Sezary’s syndrome: A case report

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ABSTRACT
A case of Sezary’s syndrome in a Saudi patient is reported. The patient presented with generalized erythroderma. Abnormal lymphoid (Sezary) cells were seen in the peripheral blood, bone marrow and skin biopsy. Its relationship to mycosis fungoides is discussed.

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Sezary’s syndrome is a rare disease. It was first described by Sezary and Bouvrain in 1938 as generalized exfoliative dermatitis with atypical lymphocytes in the peripheral blood. Since then, it has become a well known clinical entity characterized by erythroderma and abnormal lymphoid (Sezary) cells in the peripheral blood. This disorder is closely related to mycosis fungoides and is considered by some authors to be the peripheral blood manifestation of the disease. To the best of our knowledge this is the first case report of Sezary’s syndrome in a Saudi patient.

Case Report. A 56 year old Saudi male presented to the Riyadh Medical Complex with generalized erythroderma associated with severe pruritus. The patient had these skin lesions for 4 months. He had had two cardiac valvular replacements 15 and 5 years previously and had a pacemaker. He had been on warfarin 9.5 mg daily since that time. Family history was not contributory. On physical examination the patient was well nourished with a temperature of 37°C and the BP was 120/80 mm Hg. On examination he had diffuse erythema with scratch marks involving mainly the face, trunc and upper limbs (Fig. 1). Lichenified eritematous rash was seen in proximal parts of lower limbs while the distal parts showed erythema with petechial rash. He had firm, palpable and non-tender lymph nodes involving bilaterally the axillary, inguinal and right epitrochlear areas. Liver was palpable 3 cm below the costal margin but no splenomegaly was noted. Computed tomography (CT) scan of the abdomen and pelvis were negative for lymph nodes. Laboratory results showed WBC 25,000 x 10^3/ul with lymphocytes 44%; LDH 1359 U/L. Peripheral blood examination revealed 19% of atypical cerebriform lymphoid cells (Sezary’s cells) (Fig. 2). The diagnosis of Sezary’s syndrome was further established by bone marrow examination and skin biopsy.

Microscopic findings. Histologic examination of the skin showed a band-like infiltrate in the upper dermis made up by mixed populations, of histiocytes, lymphocytes and atypical hyperchromic lymphoid cells. Some of these atypical cells revealed indented nuclei with cerebriform appearance. The epidermis was infiltrated by these cells singly as well as in the form of small aggregates appearing as Pautrier micro-abscesses (Fig. 3). The bone marrow examination revealed cellular marrow with scattered atypical lymphoid cells with irregular indented nuclei identified as Sezary cells.

Comment. T-cell lymphomas are relatively rare neoplasms comprising of 5% of Non-Hodgkin’s lymphomas; however, these are fairly common in Japan. All the lymphoproliferative disorders involving the skin and characterized by cells with deeply convoluted nuclei on E.M. are grouped under one name, “Cutaneous T-cell lymphomas” (CTCL). These include mycosis fungoides, Sezary’s syndrome, actinic reticuloid, pagetoid reticulosis and lymphomatoid papulosis. Only a few T-cell lymphomas have been recognized as clinicopathologic entities: mycosis fungoides, Sezary’s syndrome, and T-cell lymphoblastic lymphomas. The remaining T-cell lymphomas are a heterogeneous group, clinically, morphologically and immunologically. Mycosis fungoides and Sezary’s syndrome are the most widely recognized members of this group. They are usually grouped together as peripheral T-cell lymphomas, though they usually present with lymphadenopathy, extranodal association is common, notably the skin being the most frequent (75%) site of involvement. Prognosis is poor in most of the cases; however, some long survivals have been reported. Mycosis fungoides, a chronic lymphoma of the skin, usually evolves through three stages: a premalignant stage with lesions similar to eczema or
psoriasis, an infiltrative or plaque stage, sometimes with generalized exfoliative erythroderma and invasion of the blood by atypical convoluted neoplastic lymphoid cells (the so-called Sezary's cells), and a nodular or tumor stage associated with a deeper invasion by the tumor and infiltration of lymph nodes and other organs.

Sezary's syndrome is essentially a leukemic phase of mycosis fungoides. It is uncommon and is characterized by generalized erythroderma, severe itching, hyperkeratosis of palms and soles, splenomegaly, superficial lymphadenopathy and atypical cells in the circulating blood, cutaneous infiltrate as well as in the bone marrow. Although the epidemiology of Sezary's syndrome is not well known, overall most patients with CTCL are between 15 and 70 years of age at diagnosis (median age 63 years) with a 1.5:1 male predominance. Various exposures to physical agent toxins and chemicals have been proposed to be etiologically responsible for the development of CTCL and presumably Sezary's syndrome.

A causative role of retroviruses have also been postulated. In our case, no hyperkeratosis of the palms or soles nor splenomegaly were present. The peripheral blood revealed leukocytosis and atypical lymphoid cells with the distinctive deeply convoluted nuclei (Sezary's cell). Histopathologically it revealed a subepidermal band of lymphocytic cells mixed with histiocytes and some Sezary's cells. These cells also were infiltrating the epidermis and producing microabscesses of Pautrier. The atypical lymphoid (Sezary's) cells are present in the skin, blood and sometimes in the lymph nodes and bone marrow. In our case, these cells were seen in the skin, peripheral blood and the marrow. Morphologically, Sezary's cells are characterized by high nucleocytoplasmic ratio, cerebriform nuclei with fine chromatin pattern and scanty cytoplasm. Immunological and functional studies of the Sezary's cell in the blood and in the cells of the skin lesion have surface markers for T cells, usually CD4 subtype. A wide variety of treatment options exist for Sezary's syndrome. These include electron beam irradiation, chemotherapy, PUVA, leukophoresis, antithymocyte globulin, monoclonal antibodies or other immune stimulants, retinoids, cyclosporine, interferon and extracorporeal photopheresis. The patient was initially treated with prednisone (60 mg daily) and cyclophosphamide with good response. In summary, we have reported a rare case of Sezary's syndrome in a Saudi patient.

References