Follicular mucinosis in a teenage girl

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SUMMARY

Follicular mucinosis is characterized by mucin deposits within the hair follicles and sebaceous glands. It can occur as a primary idiopathic benign disorder; it can be found as an incidental phenomenom in rare cases of different dermatoses; or it can arise as a secondary symptom of a malignant disorder, most commonly mycosis fungoides. Youth and localization in a single area used to be regarded as indicative of the benign nature of follicular mucinosis. However, recent reports demonstrate that no clear-cut criteria allow for the differentiation of idiopathic from lymphoma-associated follicular mucinosis.

We report the case of a 16-year old girl who presented with a 2-year history of a single, slightly pruritic, erythematous patch on the left side of the neck. Different local treatments (local corticosteroids, antibiotics, antifungal ointments and moisturizers) were ineffective. Overall, she was healthy. Histopathological examination of the lesion showed the typical histological picture of follicular mucinosis.

KEY WORDS

Follicular mucinosis, young age, mycosis fungoides

Introduction

Follicular mucinosis (FM) is a rare disorder, characterized histologically by mucin deposits within the hair follicles and sebaceous glands. Alopecia mucinosa is another term that describes the same condition (when lesions affect hairy skin, alopecia is prominent) (1).

Follicular mucinosis can occur as an idiopathic, benign disorder, but can also be associated with malignancy. Youth and localization in a single area used to be regarded as indicative of the benign nature of this disease. However, recent reports demonstrate that there are no reliable criteria for differentiation of idiopathic from malignancy-associated follicular mucinosis (2). The therapeutic options currently available for treatment of follicular mucinosis are of limited efficacy (1).

We report the case of a 16-year old girl with follicular mucinosis.

Case Report

A 16-year old girl was referred to our department for evaluation and treatment of an erythematous patch on the left side of the neck. She had noticed a single,

slightly pruritic, erythematous patch on the left side of the neck 2 years before and had been treated unsuccessfully with topical corticosteroids, antibiotics, antifungals and moisturizers. The patient had no other skin or general diseases.

Physical examination revealed an erythematous patch of 7x6 cm on the left side of the neck. Fine scaling of the lesion and a narrow ring of slightly hypopigmented skin around the lesion were observed. However, there was no clinically evident infiltration of the lesion. General physical examination revealed neither lymph node involvement nor any other abnormalities. Dermatophytosis was ruled out by direct microscopy examination and fungal culture tests.

Histopathological examination showed somewhat irregular hyperplasia of the epidermis with some scale-crusts and both superficial and deep perivascular, perifollicular and follicular inflammatory cell infiltrates of lymphocytes, histiocytes and a few eosinophils. Mucin deposits were present within follicular infundibula and outer root sheaths. Atypical lymphocytes and Pautrier microabsceses were not observed.

All laboratory examinations, including erythrocyte sedimentation rate, blood cell counts and urinanalysis were within the normal range. Chest x-ray and abdominal ultrasound showed no lymph node enlargement or other pathological findings (except a cyst in the left ovary).

Based on these findings, the possibility of malignancy was ruled out. No specific treatment has yet been introduced because of the benign nature of the lesion and because the earlier local treatments had caused burning sensations. However, a close follow-up with repeated biopsies is going to be instigated.

Discussion

FM is a histological reaction pattern (3). It can be associated with a wide range of benign and malignant conditions (1,4).

Clinically, FM typically presents with erythematous, infiltrated plaques with scaling and sometimes follicular prominence on the head and neck (1,4). When lesions affect the skin regions covered with terminal hair, non-scarring alopecia is prominent (1). Anesthesia and dysesthesia of the lesions has been reported (1). Our patient presented with a pruritic and burning sensation in the lesion and did not tolerate any local treatment very well.

However, many other clinical presentations have also been described: acneiform eruption, hypopigmented, erythematous and eczematous plaques, flesh colored follicular papules and indurated nodules (5,6).

FM can be differentiated into three clinical types. The most common is idiopathic (primary or benign) FM, that is usually to be found in young patients without concomitant cutaneous or extracutaneous disorders, and typically presenting as solitary lesions which tend to resolve within a few years. Secondary or malignancy associated FM usually occurs in the elderly, and presents as widespread lesions. The third type is persistent or chronic benign FM, comprising a combination of the clinicopathologic characteristics of the first two types (1,2,5).

The existence of the third type is questionable as it is probably a form of malignancy associated FM (2).

The most common malignancy observed in association with FM is cutaneous T-cell lymphoma (mycosis fungoides). The reported incidence of T-cell lymphoma ranges from approximately 15 to 30% of patients with FM (7–9). Although mostly seen in adults, cutaneous T-cell lymphoma may also occur in very young adults and children (10). Other malignancies that have been associated with FM include leukemia, Hodgkin's disease, renal carcinoma, cutaneous B-cell lymphoma, lymphosarcoma and squamous cell carcinoma of the tongue (1).

Mycosis fungoides may precede FM, occur simultaneously with FM or develop months to years after a diagnosis of FM has been made. It is usually made within one year, but cases have been reported of mycosis fungoides developing up to eight years after the primary diagnosis of FM) (7,11).

It was believed that idiopathic FM occurs in young patients, with lesions limited to the head and neck region, which resolve spontaneously within two years (4). Recent literature shows however that adults over 40 years of age with widespread persistent lesions are at greater risk of developing mycosis fungoides (7).

It has also been suggested that a clonal T-cell receptor gene rearrangement could help to differentiate malignancy associated FM (4).

However, recent reports have demonstrated no reliable clinical and histopathologic features or criteria for differentiation between idiopathic and malignancy-associated FM (2,4,10). There is a considerable overlap between the groups in terms of the age of the patients, the location of the lesions and the degree of generalization of the disease (solitary vs. multiple lesions), and, more surprisingly, in terms of histopathologic findings and immunophenotype (2). These results suggest that idiopathic FM may represent a variant of mycosis fungoides with localized lesions and an excellent prognosis. Therefore, in cases of "idiopathic or benign FM" a long term follow-up with repeated biopsies is required, and a minimum period of five years has been proposed (2,4).

Generally, no treatment has been described as consistently effective. A wide range of therapeutic options have been tested: topical and oral antibiotics, topical retinoids, isotretionin, steroids (topical, intralesional, oral),

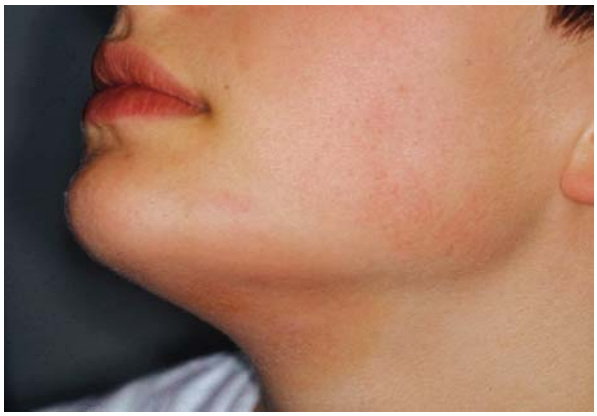

Figure 1. Slightly erythematous, infiltrated patch on the neck.

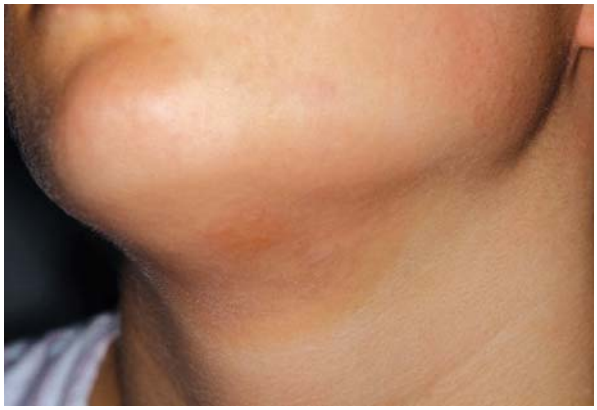

Figure 2. 7x6 cm large, rather well circumscribed erythematous patch with fine scaling and a narrow, slightly hypopigmented ring around it.

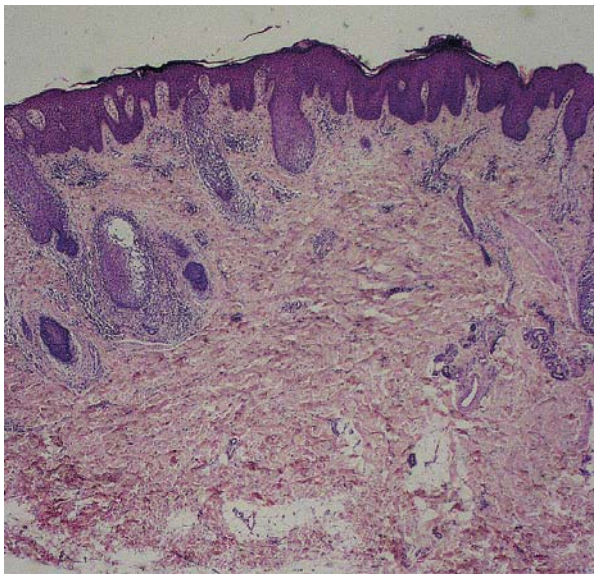

Figure 3. Epidermal hyperplasia, scale-crusts, spongiotic folliculitis and perifolliculitis. HE x 40.

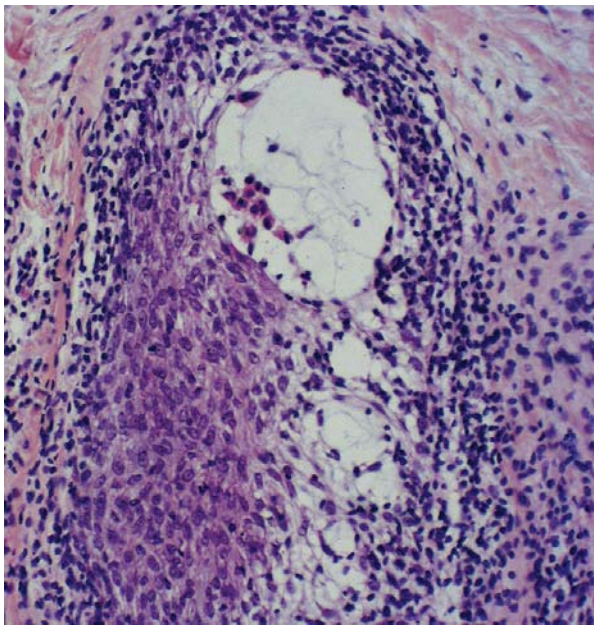

Figure 4. Accumulation of mucin within the hair follicle and inflammatory infiltrate located adjacent to and within the follicle. HE x 200.

dapsone, methotrexate, immunosuppressive drugs, nitrogen mustard, PUVA, UVA, excision, X-rays and others (1,4). The treatment of malignancy associated FM is directed toward the underlying malignancy (1).

Considering the lack of reliable criteria allowing a differentiation of idiopathic from lymphoma-associated follicular mucinosis, we shall make a close, long-term follow up with repeated biopsies.

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