

Effect of Specific Prognostic Markers on Survival and Development of a Prognostic Model in Advanced Stages of Mycosis Fungoides and Sezary Syndrome, by the Cutaneous Lymphoma International Consortium

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Introduction

FM is a rare idiopathic skin condition that typically manifests as infiltrated plaques, nodules, hypopigmented or erythematous patches, and eshcolored follicular papules. Alopecia areata-like symptoms, scarring alopecia, folliculitis, acneiform eruptions, linear lesions that follow Blaschko lines and urticaria-like follicular mucinosis are examples of unusual presentations. In primary FM, the face, head, and neck are frequently affected, whereas the trunk and extremities exhibit extensive lesions. Clinical characteristics, however, are insufficiently specific to discriminate between the secondary form linked to mycosis fungoides and the benign main type. Follow-up is therefore crucial. On the right side of her forehead, a 16-year-old girl had a steadily expanding, erythematous, somewhat irritating patch for the previous two years. The patient's family history and previous medical history were both normal [1,2].

Well-defined, alopecic, and erythematous patch measuring 1 by 2 cm with significant follicular plugs in the middle was discovered during a dermatological examination. Around the lesion, a small ring of mildly hypopigmented skin was seen. A general physical exam revealed nothing unusual. Complete blood count, erythrocyte sedimentation rate, liver and kidney function tests, and urinalysis were among the laboratory tests that all fell within the normal range for. There were no organomegaly, enlarged lymph nodes, or any abnormal signs on the chest x-ray or belly ultrasound. There was a skin punch biopsy done. A portion of the sample that underwent Hematoxylin and Eosin (H&E) staining exhibited a mixed inflammatory infiltration that was primarily made up of lymphocytes and included and surrounded the follicular epithelium as well as a minor focal follicular spongiosis. The mucinous character of the infiltration in the follicular epithelium was revealed by alcian blue staining. The histopathology analysis revealed FM characteristics. An accurate diagnosis of follicular mucinosis was obtained based on the results of the clinical and histological testing [3,4].

Description

Pinkus first identified FM as alopecia mucinosa in 1957. It is a rare idiopathic disorder with an unknown aetiology that mimics folliculitis, alopecia areata, scarring alopecia, chronic eczema, acne, urticaria, and erythrodermic forms by causing mucin deposition within the hair follicles and sebaceous glands

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of the pilosebaceous unit in addition to a superficial and deep perivascular. There are two distinct clinical kinds of dermatosis. Idiopathic (primary or benign) FM is the most prevalent kind and typically affects children and young people. Primary FM, which exhibits spontaneous resolution, is regarded as a temporary kind of disease that often clears up within a few years. Secondary FM, which typically affects older patients and manifests as widespread lesions, is linked to underlying inflammatory and malignant processes, with mycosis fungoides (MF) being the most prevalent malignancy. Despite the fact that Hodgkin disease has been recorded in children and young adults with longer lengths of follow-up, MF is the lymphoma that is most frequently associated with it. The distinction between main and secondary variations must be made using clinical and histological criteria; however these criteria are not sufficiently precise, necessitating patient follow-up. Additionally, it has been suggested that a clonal T-cell receptor gene rearrangement may aid in differentiating FM from FM associated with malignancy. Additionally, it has been observed that there are no definitive criteria for separating idiopathic FM from FM associated with lymphoma, and idiopathic FM may be one of the variant forms of MF that have a protracted, nonaggressive clinical course. Patients with 'idiopathic FM' should be closely monitored over an extended period of time [5].

Conclusion

Since there is no established therapy for primary FM, a variety of anecdotal evidence-based approaches have been tried, including topical, intralesional, and systemic corticosteroids, topical and systemic retinoids, dapson, methotrexate, cyclophosphamide, minocycline, hydroxychloroquine, interferons, indomethacin, topical pimecrolimus, ultraviolet A, superficial radiation The lesion's appearance has improved while our patient is still being monitored clinically thanks to the topical corticosteroid treatment.

Acknowledgement

None.

Conflict of Interest

None.

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