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 relation to other lymphomas was discussed.

Keywords: Sezary’s syndrome, erythroderma, bone marrow, cutaneous lymphoma, skin biopsy.

Sezary’s syndrome - a rare disease. It was first described by Sezary and Rappaport in 1938 as a generalized cutaneous dermatosis with abnormal 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 Sézary’s syndrome 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 symptoms for 4 months. He had had a varicose vein operation 5 years previously and had a sebaceous. He had been on warfarin 50 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 abdominal examination he had diffuse edema with scratch marks involving the face, trunk and lower limbs and a 3 cm right axillary lymph node. On chest examination the patient was asymptomatic and no abnormalities were noted. Computed tomography (CT) scan of the abdomen and chest was negative for lymph nodes. Laboratory results showed WBC 5,000 x 10^9/L with lymphocytes 75%. DD 1359 11:14. Peripheral blood examination revealed 16% of atypical erythrodermic lymphoid cells (Sezary syndrome). The diagnosis of Sezary’s syndrome was further established by bone marrow biopsies. Microscopic findings. Histopathological examination of the skin showed a band-like infiltrate in the upper dermis made up of mixed populations of histiocytes, lymphocytes and atypical hyperchromatic lymphoid cells. Some of these atypical cells revealed indented nuclei with a cerebriform appearance. The epidermis was infiltrated by these cells singly as well as in the form of small aggregates appearing as Pautrier microabscesses in Fig. 2. The bone marrow examination revealed cellular marrows 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. As the lymphoproliferative disorders involving the skin and characterized by cells with deeply atypical nuclei, in cutaneous T cell lymphomas (CTCL). These include mycosis fungoides, Sézary’s syndrome, acanthosis nigricans, pseudolymphoma, and lymphomatoid papulosis. Only a few T cell lymphomas have been recognized as morphologically distinctive mycosis fungoides, Sézary’s syndrome, and T cell lymphomatous lymphomas. The remaining T cell lymphomas are a heterogeneous group, clinically, morphologically, and immunologically. Mycosis fungoides and Sézary’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 involvement is common, notably the skin being the most frequent 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
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

A causative role of retroviruses have also been postulated. In our case, no hyperkeratosis of the palms or soles nor splenomegaly was present. The peripheral blood revealed 1% 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 chromatm 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