MYCOSIS FUNGOIDES
(A case report)

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Summary

A case of mycosis fungoides is presented. The unusual features observed in this case were occurrence at a relatively young age, starting with a tumour stage and a possible involvement of the liver.

Introduction

Mycosis Fungoides was first described by Alibert in 1806. A member of the lymphoma group, it has certain characterstic clinical features by which the diagnosis can often be strongly suspected before the histological proof is available[1]. The disease occurs usually in adults and mostly in males[2]. It usually evolves through three stages namely; (1) Premycotic stage characterised by intense pruritis, poikiloderma parapsoriasis, exfoliative dermatitis and eczematoid dermatitis. (2) Infiltrative stage where the infiltration in the skin gives rise to erythroderma and histopathologically non-bacterial microabscesses known as Pautrier's abscess and (3) Tumour stage characterised by development of nodules of various sizes over the infiltrated plaques as well as over the healthy skin, with a tendency to break down and form ulcers. The diagnosis is confirmed by histopathological examination. The disease runs a variable course and ends fatally. Response to treatment is usually unsatisfactory.

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Case report

A 21 years old Hindu male was admitted with the complaints of nodular swellings all over the body of two and a half month's duration. The nodules when first noted in the umbilical region were small in size and non-tender. After few days nodules appeared in the neck, extremities and axillae and gradually increased in size (Fig. 1 Page 110). Patient gave history of irregular fever of two and a half month's duration. Few weeks prior to admission the patient started feeling weak with loss of appetite and weight.

On examination he was found to be febrile with pulse of 129/mt. There were multiple, purplish nodular swellings of various sizes on the skin mostly of the abdomen, back, extremities, neck and axillae. The size of the nodules varied from a pea to that of a lemon. The skin over the abdominal swelling was dusky red in colour and marks of itching were present. The nodules were tender, their consistency varying from soft to firm. Feet were odematous. Cervical, axillary and inguinal lymphnodes were enlarged, non-tender, and soft to firm in consistency. Skin over the lymphnodes was

smooth and shiny. Patient was anaemic. There was no sternal tenderness, purpuric spots, bleeding spots or ulcers in the oral cavity. Tonsils were not enlarged. Liver was enlarged to 4 c. m. below the costal margin smooth non tender and soft to firm in consistency. There was no splenic enlargement. Respiratory, cardiovascular and central nervous systems were normal.

Laboratory examination revealed the total leucocyte count to be 11,000/-cumm, neutrophils 58%, lymphocytes 40%, eosinophils 2%, erythrocyte sedimentation rate 42 mm in one hour, hemoglobin 8 gm% and R. B. C. count 4.2 millions. Peripheral smear of blood showed marked hypochromia, polychromasia, aniscocytosis and poikilocytosis. Platelet distribution was scattered. Direct platelet count was 1.2 lacs. No premature cells or parasites were seen in the peripheral smear. Urine examination was normal. Bone marrow was normal. X-ray chest was normal.

Nodule biopsied from the abdominal region showed reticulum cells and lymphocytes which were infiltrating the fatty tissue and fibrous tissue (Fig. 2 Page No. 110). This histology was consistent with that of tumour stage of mycosis fungoides.

Patient was treated with 200 mg of endoxan intravenously on alternate days for about a fortnight without any apparent improvement. He was discharged against medical advice and could not be followed up further.

Discussion

Sequeira[3] in his series of 74 cases reported that the disease occurred commonly in males and between 30–50 years of age. Our case was 21 years old. Singh and Shah[4] have reported a case of mycosis fungoides in a 19 years old Hindu male.

The disease is known to run an insidious course. Nonspecific skin lesions like eczema, parapsoriasis, erythroderma, exfoliative dermatitis and poikiloderma may occur for as long as 27 years[5]. In 13% of cases the disease may start with a tumour stage[6] and the skin nodules are usually not tender[2]. Our case presented for the first time in the tumour stage. The distribution of the skin nodules was mainly over the trunk. Some of these were tender and showed signs of inflammation. However aspiration from the nodules showed no pus.

Lymphnode involvement is an early feature. They are characteristically non-tender, firm and freely mobile[6,7]. Bone marrow involvement occurs in about 18% of cases[6]. Involvement of viscera e.g. liver, heart, kidney, lungs, gastro intestinal tract and central nervous system is an occasional feature[4]. Liver enlargement was seen in our case but histopathological study could not be done as patient refused to have a liver biopsy. Anaemia, eosinophilia, leucocytosis and abnormal lymphocytes have been reported. In our case anaemia was present whereas lymphocytosis and abnormal lymphocytes were conspicuously absent.

Osborn et al[8] noted a dramatic response to Nitrogen mustared in two very advanced cases but it was temporary. Apart from nitrogen mustard and cyclophosphamide, methotrexate, vinblastine sulphate, actinomycin–D, azotepa and streptonigrin have been found to be effective[9]. Various forms of treatment varying from topical application of Fluocinolone acetonide[10] to systemic administration of cytotoxic drugs have been tried.

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TRUE

The condition of EFA deficiency was first observed and reported by Burr and Burr in 1929 who described the scaly skin condition and impaired rate of growth of rats maintained on fat-free diets. Sinclair showed that EFA deficient rat skin was abnormally permeable to H_2O. Van Drop in 1974 reported a reduction of the abnormally high rate of transepidermal water loss in deficient rats by rubbing esters of linoleic and arachidonic acid on the skin. Since then several workers have shown that cutaneous application of sunflowers seed oil (rich in the triglyceride form of linoleic acid) correct high rate of transepidermal water loss in EFA-deficient skin thus restoring it to normal appearance.

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