MYCOSIS FUNGOIDES
(Case reports)

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Summary

Two cases of MF have been reported, one in a middle aged male having MF with erythroderma, poikiloderma and tumours, developing in a short period, and second in a middle aged female having d' emblee form of the disease.

Mycosis fungoides (MF) is an uncommon, chronic, fatal disease of the lympho-reticular system, first described by Alibert in 1806. It occurs in all races, is most frequent between 40 and 60 years, males predominating over females. It begins in a number of different ways but the majority follows definite pattern which has three stages. It remains confined to the skin for many years and in late stages lymph-nodes and internal organs may be involved[1,2].

The initial stage (premycotic) is usually seen as ill-defined erythematous patches of bizarre shape, size and varying colour. Itching is usually severe. Some cases persist for years with little change, but more often, after sometime progress to more infiltrated patches (plaque stage) which are very itchy. The lesions are usually irregular, raised and indurated, some showing ring like pattern. In some patients poikiloderma may replace the early stages of mycosis fungoides but subsequent progress is same. The late stage (tumour stage) sets in with tumour formation and ulceration. Some patients may present with erythrodermia, seborrhoeia, pityriases rosea like lesions poikiloderma atrophicans vasculare[3,4] or the so called tumour D' emblée form first described by Vidal & Brocq[5]. References in Indian literature are very few [6-10]. We report two cases of mycosis fungoides, for the first time from Medical College and Hospital, Rohtak.

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Case Reports

Case 1: A 40 years male presented with complaints of generalised itchy, scaly, red patches, over trunk of 2 years duration and cutaneous swellings on the trunk and extermities of 2 months duration. Treatment from local practitioners gave no relief. The past, family and personal histories were noncontributory.

At the time of admission the patient looked very ill, restless and emaciated, with a pulse rate 116/mt. which was regular and of moderate volume. B.P. 110/80 mm Hg. Body temperature was 101°F, and there was oedema over the feet. The axillary and inguinal glands were slightly enlarged, non-tender and freely mobile. Liver and spleen were not palpable.

The skin was erythematous and scaly with a poikilodermal patch on the chest. (Fig. 1). Telangiectasia in

this patch was prominent. There were many nodular swellings varying in size from a pea to a lemon, scattered irregularly over the trunk and extremities. Tumours over the lower limbs were painful and ulcerated with foul smelling blood stained discharge.

Investigations

Hb, 9 gms%, T. L. C. 11,500/- with normal differential count. E. S. R. 73 mm 1st hour/WG. Peripheral blood film did not show any immature or atypical cells. S. T. S. negative. Serum proteins 5.5 gms%. X-ray chest N. A. D. Urine and stool examination N. A. D: L. F. T. were within normal limits. Pus culture report-E-Coli, when repeated—no pyogenic organism seen.

As the patient's condition was very poor, he was put only on supportive therapy and was given two units of blood. His condition did not improve and he sought discharge from the hospital.

Case No. 2

A 45 years old female presented with multiple swellings all over the body of 2 years' duration. It started with a few lesions on the trunk and severe itching. These nodules gradually increased in size and many new lesions appeared. 20 days before admission some of the lesions ulcerated, discharging foul smelling material. Examination revealed a large number of nodular indurated plaques and vegetating masses distributed all over the body (Fig. 2). They were red to purplish in colour and varied from 1 cm to 8 cm in diameter. The vegetating masses were moist and foul smelling. There was no atrophy or telangiectasia. The axillary and inguinal glands were enlarged on both sides. They were nontender, discrete and mobile. Liver and spleen were not palpable.

Investigations

HB 12 gms% T. L. C. 11,300/cmm with normal differential count. No immature or atypical cells were seen in the peripheral blood film. E. S. R. was 15 mm 2 hour (W. G.): S. T. S. was negative. Urine and stool examinations were normal. X-Ray chest-N. A. D. On the fifth day of admission patient left the hospital against medical advice.

Histopathology

Skin biopsies were done from tumors in both the patients and chest lesion (poikilodermic patch) of case No. 1: Histopathology of tumours in both the cases was similar showing irregular acanthosis of the epidermis, basal layer destroyed at places, and mononuclear cells invading the epidermis. There was tendency to form micro-abscesses in the deeper layers of epidermis (Fig. 3). The superficial and middermis showed pleomorphic inflammatory infiltrate dominated by histiocytes and lymphocytes. Occasional large mononuclear cell with darkly stained large nucleus (Mycosis cell) was seen (Fig 4).

Skin biopsy from chest lesion of case No. 1 showed atrophy of the epidermis and flattening of the rete pegs with degeneration of basal layer at places. The dermis showed diffuse chronic inflammatory infiltrate with increase of capillaries many of which were dilated. The upper dermis showed incontinence of pigment and melanophages. Lymph gland biopsies were not done in either case.

Comments

M. F. is considered to be a type of reticulosis which begins in the skin and usually confines to it for months to years. Systemic involvement is considered to be rare, though it has been reported to be frequent in the tumour stage of the disease (Post and Lincoln, loc, cit; Block et al[11]; Domonkos, loc cit. and Singh and Shah, loc cit).

The histopathologic changes were typical as described by Lever[12], (Mycosis cell). The sections from the chest

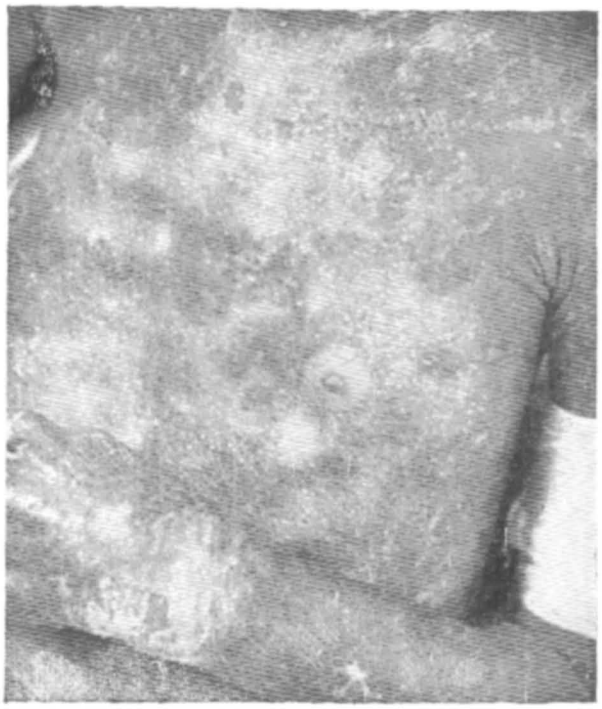

Fig. 1 Skin of the chest showing atrophic patches, erythema, telangiectasia and scales.

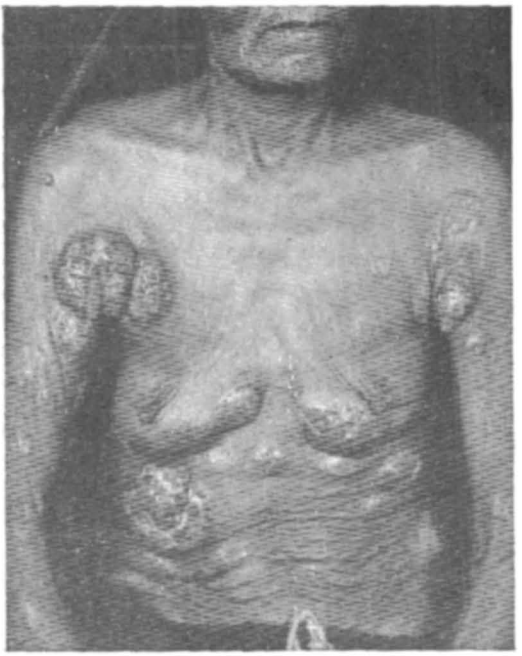

Fig. 2 Nodules and infiltrated plaques of d'emblee type of M. F. are seen.

Fig. 3 Irregular acanthosis of epidermis, with basalcell degeneration; Dermis heavily infiltrated with chronic inflammatory cells, invading the epidermis, with a tendency to form pauterier's microabscesses.

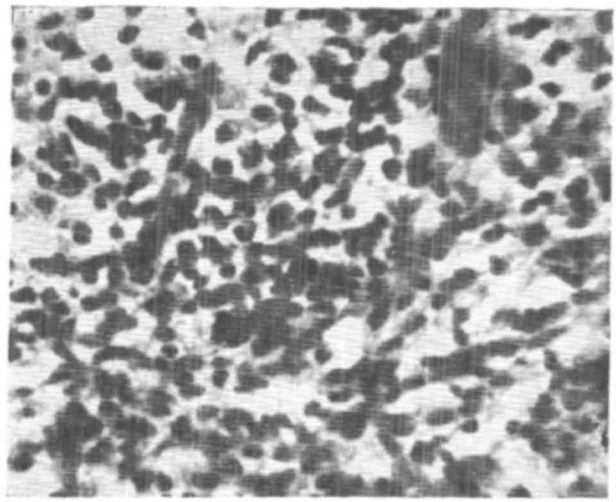

Fig. 4 High Power of part of Fig. 3, showing Mycosis cells.

skin biopsy of case No. 1 were suggestive of poikiloderma.

The aetiology of this disease remains undefined. There are controversies as to whether M. F. is a distinctive pathological entity separable from other lymphomas. Tan et al[13] and Rona et al[14] have reported elevated levels of Ig E immunoglobulin. Abnormal lymphocytes with T-cell properties[15],[16] have been described.

Greim et al[17], Long and Mihm[18] and many others have reported visceral involvement as a relatively frequent occurrence in Mycosis fungoides. Laparotomy done in these cases have shown involvement of liver, lymph-nodes, bone marrow and spleen. In our cases investigations could not be carried out to study any possible visceral involvement. Value of lymph-angiography and laparotomy has been suggested by Greim et al[17], Long and Mihm[18] and many other workers to assess the stage of the disease and for proper planning of treatment. Laparotomy as a routine diagnostic procedure probably is not advisable in all cases.

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