CASE REPORT

Folliculotropic mycosis fungoides mimicking leprosy: a case report and review

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Summary  Folliculotropic mycosis fungoides (FMF) is an uncommon subtype of MF that is characterised by an atypical lymphoid infiltrate of the hair follicle epithelium. It involves the head, neck and is often associated with alopecia and occasionally with mucinorrhea. We report this case for its rarity, its association with hypohidrosis, to highlight its resemblance to lepromatous leprosy and to review the FMF–leprosy connection.

Keywords: folliculotropic mycosis fungoides, mimic, leprosy

Introduction

Mycosis fungoides (MF), the most common variant of primary cutaneous T-cell lymphoma, accounts for nearly 50% of all primary cutaneous lymphomas. MF usually presents as slowly progressive lesions that grow over years or even decades from patch to plaque and then to tumor stage. The involvement of lymph nodes and viscera can occur in the later stages of the disease.

Folliculotropic MF (FMF) is an uncommon subtype of MF that is characterised by an atypical lymphoid infiltrate of the hair follicle epithelium. It involves the head and neck and is often associated with alopecia and occasionally with mucinorrhea. We report this case for its rarity, its association with hypohidrosis, to highlight its resemblance to lepromatous leprosy.
Case Report

A 44 year old Chinese man presented with complaints of itchy skin lesions over the trunk and face for 8 years, loss of hair from the head, pubic area, and axillae for 5 years, and decreased sweating with generalised redness and scaling all over the body for 2 years. He had earlier been clinically diagnosed with eczema or lichenoid dermatitis, but both the anti-inflammatory and antihistamines he received failed to reduce his skin lesions or pruritus. There was no family history of similar skin lesions.

On examination he had a leonine facies with predominant infiltrating plaques and loss of eyebrows. Lichenified dermatitis was also present on the upper trunk and thigh and diffuse follicular erythematous-to-violaceous papules were observed all over the body (Figures 1a–1c), except the upper eyelids, palms, and soles.

Clinical features, including hypohidrosis, were suggestive of leprosy, and a skin biopsy and slit skin smear test were done. The slit skin smear test was negative. The skin biopsy showed a predominance of folliculotropism (Figure 2c), compared to a less-remarkable epidermotropism, with intraepidermal clusters of atypical lymphocytes larger than benign recruited dermal lymphocytes, as well as perinuclear vacuolization seen in the patch- and plaque-stage of MF (H&E stain, 2a, b.)

A hemogram revealed a marginally raised erythrocyte sedimentation rate (36 mm at the end of 1 hour) and white blood cell count (≥ 10,800 cells/mm). The remaining investigations,
including liver and kidney function tests, were within normal limits. His serology was negative for HIV as well as for HBsAg and the hepatitis C virus.

Immunohistochemical analysis revealed that most of the lymphocytes were T lymphocytes of the CD2+, CD3+, CD4+, CD5+, CD7+, CD8+, CD20−, CD56−, CD79a−, and CD1a− immunophenotypes (Figure 2e, f), CD30 and Ki-67 sporadic positive, less than 10%. A diagnosis of FMF was made based on histological changes and clinical manifestations such as follicular papules and alopecia.

Figure 2. (a, b) Skin biopsy showing a predominance of folliculotropism, with intraepidermal clusters of atypical lymphocytes larger than benign recruited dermal lymphocytes. (c) Atypical lymphocytes infiltrate in the follicle infundibulum (HE 400x). (d) In the deep dermis, lymphocytes patchy infiltration into the sweat gland (HE 200x). (e, f) Immunohistochemical analysis of CD3+, CD4+ in the epidermis (EnVision two step 400x).
This patient was admitted for 20 days and treated with steroids and narrow-band UVB. After 3 months of treatment, his disorder went into remission, and the hypohidrosis had improved.

Discussion

Classic MF is traditionally divided into three stages: patch, plaque, and tumoral. However, not all patients develop all stages, as some may never progress beyond the patch stage, whereas others may present with lesions of the patch, plaque, and tumoral stages of MF simultaneously. Patients with FMF often present with a more advanced disease stage, although skin staging can be difficult to assess, and patients are often unresponsive to standard treatments. Clinically, the head and neck are the common sites of involvement, though extensive erythematous patches and plaques have been reported on the trunk. Features unusual for MF but seen in FMF are acneiform lesions, cystic comedones, alopecia, and follicular papules. Crusted purulent plaques and infiltration of the tongue are other rare features reported. Histopathologically, in addition to folliculotropism, features such as epidermotropism, syringotropism, follicular mucinosis, and granulomatous inflammation can be seen as per the spectrum of atypical cutaneous lesions.

Several studies have indicated that the prognosis of FMF is poorer than that of conventional MF. The overall survival rates of FMF at 5 years ranged from 60% to 87% compared to >90% for the classic patch- and plaque-stage MF, which confirms the importance of early diagnosis.

The co-existence of leprosy and lymphoreticular malignancies has been reported by several authors. Grossman et al. reported a patient with mycosis fungoides that had progressed to the tumor stage, which responded to chemotherapy and electron beam therapy. Six years later he developed leprosy, confirmed by slit-skin smear and a biopsy done during a reversal reaction showing features of both diseases. The leprosy lesions responded to dapsone and rifampicin.

A connection between leprosy and MF has also arisen, because of morphological similarities in both the diseases. Several authors have reported one being initially misdiagnosed for the other, infiltrative leonine facies being a finding common to the two diseases.

Wen et al. reported a case of a 45 year old man who presented with asymptomatic irregular erythema on the trunk and extremities, with no nerve thickening or sensory loss, who was initially clinically diagnosed as mycosis fungoides. However a biopsy of the erythematous skin lesions showed foamy granulomas perineurally and periadnexally, and Fites staining revealed a large number of acid fast bacilli confirming the diagnosis of lepromatous leprosy. On the other hand, Siriphukpong et al. reported a case of granulomatous mycosis fungoides (GMF) stage IIB that initially presented with leprosy-like features of chronic left peroneal neuropathy, causing a dilemma in the diagnosis. The patient, however, responded to chemotherapy and electron beam therapy. Gutte et al. reported a case of a 38 year old male patient who presented with generalised erythema, scaling and poikiloderma, with multiple nodulo-ulcerative lesions for 2 years, suggestive of lepromatous leprosy. However, histopathology and immunohistochemistry confirmed a diagnosis of GMF, which responded to chemotherapy.
Although there are some similarities in the clinical features of FMF and lepromatous leprosy, and the courses of both diseases are long, they have different histopathological features, and their treatments are completely different. No association has yet been established, and the overlap of clinical features such as the leonine facies and hypohidrosis seen in our case should therefore alert the physician to investigate both diseases, in case the results for one or the other are inconclusive.

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Author’s contributions

N. W. participated in clinical history collection, and analysed the data. X.M.L gave the pathology diagnosis and F.R. Z conceived and supervised the paper.

References