CASE REPORTS

SEZARY SYNDROME IN A YOUNG WOMAN

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A young female patient suffered for 4 months from a widespread erythrodermic rash of unknown origin, marked by a peculiar hyperaesthesia. Haematological and biopsy findings were initially non-specific. A rapid deterioration, accompanied by infiltration of the skin, lymphadenopathy and hepatosplenomegaly was suggestive of Sézary syndrome, which was confirmed by finding of 40% of the lymphocytes being atypical (Sézary cells).

Key Words : Erythroderma, Sézary cells, Sézary syndrome

Introduction

The Sézary syndrome is diagnosed by the presence of the triad of erythroderma, lymphadenopathy and more than 10% of the circulating peripheral lymphocytes being atypical. It is most often seen in elderly males and has a poor prognosis.

We present an unusual case of typical Sézary syndrome developing de novo in a young female patient with a rapid evolution over a few months and a fatal outcome.

Case Report

A 35-year-old housewife presented with an innocuous, though widespread, skin rash which was red and scaly, lasting for four months. This was associated with persistent swelling of the face and extremities. A peculiar finding was the constant presence of generalised hyperaesthesia; she complained of a burning sensation all over the body with discomfort on palpation. She also suffered from low grade fever, malaise, anorexia and weight loss.

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Three and a half years prior to this, she had received a course of anti-tuberculous drugs for a cervical swelling. At that time, she had

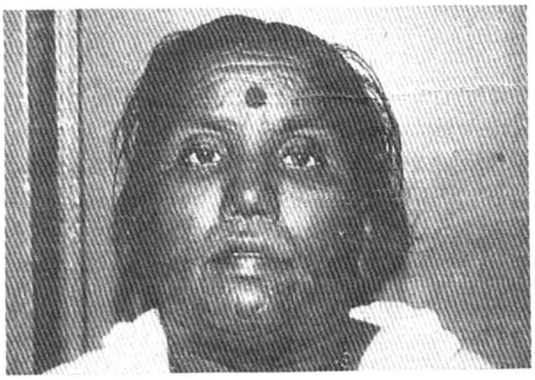

Fig. 1. Diffuse thickening and infiltration of the face giving a 'leonine' appearance.

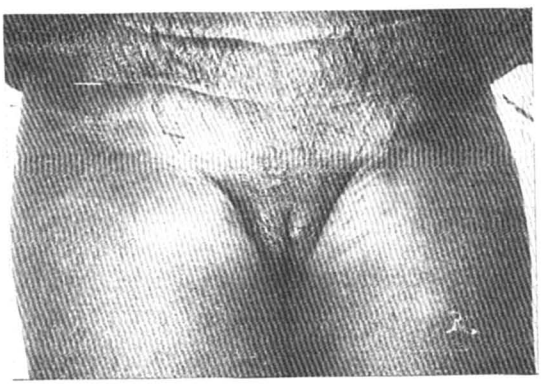

Fig. 2. Infiltration of body folds and striae gravidarum on a background of erythroderma.

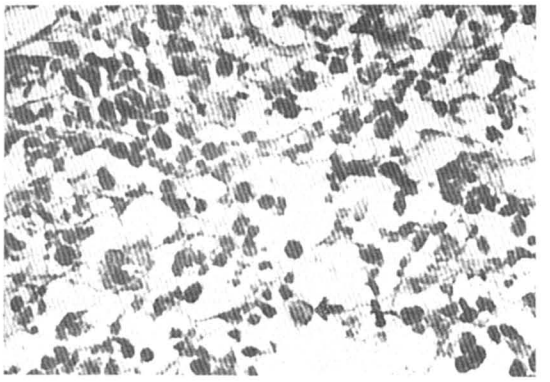

Fig. 3. Histopathological section showing abnormal lymphocytes (40X).

also complained of generalised 'pruritus' unresponsive to treatment which, however, subsided spontaneously. She was asymptomatic during the intervening three years.

On examination, the patients was febrile. The rash was generalised with an erythematous hue and fine scaling. The skin appeared thickened on palpation. Marked hyperaesthesia was a distinctive feature, but no peripheral nervous system abnormality was detected. Generalised lymphadenopathy and moderate hepatosplenomegaly were also noted.

Investigations at this point revealed anaemia (Hb 9g%). The WBC count was 5200/cmm, comprising 63% PMNs and 27% lymphocytes; 50% of the lymphocytes were atypical. Skin smears for M. leprae were negative. Skin biopsy showed a non-specific histology.

She was treated symptomatically. There was no improvement and in fact, she deteriorated over the next one month. The infiltration increased significantly leading to leonine facies, nodulation of the ears, prominence of all body folds as well as striae, and succulence of fingers and toes. The skin developed a deeper erythematous hue and was now scaly, warm and tender. Diffuse alopecia was seen. The lymph nodes enlarged to form visible swellings in the axillae and groins. An increase in the size of liver and spleen was noted.

Reinvestigation showed the haemoglobin to be the same but the total count had risen to 12,400/cmm, with a neutrophilia of 80%. Of the total lymphocytes 40% were atypical. Large cerebriform nuclei with a scanty cytoplasm confirmed these to be Sézary cells on electron microscopy. Marker studies for T-lymphocytes showed a helper-suppressor ration of 9:1 (normal 2:1).

The skin biopsy, this time, was dramatically different and revealed a cutaneous lymphoma with mild epidermotropism. The lymph nodes and bone marrow showed no malignant change. Enlarged liver and spleen with normal echo-texture were visualised on ultrasonography of the abdomen. Having confirmed a diagnosis of Sézary syndrome, the patient was treated with a regimen of cyclophosphamide, vincristine and proednisolone, to which methotrexate was added. However the patient showed only a temporary response. Her condition progressively worsened and she eventually succumbed to the disease after 8 months.

Discussion

Sézary syndrome presents with the classic triad most commonly in elderly males. It may develop ab initio or following mycosis fungoides.[1-4]

The condition, which was earlier thought to be neoplastic from the outset, is now considered to be a malignant transformation of a chronic immunological process, involving the interaction of T-lymphocytes and Langerhans cells.[2] The resultant release of circulating vasoactive lymphokines and macrophage inhibiting factor is responsible for the erythroderma in this condition.[3]

Erythrodermas of short duration and uncertain aetiology in young adults are not uncommon, and rarely necessitate a search for an underlying malignancy. In the absence of any preceding premycotic lesion, the skin rash in the present case was hardly indicative of Sézary syndrome, especially in its early stage. However, the presence of hyperaesthesia and burning sensation, as against the expected pruritus, was a distinctive finding and an indicator to the diagnosis.

References

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