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Case Report

Pilomatricoma with a bullous appearance or secondary anetoderma involving a pilomatricoma?

水疱型毛髮基質瘤抑或是毛髮基質瘤伴有繼發性皮膚鬆弛症?

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Pilomatricoma or benign calcifying epithelioma of Malherbe is a benign appendageal tumour originating from primitive cells of the hair matrix. Perforation (transepidermal elimination), bullous or anetodermic cutaneous changes overlying pilomatricomas are rare events. In this report, we describe a case of pilomatricoma with bullous appearance or secondary anetoderma. The tumour was totally excised and the wound healed well with no recurrence.

毛髮基質瘤或稱良性馬勒布氏鈣化上皮瘤是一種衍生自毛髮基質原始細胞的良性皮膚附屬器官腫瘤。 而表面帶有穿孔（經表皮排出）、水疱或皮膚鬆弛變化的毛髮基質瘤則屬罕見病例。我們在本文報告水疱型或繼發性皮膚鬆弛症的毛髮基質瘤一例，該腫瘤在完全切除以後，傷口癒合良好及沒有復發。

Keywords: Anetoderma, bullous appearance, calcifying epithelioma of Malherbe, pilomatricoma

關鍵詞：皮膚鬆弛症、水疱型、馬勒布氏鈣化上皮瘤、毛髮基質瘤

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Introduction

Pilomatricoma or benign calcifying epithelioma of Malherbe is a benign appendageal tumour originating from primitive cells of the hair matrix. It is usually a solitary, firm to hard, deep dermal or subcutaneous tumour that is covered by normal skin. Rarely, perforation (transepidermal elimination), bullous or anetodermic cutaneous changes overlying pilomatricomas have been reported.[1,2]

Case report

A 30-year-old woman had a tumour on the right shoulder that had been slowly growing for one year. It was noted to have a bullous appearance three months prior to consultation. There was no history of trauma. On examination, a single, slightly tender, flaccid bulla, measuring 3 x 2 cm was detected. Within the bulla there was a single hard tumour (Figure 1a). The skin over the bulla could be pushed deep into the dermis, which popped out with release of pressure (Figure 1b). Systemic examination was normal. Laboratory examination of haematological, biochemical parameters and urinalysis were normal. Excision of the tumour was performed and the wound healed well with no recurrence.

Histopathological examination of the lesion demonstrated a normal epidermis and a well demarcated tumour in the dermis. There was a large space in the dermis over the tumour. The tumour was composed of irregular nests of basophilic cells, shadow cells and transitional cells in a fibrotic stroma. Focal areas of calcification were present (Figure 2a). In the overlying dermis, the collagen bundles were attenuated and there were some dilated endothelium-lined vascular channels. The alcian blue stain was negative. Verhoeff van Gieson stain showed a marked reduction in elastic fibres (Figure 2b). The findings were consistent with pilomatricoma.

Discussion

Although pilomatricomas are common tumours and there are multiple large series in the medical literature, bullous appearance or secondary anetoderma overlying pilomatricoma was described in only 24 patients in English literature. An additional 19 cases were reported in French and, as a matter of fact, the first case was reported in French.[1,3-12] The incidence of a bullous apperance was reported as 6.3% in Japan.[6]

The majority of the patients with bullous apperance of pilomatricomas were young women in the

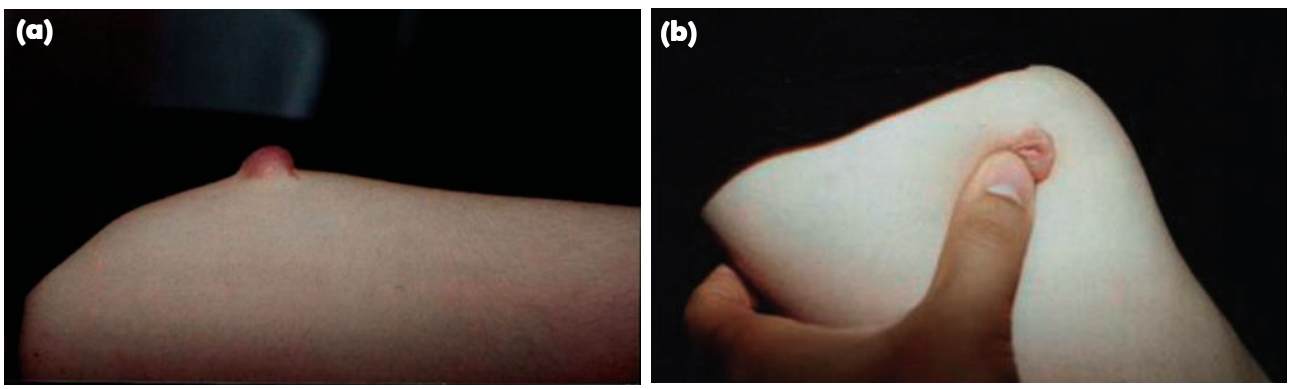

Figure 1. (a) Single flaccid bulla, measuring 3 x 2 cm on the right shoulder. (b) The skin over the bulla could be pushed deep into the dermis.

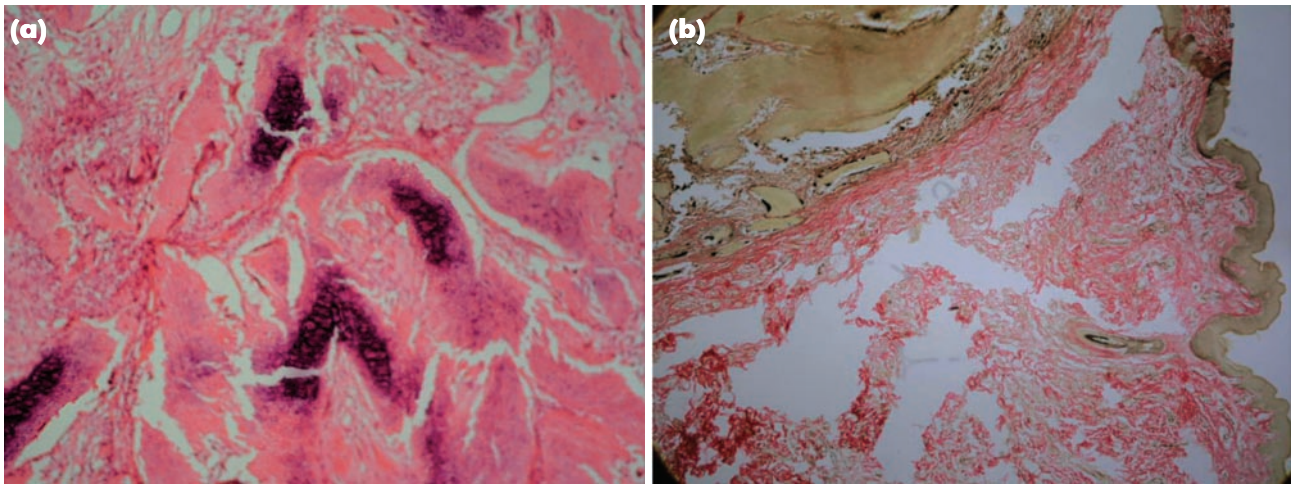

Figure 2. (a) The tumour is composed of irregular nests of basophilic cells, shadow cells and transitional cells in a fibrotic stroma with focal areas of calcification (H&E, Original magnification x 400). (b) Marked reduction in elastic fibres (Verhoeff van Gieson stain, Original magnification x 100).

second or third decade of life. Most of the lesions were located on the arms or trunk. Usually the lesions manifested as soft, wrinkled, protuberant lesions with a firm underlying subcutaneous mass. The treatment of choice was surgical excision and recurrence was not reported.[3-12] The size of common pilomatricomas ranges from 1 x 1 cm to 6 x 3 cm. While it was reported that the size of pilomatricomas with a bullous apperance was larger than that of common pilomatricomas, there was no reported association between giant pilomatricomas and bullous appearance.[1,10]

As pilomatricoma with bullous appearance is rarely reported, this diagnosis may not be immediately apparent to clinicians. Bullous morphoea, lymphangioma and bullous cutaneous lymphoma should also be considered in the differential diagnosis. Other less likely possibilities include bullous lichen planus, bullous lupus erythematosus and other autoimmune bullous disorders.[8,11,12] The diagnosis can be clearly established with histopathological examination.

In this rare and atypical variant of pilomatricoma, secondary anetoderma and bullous appearance may be found.[3-10,13,14] Secondary anetoderma may develop within lesions of various cutaneous inflammatory disorders, infections and tumours. It is characterised clinically by soft sac-like protrusion and pathologically by the focal loss of elastic fibres in the dermis.[15] Marked reduction or absence of elastic tissue in the dermis overlying pilomatricoma lesions was shown in the majority of cases.[3-5,8,13,14] However, in one case reported by Prasad et al, no changes in the elastic tissue were seen,[9] and some authors did not mention about the elastic tissue stain in their reports.[1,6,7,10] In our case, there was a marked reduction in elastic tissue.

Several hypotheses have been proposed to explain the bullous appearance. Although Piguet and Bolgert proposed that mechanical irritation (trauma) was an inciting factor and some authors support this hypothesis,[3,6,10] there were no history of trauma on the lesions in most of the cases including our patient. The second hypothesis suggested by Jones and Tschen was that tumour cells and/or infiltrating inflammatory cells could be producing elastolytic enzymes.[3] Many authors think that the pressure on the area around the

hard core of the pilomatricoma plays a main role in the development of bullous appearance as it induces obstruction and congestion of lymphatics leading to dilatation and oedema.[6-10]

In pilomatricomas with a bullous appearance, apart from secondary anetoderma, there were other significant histopathological findings. Julian and Bowers described the tumour in the bullous variant to be appearing to float in a sea of fluid.[1] Fugioka et al reported that the fluid in the dermis was lymphatic,[13] and in all cases the dermis above the pilomatricoma lesions was oedematous and there were numerous dilated endothelium lined vascular channels, possibly lymphatics.[3-10,13,14,16] Lao et al described an unusual case with pathological changes of papillary endothelial hyperplasia in dilated lymphatic vessels overlying a typical pilomatricoma.[17] As fluid is not present in anetoderma, we think that there should be other mechanisms that account for the presence of lymphatic fluid in pilomatricomas with a bullous appearance and secondary anetodermic features. We suggest that the term "bullous apperance of pilomatricoma" should be used instead of "secondary anetoderma involving a pilomatricoma".

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