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SEZARY SYNDROME

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A case of Sezary syndrome in a 56 year old female who presented with generalised erythroderma associated with pruritus and lymphadenopathy of 2 years duration is described. The disease was diagnosed by the presence of Sezary cells in the skin biopsy and peripheral smear. The patient was referred to cancer institute for further management.

Key Words : Sezary Syndrome, Exfoliative Dermatitis

Introduction

Sezary syndrome is an uncommon form of cutaneous T cell lymphoma which is considered to represent the leukemic stage of Mycosis fungoides. It was first described by Sezary and Bouvrain in 1938 in a patient with erythroderma, intense pruritus, adenopathy and abnormal 'monster' hyperconvoluted mononuclear circulating cells in the peripheral blood. The less constant features include cutaneous oedema, alopecia, onycho dystrophy, palmar & plantar keratoderma, hepatomegaly and lymphadenopathy.[1]

The aetiology remains doubtful though environmental factors and retroviral (HTLV - I) infections have been suggested. Sezary syndrome differs from other forms of leukemia in that the sezary cells do not originate from the bone marrow which is in fact normal in appearance. The exact origin of sezary cells is not clarified though skin and lymph node have been suggested, the later being more appropriate as radioisotope scanning studies have suggested that these cells migrate from blood into the skin.[2]

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Case Report

A 56-years-old female was suffering from generalised erythroderma with intense pruritus since 2 years. Initially the lesions were localised to medial aspect of right upper thigh which gradually spread to involve other sites within 2-3 months. She was diagnosed to have exfoliative dermatitis else where and treated with corticosteroids which did not show complete remission at any time. With repeated episodes of such rashes she developed hyperpigmented patches, erosions, plaques, nodules and ulcers, crusting and oozing. She had alopecia and transverse ridging nails, pigmentation of proximal nail plate and onychomadesis. There was generalised lymphadenopathy with discrete, non tender, firm, mobile and moderately enlarged lymphnodes. (Fig. 1 & 2)

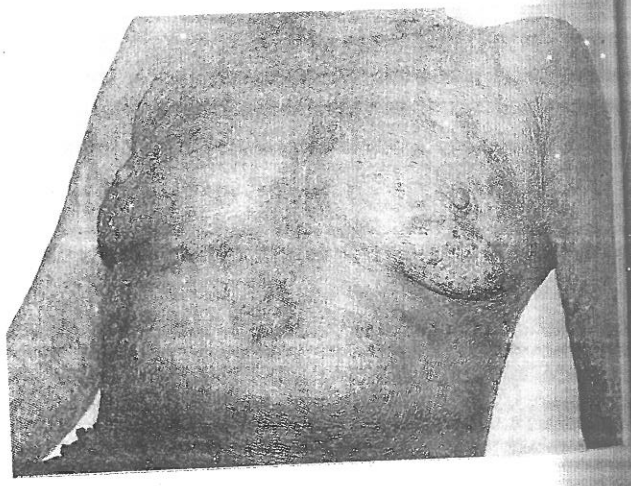

Fig. 1

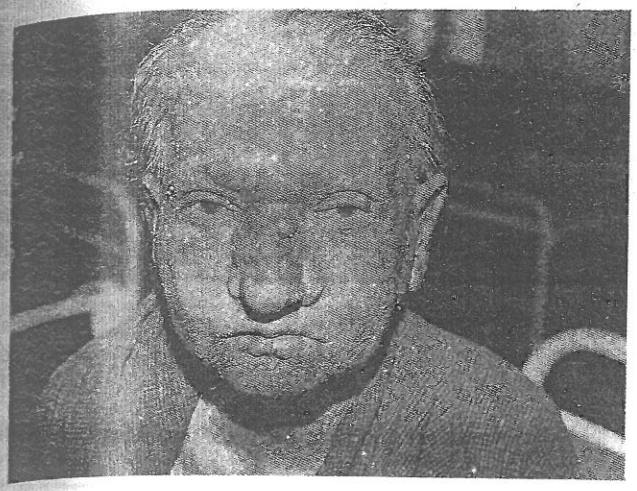

Fig. 2.

Haematologic investigations revealed haemoglobin 8 gms%, PCV 22%, ESR 80mm/Ist hr. The total leucocyte count was 23,200 cells/cμmm with lymphocytosis. Atypical lymphocytes having convoluted nuclei, coarse chromatin and inconspicuous nucleoli with scanty basophilic cytoplasm were 46%.[3] Other parameters were normal.

Periodic acid Schiff (PAS) stain showed PAS positive granules[4] in the atypical lymphocytes of peripheral smear. (Fig. 3) Skin

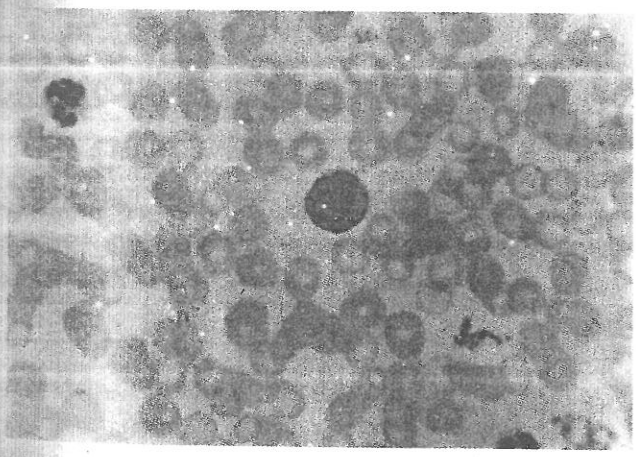

Fig. 3.

biopsy showed lichenoid infiltrate of lymphoid cells in the upper dermis many of which were large with hyperconvuluted nuclei admixtured with small lymphoid cells. (Fig. 4 & 5).

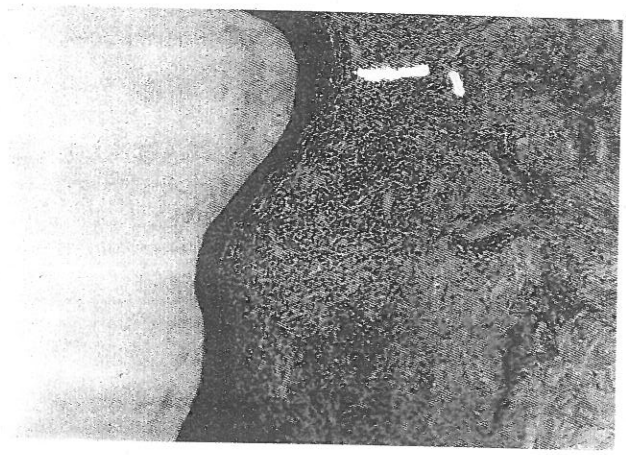

Fig. 4.

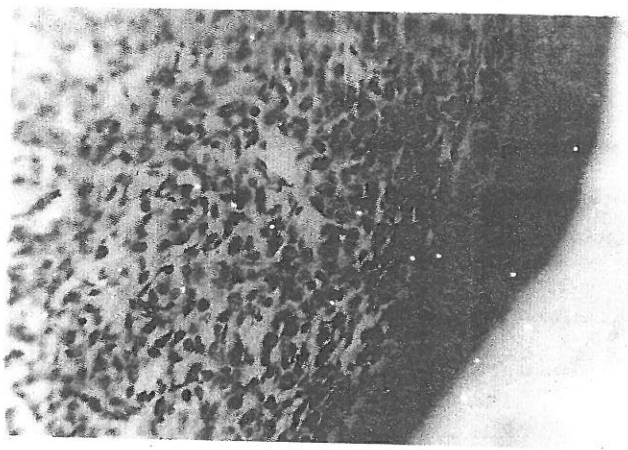

Fig. 5.

Comments

Sezary syndrome remains a challenging disease in terms of its proper diagnosis, classification and treatment. The prognosis for patient with sezary syndrome is poor. The best estimates of survival are 2.5 to 5 years. It is notorious[1] for its resistance to various treatment options, the most reasonable being extra corporeal photopheresis, chlorambucil and prednisolone or low dose methotrexate.[5] Interferron, electron beam irradiation, monoclonal antibodies, cyclosporine and other immunostimulants have been effectively tried.

The appearance of cells in the peripheral smear and in the skin biopsy of our case corresponded to that described in sezary syndrome. The patient was lost for follow up as she was referred to cancer institute.

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