CASE REPORT

Folliculotropic mycosis fungoides in a child: A rare case

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Abstract

Folliculotropic mycosis fungoides (FMF) is a variant of mycosis fungoides characterized by infiltration of hair follicle epithelium by neoplastic lymphoid cells. Generally, it is usually typified by indurated plaques and tumours mainly on the head and neck. However, a wide range of clinical signs have been noted. The clinical presentation of FMF may include prurigo-like lesions, acneiform lesions, cysts, nodules, areas of scarring alopecia, and hypopigmented plaques or papules with follicular prominences. The average age of diagnosis is 60 years while it is rare in childhood and adolescence. We discuss the case of a 12-year-old male patient who had an asymptomatic, erythematous, infiltrating plaque across his left nasolabial fold for three months prior to presentation. Histological assessment of lesion showed characteristic findings of follicular mucinosis with predominance of CD4+ lymphocytes and immunohistochemical studies were positive for CD3+ stains. An increased CD4:CD8 ratio and negative CD20 was also shown. Both findings were consistent with diagnosis of FMF.

Keywords: Childhood tumors, hypopigmentation, melanocytes, lymphoma, CTCL, mycosis fungoides.

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Introduction

Mycosis Fungoides (MF) is a variant of primary mycosis fungoides characterized by infiltration of hair follicle epithelium by neoplastic lymphoid cells. It typically presents in its early stage as inflammatory erythematous patches or plaques, with epidermotropism as the histopathologic hallmark of the disease. It pursues an inert clinical path and is characterised by well-defined clinico-pathological features. There is a large number of clinical variants of MF. Some plaques have verrucous or hyperkeratotic appearance while rare icthyosiform and bullae variants have also been described. The most common variants are folliculotropic, poikiloderma, hypopigmented, capillaries-like, verrucous, psoriasiform, icthyosiform and bullous. The prognosis for FMF, a relatively uncommon but aggressive form of mycosis fungoides that typically develops in old age, is worse than that of classical mycosis fungoides.[1] The average age of presentation is near to 60.[2] Early lesions are non-specific, which delays an accurate diagnosis. It primarily presents on the face, neck, and upper torso as acneiform lesions, follicular papules, or erythematous plaques. The plaques exhibit follicular prominence and histological evidence of neoplastic T-cells infiltrating hair follicles, which is attributed to the former's partial destruction and mucinous degeneration.[3] Both dermatologists and pathologists frequently underdiagnose the disease, especially in its initial stages. Folliculotropic mycosis fungoides primarily affects the elderly, and there are relatively few documented examples of it presenting in children or adolescents. We present the case of a 12-year-old child who was diagnosed as a case of FMF on the basis of histopathological examination.

Case Report

A 12-year-old boy was presented at the out-patient department of PNS Shifa hospital Karachi, in August 2022, with a solitary, painless, hypo-pigmented plaque on his left nasolabial area for three months with overlying hair loss. The plaque was noted to steadily getting larger. (Figure 1 & 2). There was no prior history of trauma, itchiness, pain, or loss of sensation on the affected area. Previous surgical

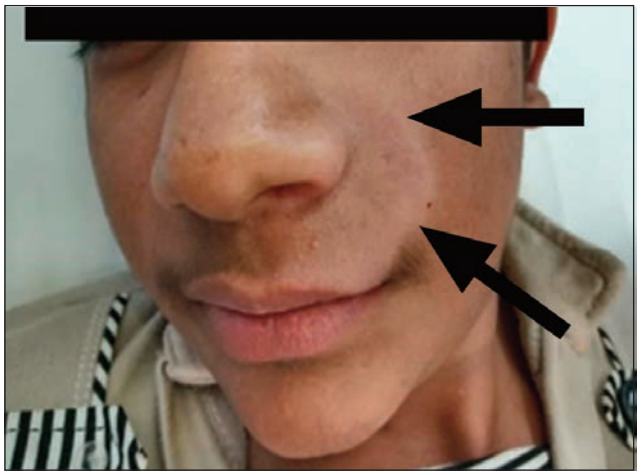

Figure-1: single, well circumscribed hypopigmented plaque involving left nasolabial fold and left lateral upper lip.

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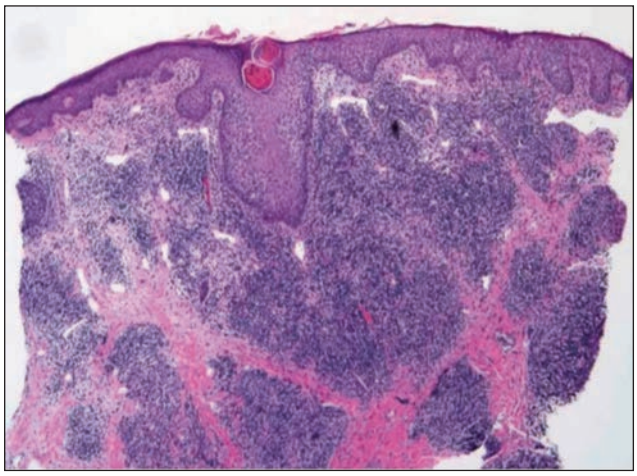

Figure-2: Histopathologic finding reveals a dense, lymphoplasmacytic infiltrate in the papillary dermis and upper reticular dermis. The lymphocytes have slightly enlarged hyperchromatic nuclei with irregular contours.

and medical histories were insignificant. Physical examination revealed a single, 4x5 cm, well-circumscribed, hypopigmented, infiltrating plaque in the left nasolabial region. It was non-tender. There were no swollen nerves identified near the lesion and the sensations over the plaque were intact. There was no lesion on any other part of the body. No associated lymphadenopathy was found. The clinical staging related to TNM classification according to American Joint Committee on Cancer staging classification of this patient was stage 1A as there was a single, 4x5 cm, well-circumscribed, hypopigmented, infiltrating plaque in the left nasolabial region with no evidence of nodal involvement as well as metastasis.[4]

The results of all haematological, biochemical, radiological, and peripheral blood smear tests were normal. Based on the history and physical examination, differential diagnosis for this patient included sarcoidosis, lupus vulgaris, orofacial granulomatosis, tinea incognito and mycosis fungoides. To confirm the diagnosis, an incisional skin biopsy was done. Histopathology revealed mild hyperkeratosis with focal parakeratosis, flattening of rete pegs with mild epidermotropism. Marked follicular mucinosis and focal dilatation associated with moderate lymphoid infiltrate of small to medium sized cells with angulated nuclei were identified. Immunohistochemical staining test showed positive CD3, markedly increased, CD4: CD8 ratio and negative CD20. Typical histopathological features of follicular mucinosis with predominance of CD4+ lymphocytes, was constant with diagnosis of FMF. The patient was given intralesional steroid and asked to follow up in one month.

Discussion

The most prevalent type of cutaneous T-cell lymphoma is mycosis fungoides. About 10% of cases of mycosis fungoides are FMF, a rare and aggressive variant that mostly affects the head and neck .[1] It is usually seen in people aged above 60 years and is not commonly seen in paediatric and juvenile population. To the best knowledge of researcher, there are very few cases of mycosis fungoides reported globally in adolescents and children. The morphological range of lesions is vast and includes nodular and prurigo-like lesions, comedonal, acneiform, and cystic lesions, erythematous papules and plaques with follicular prominence with or without alopecia, plus alopecic lesions with or without scarring.[5] Additionally, hypopigmented plaques have been observed.[6] Leprosy, sarcoidosis, leishmaniasis, alopecia mucinosa, and deep fungal infections are differential diagnoses for plaque lesions. Acne and milia are viewed as differentials for acneiform lesions.

Only a few sources in literature address FMF in juvenile population. In seven individuals, Amitay-Laish identified a unilesional FMF.[7] Two of them were under the age of 18. The lesion appeared as single patch with follicular accentuation, an infiltrating plaque with follicular nodules in one, and a patch without hair in another. Following treatment with topical psoralen and UVA in the 09-years old patient out of six patients there was no recurrence, in up to 10-years follow-up.

Meeta Deepak Mank described FMF in 16-year old boy who presented with an asymptomatic, infiltrated and erythematous plaque on the forehead. The results of histopathological analysis supported the FMF diagnosis. He had successful electron beam therapy treatment.[8] Julian Boyx Volanova discussed the case of a 13-year-old boy who presented with an infiltrated plaque with well-defined borders made of confluent follicular pink-yellowish, slightly scaly papules that had first appeared on his left cheek four months prior and were initially misdiagnosed as eczema by other consultants. Later on, biopsy was performed and it showed features of FMF.[9]

The above-mentioned cases prove that FMF can rarely be seen in juvenile and paediatric population. Histopathological examination of skin biopsy of lymphocytes sample shows a predominance of CD4+ lymphocytes[10] and paucity of CD4+ lymphocytes around the follicle with or without follicular mucinosis and epidermotropism.[11] The term "syringotropic mycosis fungoides " refers to FMF in which hair follicles and eccrine glands are both infiltrated.[12] Psoralen plus UVA has been shown to be effective for FMF in individuals with extensive patches & plaques among the many skin-targeted therapy.[13] Whereas radiation would be advised for those with localised plaque. The prognosis varies, and in general,

cutaneous confined disease in young people tends to wax and wane over decades.

Conclusion

FMF is a rare and aggressive variant of cutaneous T-cell lymphoma, with an average diagnosis age of 60 years. It appears as a hypopigmented asymptomatic plaque most commonly on the face. This variant differs from classic mycosis fungoides as being seen in the younger population, slower progression and the majority of patients remaining in Stage I with treatment. This study underlies that FMF can occur in young people as well. It further shows that histopathological examination and immunohistochemical studies further help in confirmation of the diagnosis.

Informed Consent: The patient's father signed the relevant consent forms, provided his agreement for their photos and other clinical information to be published in a scientific journal Patient was aware that his identity will not be disclosed and that every effort will be carried out to keep it anonymous.

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Conflict of interest: None.

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