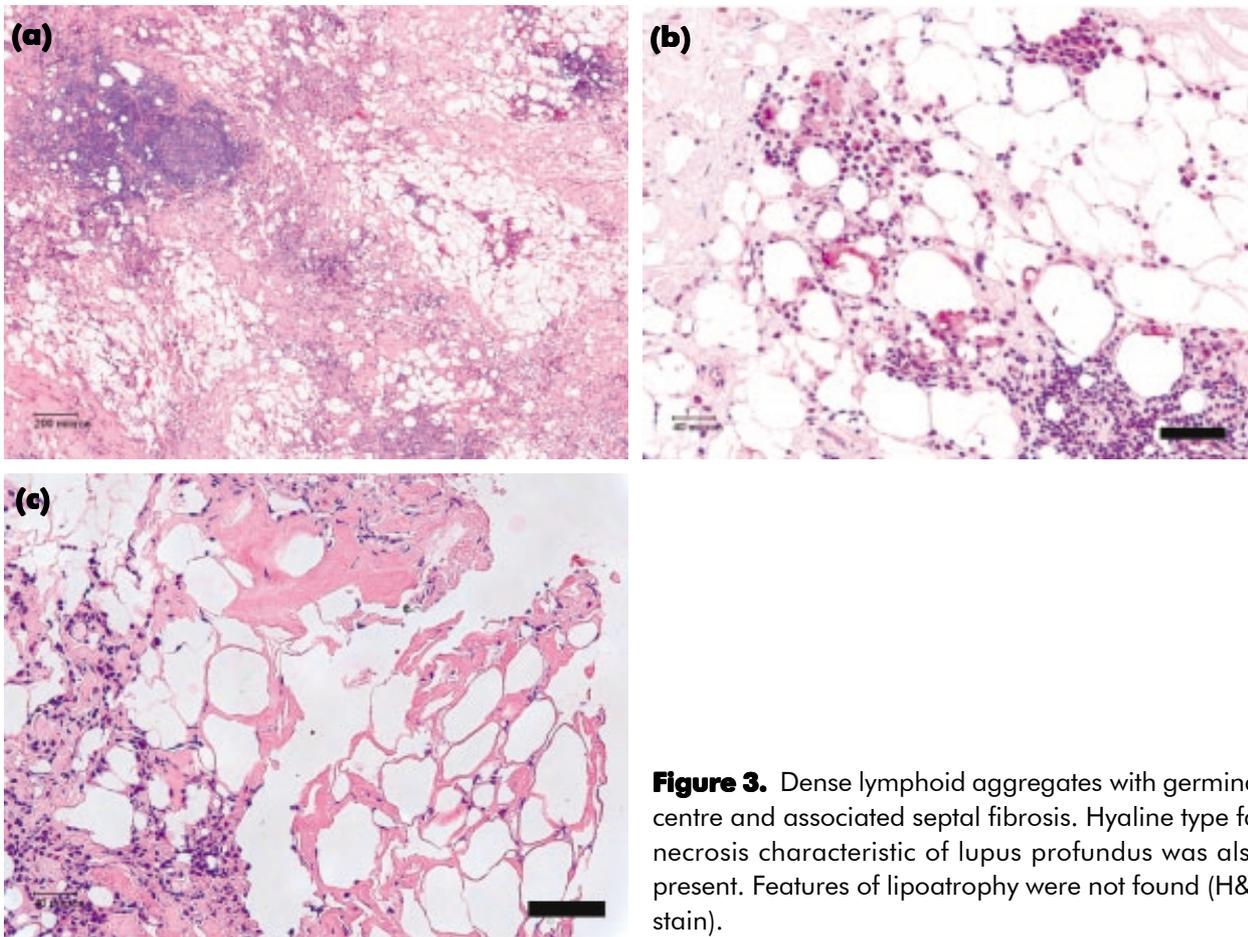
Lupus profundus (lupus panniculitis) is a subtype of chronic cutaneous lupus erythematosus (CCLE). It is associated with CCLE and systemic lupus erythematosus (SLE) in at least one third and 10-15% of the cases respectively. This condition



**Figure 1.** Facial asymmetry with left cheek atrophy.



**Figure 2.** Low power view: Dense lymphoid aggregates with fibrosis in the subcutis. The epidermis is relatively unremarkable (H&E stain).



**Figure 3.** Dense lymphoid aggregates with germinal centre and associated septal fibrosis. Hyaline type fat necrosis characteristic of lupus profundus was also present. Features of lipoatrophy were not found (H&E stain).

constitutes 2-3% of all cases of lupus erythematosus (LE).<sup>1</sup> It usually occurs in adults with a median age of onset of 30-40 years with a female predominance.<sup>2</sup>

Clinically, patients present with tender, subcutaneous nodules and plaques occurring predominantly over the face, upper arms, upper trunk, breasts, buttocks and thighs. Lupus profundus runs a chronic, relapsing course and the inflammation in the fat leads to indurated plaques which can evolve into disfiguring, depressed areas or lipoatrophy. Changes in the overlying skin range from a light pink colour to an erythematous plaque with scarring and follicular plugging as seen in discoid lupus erythematosus (DLE). The overlying skin may be "tethered" to the subcutaneous lesion, causing a surface depression and ulceration.<sup>3</sup>

Histologically, lupus profundus is characterised by the following features: predominantly lobular panniculitis, characteristic hyaline fat necrosis and lymphoplasmacytic infiltrates. Lymphocytic vasculitis and mucin deposition may be present and direct immunofluorescence may be positive. Epidermal or dermal changes of CLE may be present in one-half to two-thirds of cases.

First-line treatment of lupus profundus are antimalarials and systemic or intralesional corticosteroids (level of evidence: E).<sup>4</sup> Antimalarials produce improvement in most patients but treatment may be required for several years.<sup>2</sup> Second-line treatment is topical corticosteroids under occlusion (level of evidence: E).<sup>4</sup> Third-line therapies include gold, bismuth, dapsone, cyclophosphamide, thalidomide, rituximab and cyclosporine A (level of evidence: E).<sup>4</sup>

In conclusion, lupus profundus is an inflammatory disorder of the deep dermis and subcutaneous fat and it is a variant of CLE with female predominance. Disfigurement and lipoatrophy may result from lupus profundus. So, early diagnosis and investigations can prevent permanent disfigurement and the ensuing psychological consequences. When we diagnose a patient with lupus profundus, we should also screen for the presence of other LE features and systemic involvement with which they may be associated.

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