Poikilodermatous Mycosis Fungoides: A Case Report.

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Abstract

Mycosis fungoides is the most common subtype of cutaneous lymphomas. Many clinical variants exist including follicular, ichthyosiform, poikilodermic, bullous, mucinous, granulomatous, pustular and pigmented purpura like lesions. We herein report a case of Poikilodermatous mycosis fungoides in a male patient. Clinical rarity of this entity prompted this communication.

Introduction

Poikiloderma denotes pigmentation, telangiectasia and atrophy of skin. These changes occur in many dermatological disorders like Rothmund Thompson syndrome, Dermatomyositis, Lupus erythematosus and Mycosis fungoides.

Poikilodermatous MF is a rare form of cutaneous T-cell lymphoma that is characterized clinically by localized or diffuse patches, consisting of telangiectasia, mottled hyper- and hypopigmentation, and atrophy. The immunophenotype of neoplastic cells is similar to that observed in classical mycosis fungoides.

Case Report

A 45 yr old male businessman, normotensive, non-diabetic reported to the out-patient department of dermatology, SMHS hospital (associated teaching hospital of Government Medical College Srinagar) with one month history of generalized pruritus and burning sensation associated with dryness of skin and progressive wrinkled pigmentation of trunk and limbs. He also complained of multiple swellings over left side of neck from the same duration, which were insidious in onset, painless and non-tender. He also gave a history of decreased appetite and generalized weakness. The patient?'s medical and surgical history was otherwise unremarkable. There was no history suggestive of photosensitivity. The patient also denied any systemic or topical medication prior to the onset of these lesions.

General physical examination of the patient was normal. Cutaneous examination revealed generalized erythema with scaling over trunk, face and extremities. Poikilodermatous changes were observed, with reticulate pigmentation, telangiectasia, intermingled with atrophic wrinkled scaly hypopigmented macules. These changes were more marked on the anterior chest (Illustration 1), neck, upper back, upper and lower limbs (Illustration 2). Scarring alopecia was present along the scalp margin (Illustration 3) with diffuse scaling of scalp. Nails and mucosae were normal. There was also left cervical lymphadenopathy. The lymph nodes were multiple, firm, discrete, non-tender, mobile, largest being 4 x 4 cm. There? was no axillary or inguinal lymphadenopathy. Systemic examination of the patient revealed no abnormality.

Complete blood count, liver, renal and thyroid function tests were normal. Blood smear for Sezary cells was negative. ANA and anti ds DNA were also negative. Chest X-ray was normal and ultrasonography of the abdomen revealed mild spleenomegaly. Skin biopsy was taken from the back which revealed epidermal atrophy with a band like dermal infiltrate of medium sized atypical lymphoid cells, which show focal epidermotropism (Illustration 4).

Immunohistochemistry was also done revealing CD3+, CD4+, CD45RO+ and CD7- pattern in the dermal infiltrate typical of mycosis fungoides. In view of the clinical, histopathological and immunohistochemical findings in this patient, a diagnosis of Poikilodermatous MF was entertained and the patient was referred to medical oncology for further evaluation and management.

Discussion

Mycosis fungoides represents the most common type of cutaneous T-cell lymphoma(1). Traditionally, it is divided into three clinical stages: patch, plaque and tumour stage. The clinical course can be protracted over a period of years or decades. The aetiology of mycosis fungoides is yet unknown. Genetic predisposition may play a role in some cases. Familial occurrences have been reported in a few instances. Associations with long term
exposure to various allergens has also been advocated, as well as exposure to environmental agents and association with chronic skin disorders and viral infections. In some countries, mycosis fungoides like disorders are clearly associated with viral infections (HTLV-1)(2).

References

Illustrations

Illustration 1

Poikilodermatous changes over neck and anterior chest

Illustration 2

Poikilodermatous changes over lower limbs.
Illustration 3

Scarring alopecia over scalp with poikilodermatous changes over neck.

Illustration 4

H & E staining demonstrating epidermal atrophy, dermal infiltration with atypical lymphocytes and focal epidermotrophism.
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