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Case Report

A 74-year-old lady with alopecia: alopecia mucinosis idiopathic type

74 歲女患者的斑禿：特發型黏液性斑禿

KK Ho 何家強 and KC Lee 李景中

Alopecia mucinosis is an uncommon cause of alopecia. It can be idiopathic or associated with mycosis fungoides. Histopathological features may help to distinguish the two. Idiopathic alopecia mucinosis usually have a good prognosis.

黏液性斑禿是斑禿的一種少見原因。黏液性斑禿可分為特發型及蕈樣肉芽腫相關型。組織病理學特徵有助區分二者。特發型黏液性斑禿通常預後良好。

Keywords: Alopecia mucinosis, mycosis fungoides

關鍵詞：黏液性斑禿，蕈樣肉芽腫

Introduction

Alopecia mucinosis (AM) or follicular mucinosis (FM) is a clinicopathological disease characterised by follicular papules with or without alopecia. Histologically, it is associated with a non-distinct epithelial reaction pattern characterised by the accumulation of mucin within hair follicles. Two types of primary follicular mucinosis are identified: mycosis fungoides associated and idiopathic type. Idiopathic follicular mucinosis is usually presented with solitary lesion which resolves within years without progression to cutaneous T cell lymphoma. The following is a case report of idiopathic type of follicular mucinosis.

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Case report

A 74-year-old lady presented with hair loss for nine months which was associated with occasional scalp itching and dandruff. There was no systemic upset or joint pain. She denied any recent intake or local application of drugs or herbal medications. The patient had essential hypertension taking anti-hypertensive medication for more than ten years.

On physical examination, a solitary 1 cm x 1.5 cm oval shaped bald patch was noted over her occipital area. The margin was not distinct. Prominent hair follicles and follicular plugging were noted over the alopecic patch (Figure 1). Neither sign of inflammation nor scar were detected. Another ill-defined erythematous patch, 3 cm in diameter, was also noted over her upper back. Her nails and mucosal surfaces were normal and no regional lymph node enlargement or organomegaly were detected. The provisional diagnoses included mycosis fungoides (MF) related alopecia mucinosis, idiopathic type follicular mucinosis, alopecia areata, lichen planopilaris and discoid lupus erythematosus.

Wood's light examination of the scalp showed no fluorescence and skin scrapping of her back lesion revealed no fungi. Results of her complete blood picture, differential cell count, liver and renal function and anti-nuclear factor were normal or negative.

A skin biopsy of the scalp lesion showed perifollicular lymphocytic infiltration associated with mucinous changes. The interfollicular zone was spared (Figure 2). High power examination showed lymphoid infiltration of a hair follicle with mucin deposit between the follicular epithelial cells. There was no definite atypia in the lymphoid component (Figure 3). Direct immunofluoresence study was negative. There was no feature of MF or lymphoma noted.

The patient was treated with fluocinolone acetonide 0.025% twice daily. Her hair totally regrew three months after the biopsy (Figure 4).

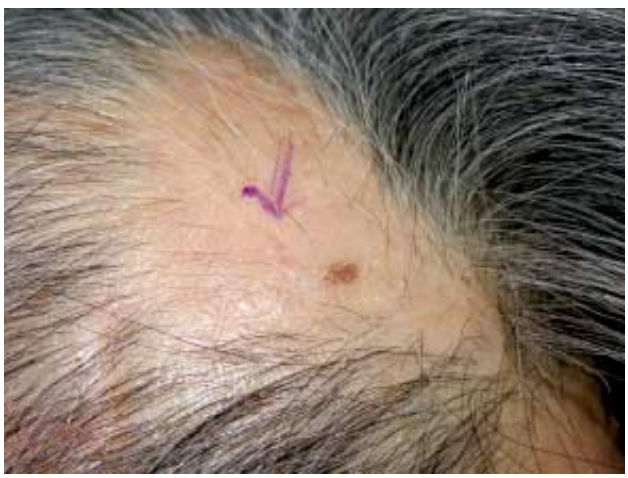

Figure 1. Prominent follicular papules over the patch of alopecia.

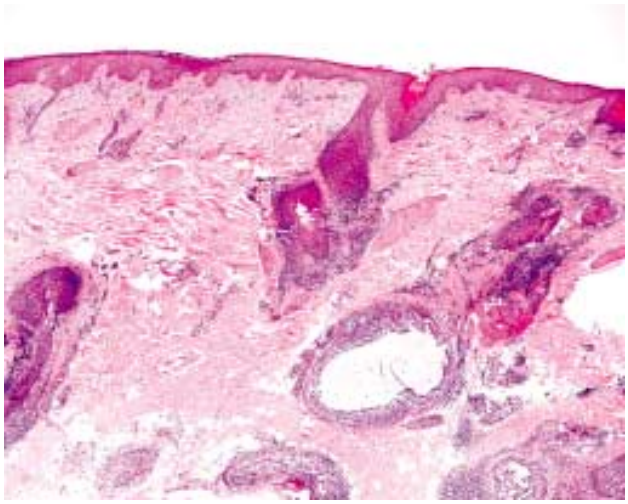

Figure 2. There is perifollicular lymphocytic infiltration associated with mucinous change in the hair follicles. Note the sparing of interfollicular zone (H&E).

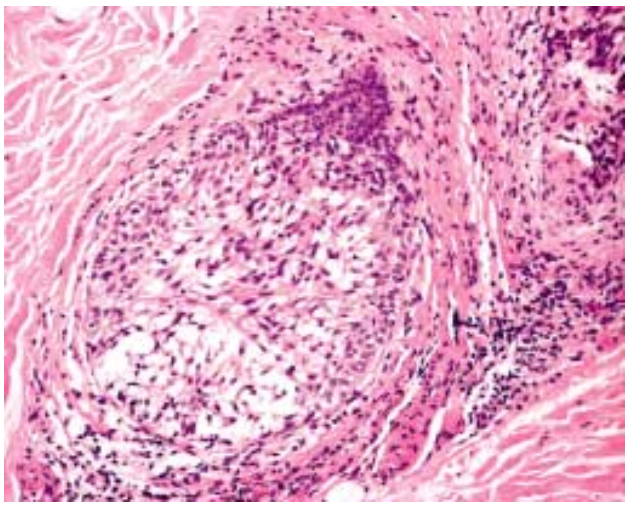

Figure 3. High power showing lymphoid infiltration of a hair follicle with mucin deposit between the follicular epithelial cells. There is no definite atypia in the lymphoid component (H&E).

Figure 4. The previous bald area had hair completely regrown.

Discussion

Pinkus et al in 1957 coined the term alopecia mucinosis to designate localised alopecia that histopathologically showed mucin deposition within hair follicles.[1] Jablonsks et al later changed it to a more histopathological term of FM.[2] The histological features of follicular mucinosis is, however, non specific. FM is classified into primary and secondary. Primary follicular mucinosis is divided into MF associated or idiopathic form. Whereas in secondary group, it can be associated with systemic lupus erythematosus,[3] arthropod bites,[4] eosinophilia associated with HIV infection,[5] non-cutaneous T cell lymphoma, chronic lymphocytic leukaemia,[6] and cutaneous B cell lymphoma.[7]

Idiopathic follicular mucinosis is usually diagnosed by exclusion. It is characterised by a solitary lesion with a tendency to resolve within years. The predictive factors that differentiate between MF associated and idiopathic follicular mucinosis were summarised in Table 1.

The presence of monoclonal T cell receptor (TCR) gene rearrangement proliferation has been suggested to be associated with MF. However, Brown et al reported no evidence of progression to cutaneous T cell lymphoma in a five-year follow-up of five young patients (all less than 40) with primary follicular mucinosis despite the presence of monoclonal TCR gene rearrangement.[8] Cerroni et al studied 44 patients[9] with either MF associated or idiopathic follicular mucinosis. No clear-cut histopathological and immunohistochemical features were noted that could help to differentiate the two. In addition, monoclonal TCR gene rearrangement was detected in about half of the cases in both MF associated and idiopathic groups. No single case of idiopathic follicular mucinosis progressed to MF upon the mean follow-up time 71.6 months was found.

In essence, the inclusion of the presence of monoclonal TCR gene rearrangement to differentiate between MF associated and idiopathic follicular mucinosis may not be reliable. Therefore, long-term surveillance of idiopathic follicular mucinosis and consider re-biopsy from most infiltrated lesions represent the best clinical

Table 1. Predictive factors that help to differentiate between MF associated follicular mucinosis and idiopathic follicular mucinosis

	Idiopathic FM	**MF associated FM**
Age	Young (< 30 years old)	Elderly
Number of lesions	Solitary or localised	Generalised
Disease duration	< 3 years	> 3 years
Histopathology	Absence of Pautrier microabscesses, Confinement of atypical lymphocyte, Absence of monoclonal proliferation	Presence of Pautrier microabscesses, Absence of inflammatory dermal infiltrates, Presence of monoclonal proliferation

approach to monitor for the development of cutaneous T cell lymphoma.

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