Case Report

Primary Follicular Mucinosis: A Case Report From Saudi Arabia With Successful Treatment And Literature Review

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ABSTRACT: Background:Follicular mucinosis is an uncommon inflammatory disorder that characteristically presents as clearly defined, erythematous plaques or papules, with follicular projections, superficial scaling, and alopecia in terminal hair bearing areas, characterized histologically by mucin accumulation in pilosebaceous units (follicular epithelium and sebaceous glands) . The condition is generally divided into primary (idiopathic) and secondary forms in association with several conditions including benign and malignant diseases. There are many local and systemic treatments.

Main observations: We report a case of 15 years old male with primary follicular mucinosis treated effectively by intralesional steroid injections.

Conclusions: This is a new case of Primary follicular mucinosis from Saudi Arabia was treated successfully with intralesional corticosteroids without relapse.

KEYWORDS:follicular mucinosis, intralesional corticosteroids, treatment.

INTRODUCTION

Follicular mucinosis is a rare condition, of unknown cause, which affects all races, ages and both sexes.[1,2]It is defined as the accumulation of mucin in the follicular epithelium and sebaceous glands.[3,5] It was initially described in 1957 by Pinkus who named it alopecia mucinosa and renamed it mucinose follicular in 1959, due to the fact that alopecia is not always present.[6,7]Clinically characterized by sharply demarcated infiltrated erythematous papules orplaques with follicular prominence, scaling and alopecia .[8,9] Less commonly, nodular lesions, cysts, chronic eczema, follicular spines and acneform lesions have been described.[10-14]It can present in isolation with an unknown etiology usually present in children and young adults, of spontaneous remission or in association with several conditions,including mainly , cutaneous T-cell lymphoma (mycosis fungoides[15]) and less commonly inflammatory or malignant diseases including (lupus erythematosus,[16,17]insect bites,[18]eczema, alopecia areata,[19,20] hypertrophic lichen planus ,[21]Sezary syndrome, leukemia cutis,[19]cutaneous B-cell lymphoma and Hodgkin's

disease).[2,22]Clinical and histopathological criteria are fundamental for the distinction between primary and secondary forms.[6,7]Effectivetherapeutic options for patients with primary idiopathic FM are limited .[2]

CASE REPORT

15 years old, male, presented with 6-months history of asymptomatic, solitary, and slowly growing plaque on the forehead.There is no history of injury, local infection, or insect bite, also no history of animal contact, photosensitivity or recent travel, Review of systems was negative, and the patient was otherwise healthy. He had no significant past medical or family history and was taking no medications.Skin examination revealed single erythematous, scaly plaques about 3x3 cm with overlying alopecia, on his forehead over the left eyebrow(figure1), all other examination was normal. Exam of the head and neck revealed no lymph node enlargement.

Skin biopsy shows prominent perifollicular inflammatory cell infiltrate, comprised mainly of lymphocytes, with mucinous degeneration of the follicular epithelium (figure3a,b). There was no evidence of lymphocytic atypia, epidermotropism or any feature of lymphoid malignancy. Follicular destruction and granulomatous inflammation were absent. No fungi seen.Alcian blue staining confirmed the presenceof intrafollicular mucin deposits (figure3 c).

According to these finding the case was diagnosed as primary follicular mucinosis. The patient was treated with intralesional triamcinolone 2.5mg/ml every 4 weeks for 3 months and shows significant improvement (figure2).

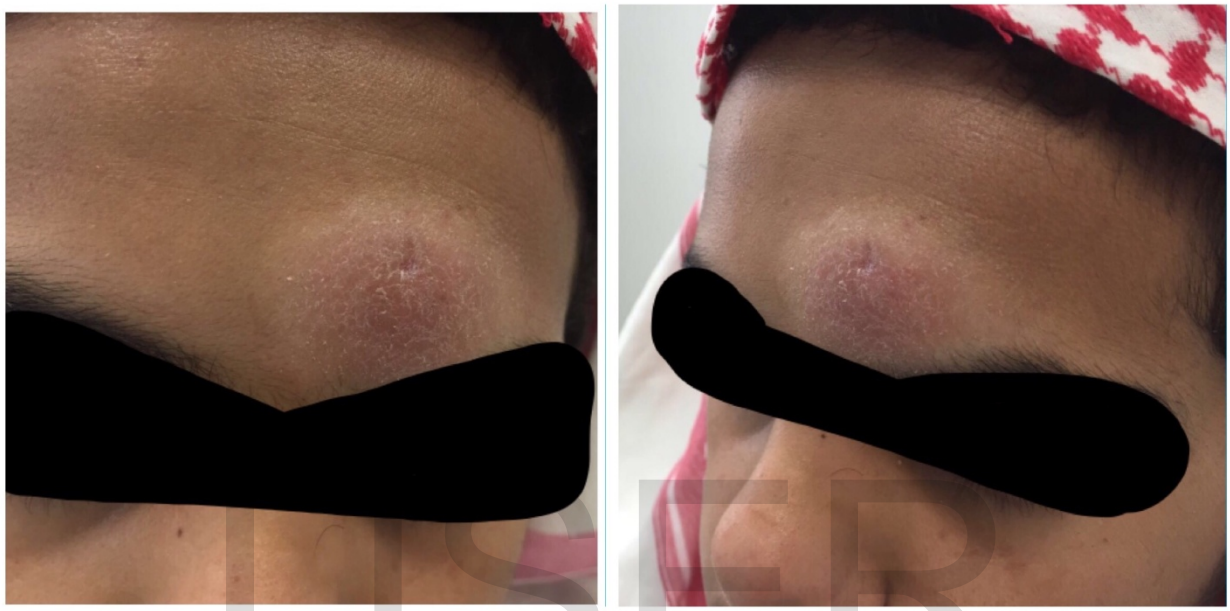


Figure 1: *scaly, erythematous plaques with overlying alopecia over the forehead*

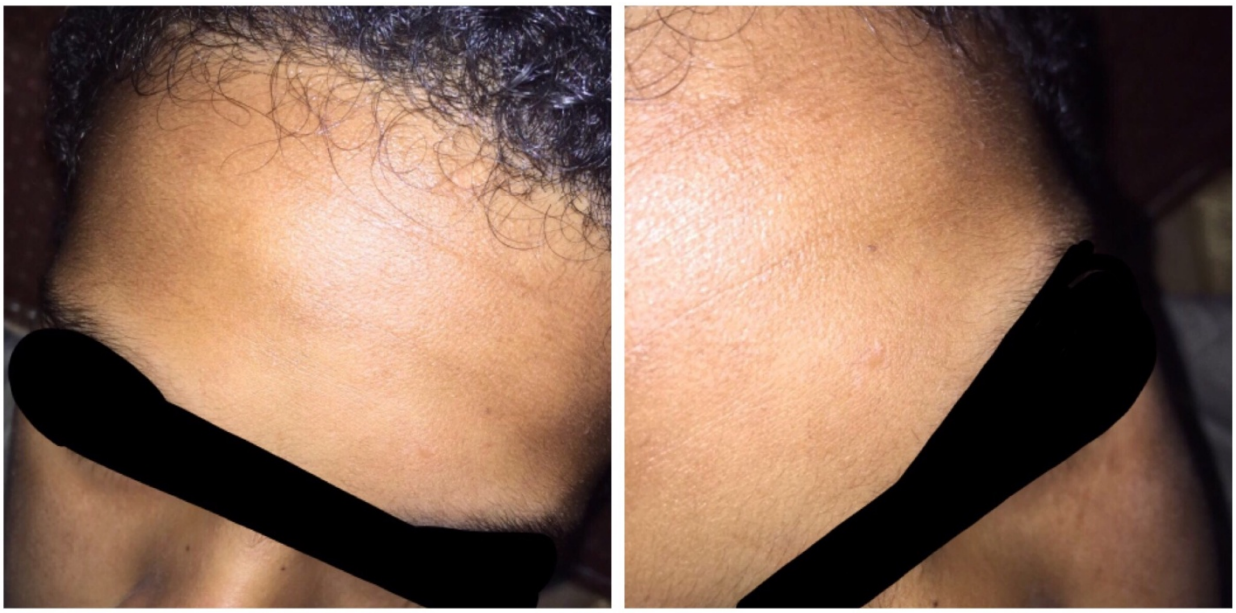


Figure 2. *Post-treatment showing complete resolution of the lesions.*

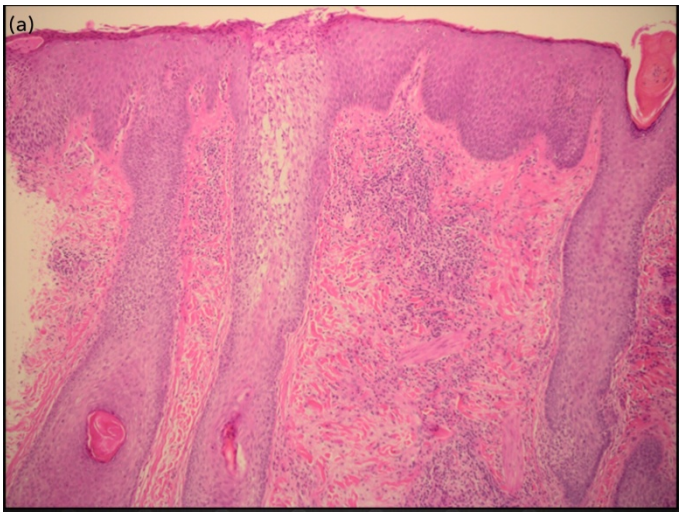
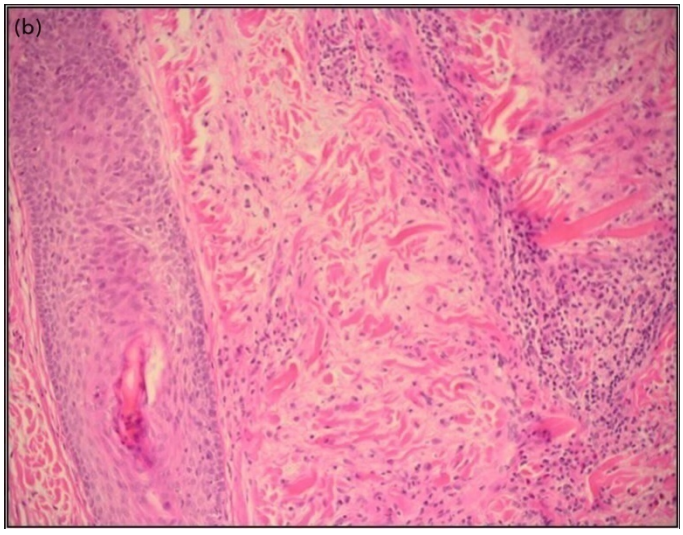
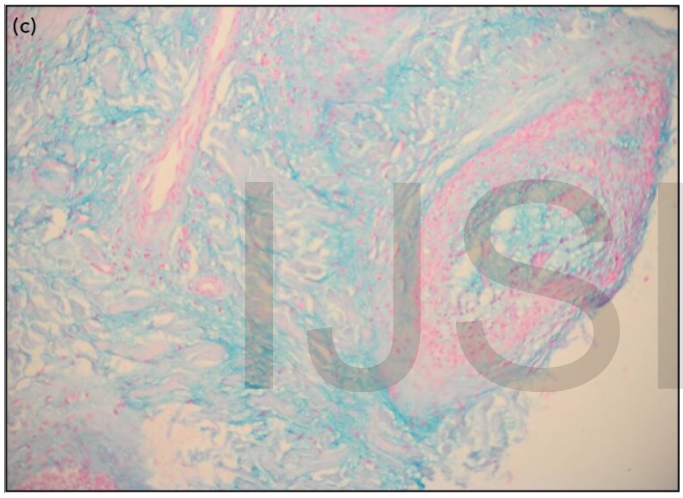


Figure 3

(a) Perifollicular inflammation and widespread mucinous degeneration of the follicular and sebaceous epithelium

(b) mucinous degeneration of the follicular epithelium. Haematoxylin and eosin





DISCUSSION

Follicular mucinosis was first reported by Pinkus in 1957 in a description of the histology findings for a series of patients with characteristic cutaneous lesions and mucin deposits in the hair follicles.[5]It was initially named alopeciamucinosa and was renamed

mucinosefolicular by Jablonska et al in 1959, due to the fact that alopecia is not always present.[10,17]

Folicularmucinosis is a relatively rare epithelial reaction pattern that is characterized by the accumulation of mucin in the follicular epithelium and sebaceous glands with a superficial and deep perivascular and interstitial mixedcell infiltrate.[16,24]FM has been observed in all races and ages, and in both sexes equally.[1,2]

The follicular keratinocytes have been considered to be the source of mucin as a response to the stimulus of cytokines released by perifollicular T lymphocytes. The exact pathogenesis is unknown, although the role of circulating immune complexes and cellmediated immunity has been considered .[24,25]

The dermatosis can have several clinical variants. Primary (idiopathic) follicular mucinosis (PFM) can occur in children and young adults as well as older adults. PFM in young people tends to involve the head and neck, with resolution after 2 to 24 months. Clinically, it is characterized by one or several lesions. Most of the cases have a benign course and demonstrate spontaneous resolution with no relapses. However, rare case reports of the development of Hodgkin's disease, other lymphomas, and leukemia have been seen.[24,26] PFM in older adults usually affects people older than 40 years. The inflammatory lesions are more

numerous, more widespread (extremities, trunk and Face), and more variable and may persist or progress for many years without evidence of associated disorders .[24-27]

Secondary follicular mucinosis (SFM) occurs in older patients (usually aged 40-70 years) and is associated with an underlying inflammatory (lupus erythematosus,[16,17]lichen simplex chronicus, and angiolymphoid hyperplasia[28])or neoplastic condition (mycosis fungoides,[15]Hodgkin disease,[2,22]Kaposi sarcoma), with mycosis fungoides (MF) being the most common malignancy.(table 1)[15]

Table 1:Conditions associated with follicular mucinosis

alopecia mucinosa	Photoinduced eruption
sarcoidosis	Arthropod bites
Lupus erythematosus	Hypertrophic lichen planus
Spongiotic dermatitis	Lichen striatus
Intradermal melanocytic nevus	Lichen simplex chronicus
Mycosis fungoides	Acne vulgaris

Pseudolymphoma Cutaneous B-Cell Lymphomas	Lymphoma Leukemia cutis

The morphology of lesions on idiopathic and secondary forms is identical. Lesions present as clearly defined, slightly infiltrated erythematous plaques or papules, with follicular projections, superficial scaling, and alopecia in terminal hair bearing areas.[1,2] Other less common presentations include acneiform lesions[13,14] Urticaria-like,[31] hypopigmented,[29,30] erythematous, eczematous plaques, flesh colored follicular papules, and indurated nodules.[8,32] Patients may experience anesthesia to cold or touch over the lesion.[33,34]

Histologically, follicular mucinosis is characterized by mucinous degeneration of the follicular epithelium, with the presence of dense fibrous material in the form of amorphous mucin deposits in the pilosebaceous units and a mixed perifollicular inflammatory infiltrate.[5,24,36]Routinely, staining- alcian blue, colloidal iron, toluidine blue for mucin is required which reveal amorphous deposits within the follicles.Clinicohistopathologic correlation favored a diagnosis of Primary follicular mucinosis in our patient.

Because of the close association with MF/CTCL and because in some cases T-cell clonality is detectable, controversy exists as to whether FM is in fact a neoplastic process or a clonal inflammatory condition.[10,15,38]To date, no consistently reliable features (ie, patient age, distribution of lesions, light microscopic/histopathologic features, molecular studies) have been shown to predictably distinguish "benign" FM from lymphoma-associated FM.[15,37,38] Features in favor of a primary form are the young age of the patient, a solitary plaque or limited number of lesions in the head and neck region, spontaneous resolution, and the absence histologically of epidermotropism and atypical lymphocytes but there is no absolute distinction .(table 2)[15,18,37,39]

Table 2. 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 Pautriermicroabscesses - Confinement of atypical lymphocyte - Absence of monoclonal proliferation	- Presence of Pautriermicroabscesses - Absence of inflammatory dermal infiltrates - Presence of monoclonal proliferation

The onset of MF might precede, occur concomitantly or years after the diagnosis of FM. In almost all cases in which there is development of MF this process happens within 5

years, one year on average.[37,40,41] There are however a few reports where lymphoma onset was late, occurring 15 years after the FM diagnosis.[4,15]

There are no standard treatment regimens have been established for idiopathic FM. A wait and see approach is usually recommended , since many of them resolve spontaneously between 2 and 24 months.[24] Several therapeutic modalities have been reported with variable results include: topical corticosteroids,[9,25,29,35]intralesional or oral corticosteroids,[11]topical retinoids,[43]topical Imiquimod 5% Cream,[44]topical calcineurin inhibitors,[45,46,36]oral isotretinoin,[27,47] dapsone,[31,48] antimalarials,[49] indomethacin,[42,50]minocycline,[23,51]PUVA,[52] interferon,[42,53]photodynamic therapy .[54]Treatment of secondary form is the treatment of the associated disorder.[2]

In patients with persistent primary FM, long-term surveillance and biopsy of atypical lesions currently represent the best clinical approach to monitor for the development of cutaneous lymphoma.

In the case presented, there was total remission of lesion with the use of intralesionaltriamcinolone 2.5mg/ml every 4 weeks for 3 months. The patient has been followed up carefully for 7 months without relapse.

Up to our knowledge there is no single case of FM successfully treated with intralesional corticosteroids has previously beenreported. The response to intralesional corticosteroids in ourcase was rapid.

CONCLUSION

We have presented a typical case of primary FM in its morphology as well as its histology and age of presentation with completeclinical remission after treatment withintralesional corticosteroids. The patient with primary FM must be orientedregarding the necessity of a follow-up for the earlydetection of alterations signs secondary to malignancies.Such orientation is mandatory, and also suggestedis patient follow-up for a minimal period of 5years. Our patient remains under clinical follow-upfor 7 months without relapse.

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TABLE LEGENDS

follicular mucinosis and idiopathic follicular mucinosis	

FIGURE LEGENDS