CASE REPORT

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Syringotropic Mycosis Fungoides, an Unusual Variant with Distinctive Features

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ABSTRACT

Syringotropic mycosis fungoides (SMF) is a rare variant of mycosis fungoides (MF). It is characterised by infiltration of ecrine glands by neoplastic lymphocytes. Clinical lesions are frequently seen as solitary, punctate erythematous papules, plaques, and nodules. Pruritus, alopecia, anhydrosis, superficial erosions and ulcers) can also seen in SMF. Histopathologically SMF is characterised by hyperplastic eccrine glands and ducts infiltrated by atypical lymphocytes and syringometaplasia, Here we present a case with SMF, to draw attention about this rare form of MF.

Keywords: Syringotropic, Mycosis fungoides, Folliculotropism

Introduction

Syringotropic mycosis fungoides (SMF) is a rare variant of mycosis fungoides (MF), and it is characterised by infiltration of eccrine glands by neoplastic lymphocytes. According to the current guidelines, SMF is classified in the group of adnexotropic MF with folliculotropic MF (FMF) [1,2].

Clinical lesions are frequently seen as solitary, punctate erythematous papules, plaques, and nodules [1,2]. Pruritus, alopecia, anhydrosis, superficial erosions and ulcers can also be seen in SMF. Colour and shape of the skin lesions may vary; they can be round, circular or irregular in shape, and red, dark red or they can be dark brown in color [1,2,3,4,5,6].

SMF was first identified by Sarkany in 1969 [1,4]. SMF has a predilection for extremities and palm and soles [3,4,7]. Histopathology is the gold standard for diagnosis of MF and STMF. Histopathologically SMF is characterised by hyperplastic eccrine glands and ducts infiltrated by atypical lymphocytes and syringometaplasia [1,3,7]. Although clinically similar to FMF, SMF is less aggressive and has a better prognosis [3,5,6,8].

Here we present a case with SMF, in order to draw attention to this rare form of MF.

Case Report

Eighty-three-year-old man presented with a 1 year history of skin lesions and severe pruritus. First lesion appeared on the arm, followd by legs and abdomen. He did not apply to dermatologist before but he took non-spesific treatment for pruritus. Dermatological examination showed diffuse kserosis, erythematous plaques on abdomen and back, erythematous plaques and nodular lesions on lower extremity with a small ulceration. There was also hair loss on arms and legs (Figure 1A-D). There was no lympadenopathy or hepatosplenomegaly at physical examination. Routine biochemistry was normal. Viral serology was negative for hepatitis B, C and human immunodeficiency virus. Excisional biopsy was taken from the lesions on arm, trunk and legs. Histopathological examination

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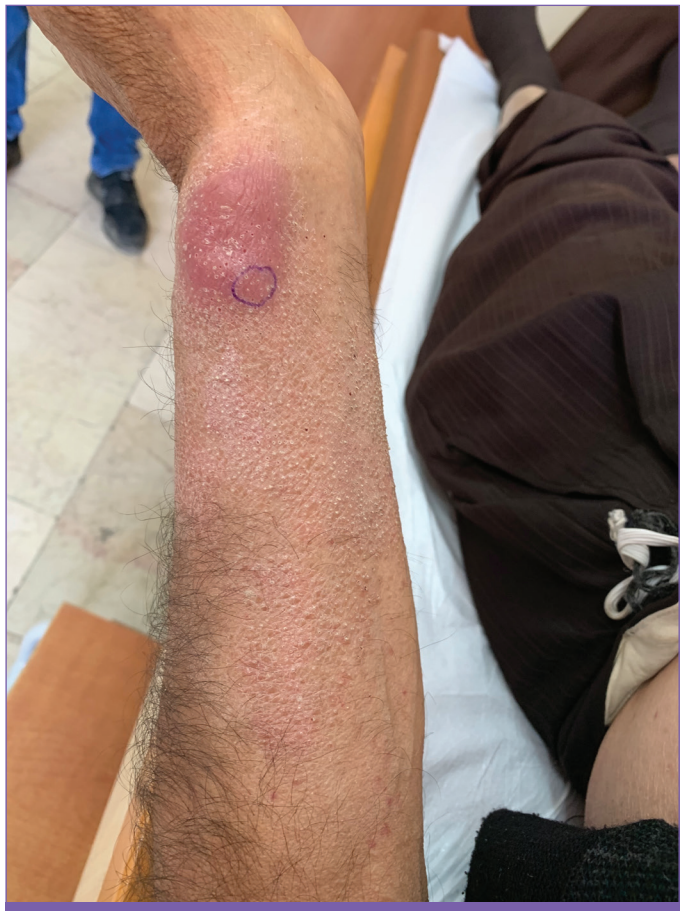

Figure 1A. Nodular lesion on the arm

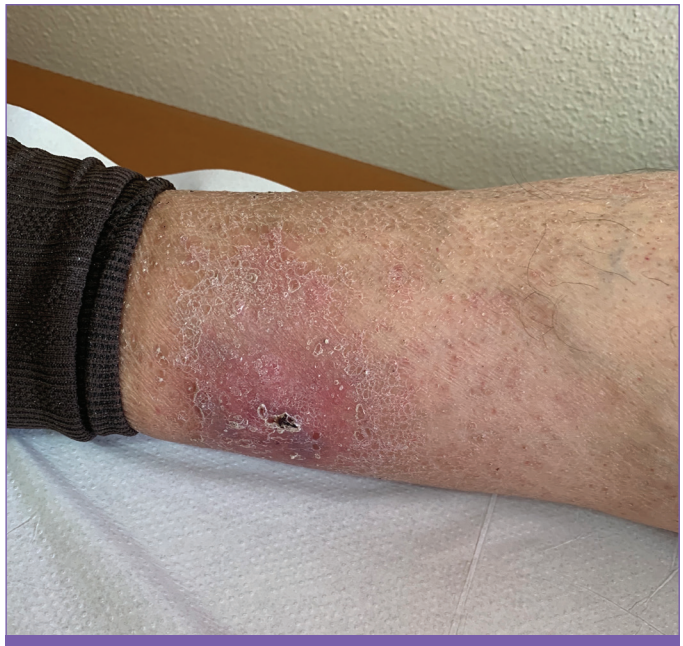

Figure 1B. Kserotik plaque lesion with ulceration

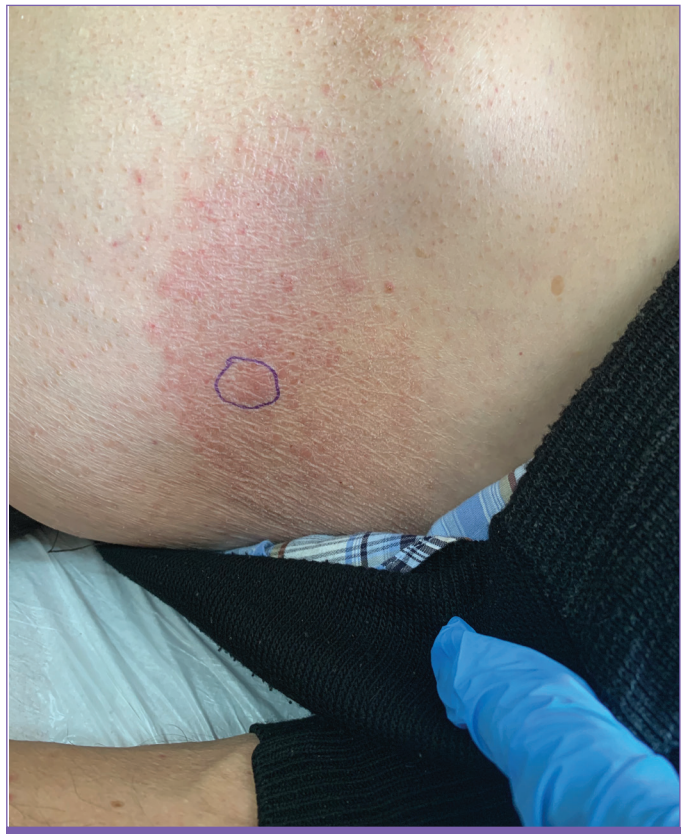

Figure 1C. Kserosis and erythematous plaque lesion on the abdomen

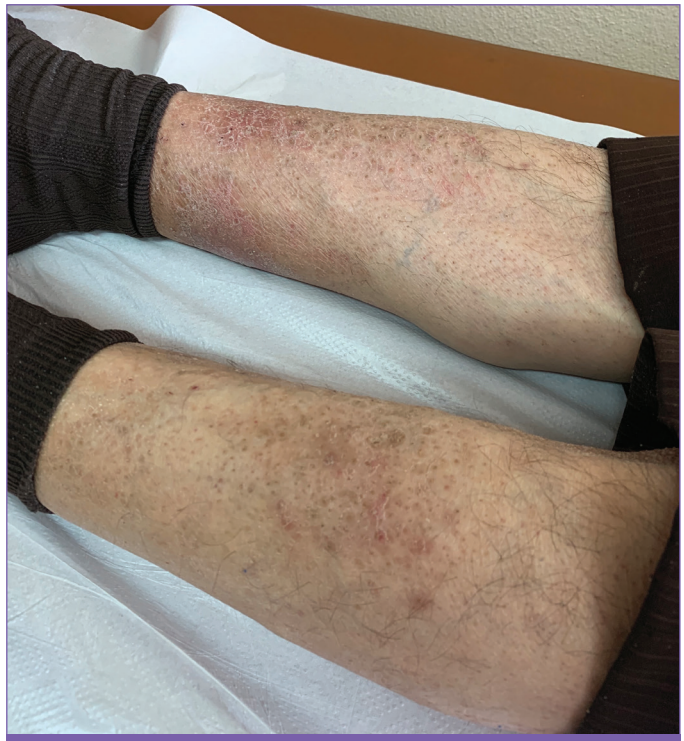

Figure 1D. Kserotic lesions and papular lesions

revealed lymphocyte, plasma cell, eosinophil and neutrophilic infiltration in the epidermis. There were lymphocytic infiltration involving eccrine glands, syringometaplasia, and granulomatous infiltration with two multi nuclear giant cells. Folliculotropism was present. Perifollicular and follicular lymphocytic infiltration were mostly positive for CD4, and CD7, and less predominantly positive for CD8 and CD20 (Figure 2A-H).

Informed consent was taken from the patient for possible case report publication. The patient was accepted as SMF. The patient didn't come to follow up and we couldn't treat the patient.

Discussion

MF is the most common type of cutaneous lymphoma, and it is classified in the group of cutaneous T-cell lymphomas (CTL), which is a heterogeneous group of extranodal non-Hodgkin lymphomas. MF compromises approximately about 50% of all CTLs [6,7]. MF has 3 well-known variants as solitary pagetoid reticulosis (Woringer-Kolopp), FMF, and granulomatous slack skin disease. Other than these variants, many other clinical and/or histopathologic forms have been described including the SMF [6,9]. Although there is a debate about SMF being a different entity or its a subtype of FMF, recent studies assumed SMF as a different entitiy with distinctive clinical and histopathological features [6,8].

Mostly seen lesions in SMF are papules, plaques and nodules [2]. Our patients had papules, plaques and nodular lesions. Ulceration is not a frequent finding in classical MF, but can be seen in SMF [4,10]. Alopecia on the lesions is a frequent finding in SMF [2,3,6]. Body hairs were lost on the skin overlying the lesions of our patient similar to the literature.

In SMF lesions are usually located usually on the extremities and palms and soles, whereas FMF usually affect head and neck regions [3,9]. Lesions were scattered on the abdomen, arms and legs in our patient, he had no involvement in palms and soles.

Solitary and localized lesions are more frequently seen in SMF than classical MF [6]. There were multiple lesions in our patient similar to the some previous reports [9].

Men are affected more than women [3,5,7,8]. Mean age of the patients was 50-55 years [7,8]. Our patient was male in accordance with the literature. He was at 83 years, with one of the oldest published case at 86-years-old patient from France [4].

Epidermotropic atypical lymphocytes can be seen in the majority of the cases [3,4,6]. Epidermotropism was found is our patient's specimens.

SMF histopathology consists of atypical lymphocytes surrounding eccrine coils. Syringometaplasia is a distinctive feature of SMF [3,4,6]. T lymphocytes can be CD2+, CD3+, and CD4+. In our case, lymphocytes were mostly positive for CD4 and CD7 and less predominantly positive for CD 8 and CD20. In the majority of the cases there are T-cell receptor gene rearrangements that are monoclonal [4]. Histopathologic differential diagnosis of SMF includes perniosis (no syringotropism

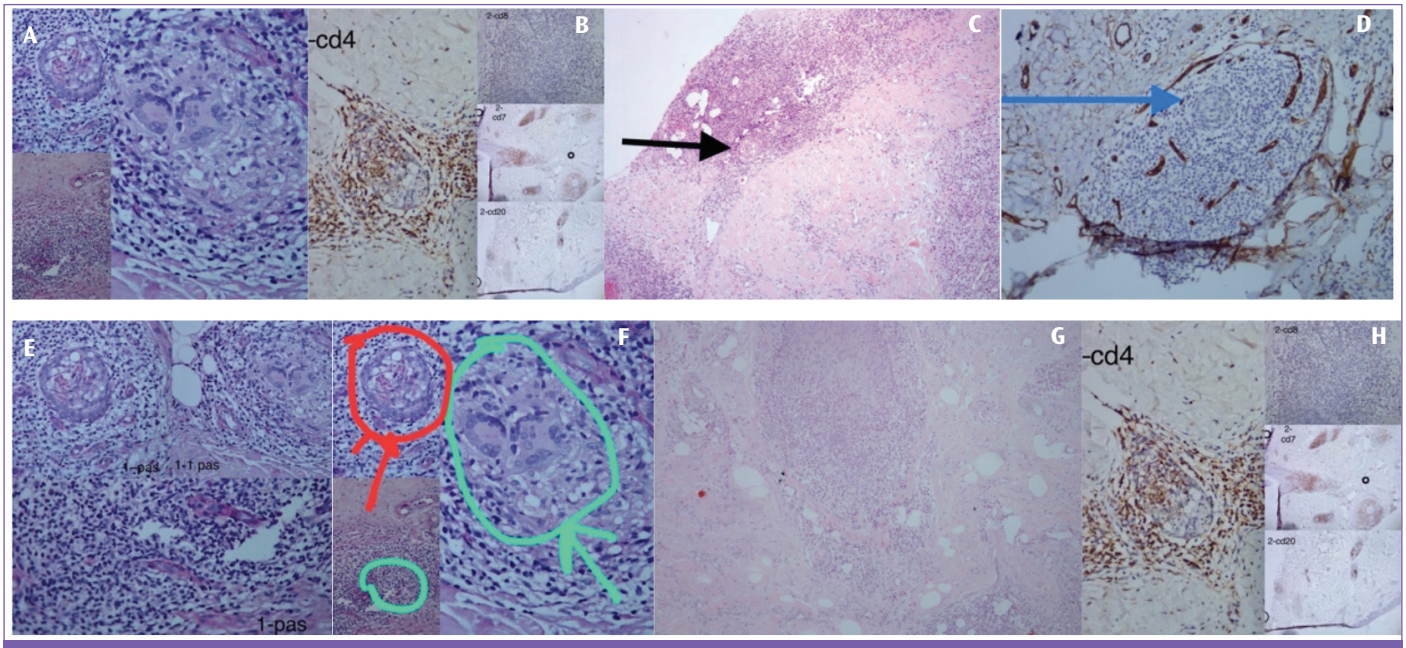

Figure 2. A: Lymphcyte, plasma cell, eosinophil and neutrophil infiltraion (H&E), B: Lymphcytic infiltration involving eccrine glands, C and D: Lymphocytic infiltration involving eccrine glands, E: Granulomatous infiltration and folliculotropism , F: Red arrow: syringometaplasia, yellow arrow: granulomatous infiltration with two multi nuclear giant cells, G: Perifollicular lymphocytic infiltration, H: Perifollicular and follicular lymphocytic. infiltration Mostly positive for CD4, and CD7, Less predominantly positive for CD8 and CD20

and syringometaplasia), neutrophilic eccrine hidradenitis (infiltrate is mostly neutrophilic). Syringometaplasia can also be seen in a number of conditions including skin reactions to chemotherapy, cutaneous ischemia, and radiation dermatitis, in which there were no prominent lymphoid infiltration cutaneous lymphoproliferative T-cell and B-cell disorders other than SMF can also cause hyperplasia of the hair follicles and/or eccrine glands [6]. In classical MF, usually in the tumoral stages, we expect lymphoid infiltrates around the eccrine glands, but syringometaplasia and syringotropism are absent [3,4,6]. Folliculotropism may also be present in some cases [8,10]. In our case, folliculotropism was present.

Distunguishing SMF from FMF is important since their prognosis is different [10]. SMF has better prognosis than its clinically undistinguishable counterpart, FMF [8,9,10]. SMF is a rare disease, thus there is no guideline about its treatment [2,8]. Since the malignant cells are located deep in the dermis, SMF is more refractory to conventional skin directed therapies, such as topical corticostreroids and narrow band UVB, which are usually more effective at classical MF [2,3,9,10]. Radiotherapy is the most effective treatment for local disease [3,8]. Oral retinoids (alitretinoin), interferon alpha, systemic chemotherapy (VELP; vincristine sulfate, etoposide, L-asparaginase, and prednisone acetate), vorinostat, extraphotopheresis, psoralen and UVA are the agents that are used to treat generalised SMF [2,3,8].

In conclusion, SMF is a clinically and histopathologically different variant of MF. It is clinically undistinguisable from FMF, since their prognosis is quite different, they should be differentiated. Clinicians and pathologist should be aware of this entity to prevent delayed treatment.

Ethics

Informed Consent: Informed consent was taken from the patient for possible case report publication.

Peer-review: Internally and externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: F.E., H.E., Concept: F.E., Design: F.E., Data Collection or Processing: F.E., H.E., Analysis or Interpretation: F.E., H.E., Literature Search: F.E., Writing: F.E.

Conflict of Interest: No conflict of interest was declared by the authors.

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