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Case Report

A lady with indurated plaques over both eyebrows

一位在雙眉位置有著硬化斑塊的女仕

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A 47-year-old lady presented with erythematous plaques over bilateral eyebrows and nose regions for about 10 years. She also had a history of systemic lupus erythematosus, idiopathic thrombocytopaenic purpura, interstitial lung disease and chronic hepatitis B. The diagnosis was confirmed to be basaloid follicular hamartoma by histological examination. The patient was treated with potent topical steriods with a satisfactory clinical response.

一名四十七歲的女仕在兩邊眉毛及鼻子位置長出紅色的斑塊，爲時約莫十年。她本身有系統性紅斑狼瘡、特發性血小板減少性紫癜、間質性肺病和慢性乙型肝炎的病史。組織學檢查後，診斷確認爲基底細胞樣毛囊錯構瘤。該患者在使用強效外用類固醇治療後，得到滿意的臨床效果。

Keywords: Basaloid follicular hamartoma, systemic lupus erythematosus

關鍵詞：基底細胞樣毛囊錯構瘤、系統性紅斑狼瘡

Introduction

Basaloid follicular hamartoma (BFH) is a benign lesion of important consideration as it can be mistaken both clinically and histologically for basal cell carcinoma. We present a case report of a middle-aged lady with slowly progressive plaques over both eyebrows and nose for 10 years. A skin biopsy performed over the right eyebrow revealed the characteristic features of basaloid follicular hamartoma.

Case history

A 47-year-old lady with underlying systemic lupus erythematosus (SLE) diagnosed since 1987, idiopathic thrombocytopaenic purpura, interstitial lung disease, chronic hepatitis B, fatty liver and depression was referred from the medical department of a local regional hospital for erythema over the glabellar region for ten years. Medications she was taking included prednisolone and azathioprine.

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It was noted that she had a rash over both eyebrows and nose for ten years which was occasionally itchy and slowly progressive. It was not related to sunlight and there was no previous history of trauma.

Physical examination showed pinkish to reddish indurated plaques over both eyebrows, glabellar and nasal regions (Figure 1), and alopecia over both eyebrows (Figures 2 & 3). Clinical differential diagnoses included Jessner's lymphocytic infiltrate, granuloma faciale, trichoblastoma, sebaceous hyperplasia, leprosy, cutaneous tuberculosis, deep fungal infection and sarcoidosis.

Incisional biopsy, which was performed over the right eyebrow, showed mild hyperkeratosis in the epidermis and thickening of the epidermal basement membrane. The dermis showed fibrosis and contained many abnormal hair follicles with basaloid cell proliferation forming irregular buds, and anastomosing cords with peripheral palisading, distorting the hair follicles. Disorganised hair papillae were also noted. These abnormal follicular structures were surrounded by a slightly myxoid, cellular stroma. There were no clefts between the basaloid cells and surrounding stroma. The histological features were consistent with basaloid follicular hamartoma (Figure 4).

In view of the unique histological findings and clinical features, the diagnosis of basaloid follicular hamartoma was made. Patient was initially given topical tretinoin cream with fair

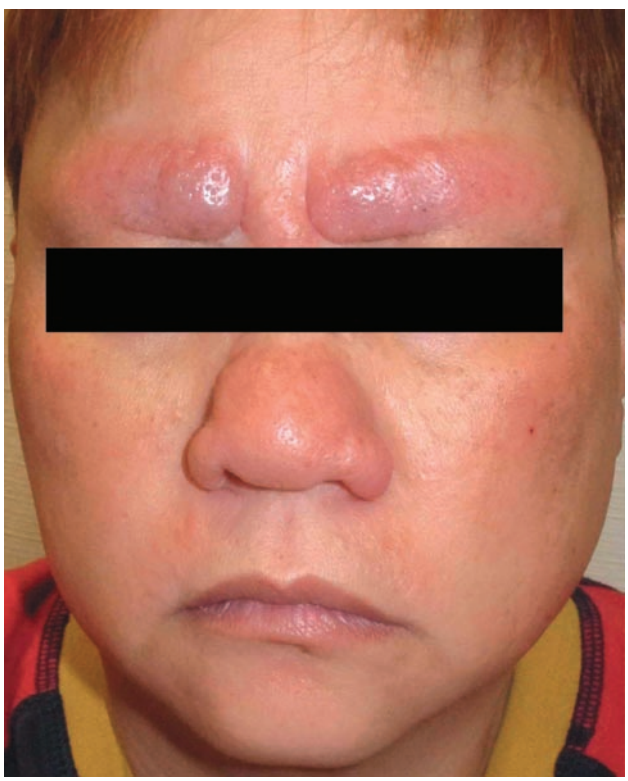

Figure 1. Flesh-coloured plaques over both eyebrows and nose.

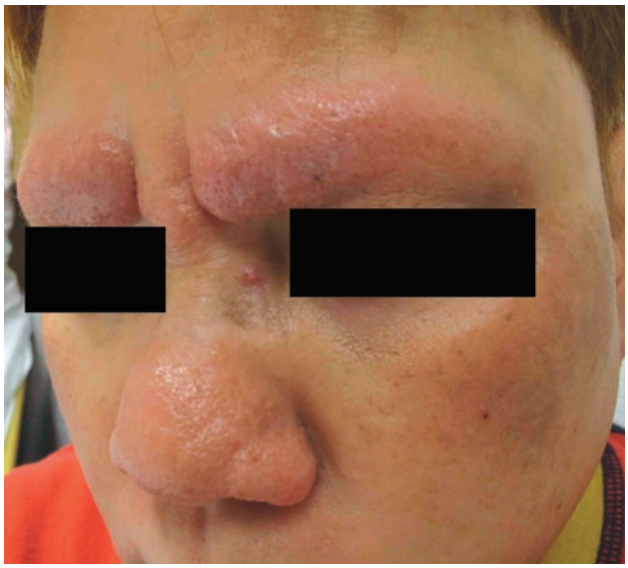

Figure 2. Alopecia over both eyebrows.

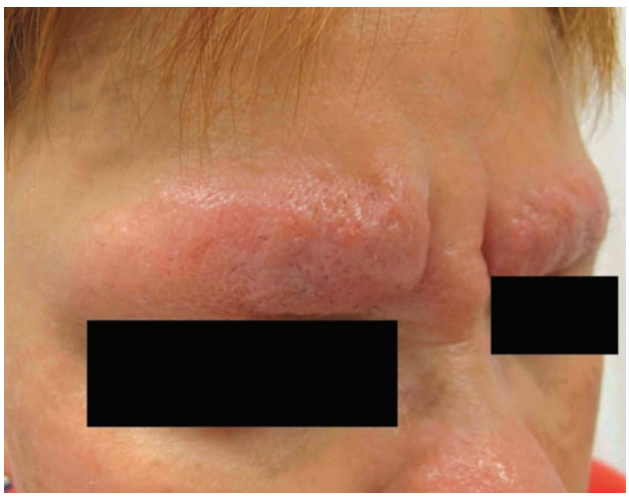

Figure 3. Close-up view of the lesions.

response. Subsequently she was treated with a topical steroid, mometasone furoate cream, daily with mild clinical improvement on subsequent follow-up.

Discussion

BFH was first described in 1969 by Brown et al as multiple papules in the nasolabial folds associated with myasthenia gravis and diffuse alopecia. In 1985, the term "basaloid follicular hamartoma" was used by Mehregan and Baker, who reported localised and solitary lesions without associated abnormalities. BFH is a rare benign adnexal tumour characterised by superficial malformation of hair follicles and distinctive histopathological features. There is a transformation of hair follicles into solid mass and branching cords of basaloid cells.[1]

Patients are usually asymptomatic. The clinical lesion is most often found on the face and scalp. There are variable clinical presentations and associations. It may be localised (solitary), generalised or of linear form.[2]

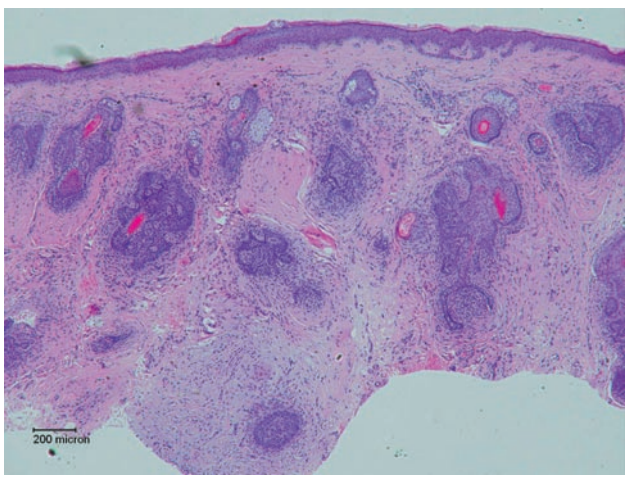

Figure 4. Hair follicles distorted by basaloid cell proliferation. Note the irregular buds and anastomosing cords of basaloid cells with peripheral palisading but no clefts in the surrounding stroma. (H&E, marker: 200 micron).

This condition is generally benign. Patients usually have a stable clinical course. The lesions in patients with multiple basaloid follicular hamartoma may gradually increase in size throughout childhood, but then stabilise and become static upon reaching adulthood. The main complaint from the patient is cosmetic concern. Rarely, development of basal cell carcinoma within lesions has been reported.[3]

The hamartoma is derived from hair follicles and consists of branching cords and strands of basaloid cells showing follicular differentiation. Basaloid proliferations in the lower portions of pre-existing follicles may cause follicular destruction. As a result, malformation of hair follicles, alopecia, and hypotrichosis are often associated with basaloid follicular hamartoma. The pathogenesis is found to be associated with the patched *PTCH* gene mutation on chromosome 9q23. The *PTCH* gene encodes a receptor involved in the Sonic Hedgehog-Patched signalling pathway. Mutations in this signal pathway lead to abnormal growth of cells.[3]

Although there are many clinical forms of basaloid follicular hamartoma, they all share the same unique histopathological feature. Basaloid follicular hamartoma is a folliculocentric lesion limited to the superficial dermis. Deep reticular dermis or soft tissue involvement is not typical for this disease. In early lesions, branching cords of basaloid cells connecting the central pilosebaceous structures can be observed. In longstanding lesions, the central pilosebaceous structures can be totally replaced by the basaloid cell proliferation.[4]

No specific tests are required to establish a diagnosis of basaloid follicular hamartoma other than skin biopsy. However, given the association between multiple BFH and autoimmune diseases such as SLE and myasthenia gravis, a comprehensive history and physical examination should be performed. Skin biopsy is needed to establish the histological diagnosis. Autoimmune disease screening would be needed.[5]

Basaloid follicular hamartoma is a rare condition. There is no standard treatment protocol. Numerous treatment options are available, like topical tretinoin gel, systemic isotretinoin and photodynamic therapy. Systemic isotretinoin has been reported to decrease the size of lesions and improve the associated alopecia in patients with multiple BFH and SLE. As basal cell carcinoma may occur in BFH, biopsy should be performed in the event of sudden enlargement of the lesions. Apart from clinical suspicion of malignant change, there is no absolute medical indication to remove the lesion, although they may be excised for cosmetic reasons.[1,6]

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