Acneiform follicular mucinosis

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Summary

Follicular mucinosis is a rare chronic inflammatory disease of unknown aetiology, presenting as mucin deposits around the follicles and sebaceous glands. It can progress to alopecia of the scalp and other hairy areas. Follicular mucinosis may be a benign primary idiopathic disorder or secondary to malignant lymphoproliferative disorders. It can present with shiny papules or sharply margined infiltrated erythematous scaling plaques, with follicular accentuation on the scalp, neck, trunk and limbs. There are many local and systemic treatments. This paper discusses the case of an adult with an uncommon acneiform follicular mucinosis controlled with systemic corticosteroids.

Report

Pinkus and Braun-Falco first described follicular mucinosis in 1957 under the term alopecia mucinosis, which was given in reference to a disease process of follicular degeneration resulting in alopecia. Histologically the process manifests as deposition of mucin in the epithelium of the follicular outer root sheath and sebaceous glands. The condition was later renamed follicular mucinosis by Jablonska, Chorzelski and Lancucki in 1959 because alopecia is not always present.

Follicular mucinosis is a chronic inflammatory disease of unknown aetiology. It is more commonly observed as shiny pink or pale-coloured follicular papules or as erythematous scaling infiltrated plaques. The involved follicles may show conspicuous horny plugs, presenting with alopecia when the scalp or other hairy regions are involved. The lesions may be localized or widely disseminated. There are various distinctive clinical presentations: alopecia areata-like, scarring alopecia, nodules, cysts, chronic eczema and acneiform eruption.

Follicular mucinosis can present as a benign primary disorder or be associated with malignant lymphoproliferative processes such as cutaneous T- and B-cell lymphomas, Hodgkin’s disease, chronic and acute lymphocytic leukaemia. Two categories of disease have been identified in patients with the benign primary form. The first occurs in younger patients: there are few lesions limited to the head and neck or upper arms and which disappear spontaneously between 2 months and 2 years after onset. The second is a disorder of adult patients: the lesions are larger and more widespread and can take a longer period to improve. In the second type, which is linked to malignant lymphoproliferative conditions, the patients are older and have widespread and infiltrated lesions that can progress to mycosis fungoides (MF).

The origin of the mucin deposits in the follicles is unknown, but immunopathological studies have shown that cytokines released from perifollicular T lymphocytes might stimulate the follicular epithelium to secrete mucin. Immunohistochemical studies have shown expansion of clonal T cells in many cases of primary ‘benign’ mucinosis. The clonality is not always synonymous with malignancy, but its presence is a reason for caution.

Various therapies have been tried for follicular mucinosis: indomethacin, topical and systemic corticosteroids, dapsone, topical tretinoin, oral isotretinoin, minocycline, tetracycline and psoralen with ultraviolet A. When the condition has been associated with lymphoma, radiotherapy, topical nitrogen mustard, electron beam radiation and immunotherapy have been tried.
We report the case of a black 36-year-old male patient, who complained of nodules on his face with slight pruritus that increased after sun exposure. The lesions first became apparent 1 year before presentation. Physical examination showed multiple red or pale isolated papules and nodules on his forehead (Fig. 1a), temples and behind his ear, without lymphadenopathy. The patient had no history of acne. At that time the diagnostic possibilities included: granulomatous rosacea, demodicidosis, cutaneous tuberculosis, sarcoidosis, Jessner–Kanof lymphocytic infiltrate, lupus erythematosus, lymphocytoma cutis, lymphoma and syphilis.

Histopathological examination showed deep and superficial dermatitis with an inflammatory infiltrate surrounding the hair follicles. A few eosinophils were present. Special stains for fungi and acid-fast bacilli were negative. Demodex folliculorum was not isolated. Syphilis and HIV serology were negative. Rosacea was the most likely diagnosis and so tetracycline, 1.5 g per day, was given for 30 days but the patient did not respond.

A second biopsy revealed an inflammatory infiltrate around the follicles composed of lymphocytes, histiocytes and moderate number of eosinophils. Follicular epithelium showed cystic holes provoked by intracellular and intercellular oedema (Fig. 2). Deposition of mucin in the follicular epithelium was demonstrated by Alcian blue stain. These findings confirmed the diagnosis of acneiform follicular mucinosis. Laboratory examination revealed a normal full blood count, renal and liver function tests and chest X-ray.

Figure 1 (a) Multiple erythematous follicular papules and nodules on the forehead and temples before treatment. The papules are not coalescent and present an acne-like aspect. (b) Complete regression of the cutaneous lesions following treatment with oral prednisone 40 mg/day.

Figure 2 Histopathology shows, in the centre of the field, a follicle surrounded by a mixed inflammatory infiltrate. Mucin deposits can be seen in the outer sheath of the follicle (highlighted in the figure inset). (Haematoxylin & eosin).
We decided to treat the patient with oral prednisone, 40 mg/day for 20 days. The patient improved quickly and the lesions disappeared entirely (Fig. 1b). Prednisone was gradually decreased by 10 mg weekly and was discontinued after 48 days. The patient has been followed up carefully for 7 months without relapse.

Deposits of mucin in the follicles have been described in various dermatoses: alopecia areata, lichen planus, melanocytic nevus, lentigo maligna, sarcoidosis, squamous cell carcinoma, lupus erythematosus, arthropod bites, pseudolymphoma and Hodgkin’s disease. Some authors have described follicular mucinosis with an acneiform clinical presentation but this raises the question of whether the deposition of mucin around the follicles is primary or secondary. We consider that further studies are necessary to clarify this aspect.

When the infiltrated lesions are generalized with a chronic course in patients over 30 years old the risk of association with MF is high. Histopathological features that draw attention to this include the presence of atypical lymphocytes, epidermotropism and a paucity of eosinophils. However, there are reports of follicular mucinosis that progressed to MF in children or in patients with only a few isolated lesions on the face and neck. Some authors have observed that MF associated with, or preceded by, follicular mucinosis is more aggressive with lymph node involvement; histopathological examination shows folliculotropism without epidermotropism, i.e. follicular MF. The folliculotropic variant of MF may require aggressive and distinct modalities of treatment because 7% of cases progress rapidly to lymph node involvement as compared with less than 3% of classic MF cases. At present the treatment options are electron beam radiation, immunotherapy and photopheresis.

Patients with follicular mucinosis, whether children or adults, presenting with uncommon clinical variants must be followed up clinically and histologically to detect any evolution into cutaneous lymphoma.

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References