

Fiery erythema in Sézary's Syndrome and an overview of the disease

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Abstract:

Sézary'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 atypical cells in the circulating blood, producing a cutaneous infiltrate as well as being present in the bone marrow.

Although the epidemiology of Sézary's syndrome is not well known, most patients with CTCL are between 15 and 70 years of age at the time of diagnosis (median age 63 years) and there is a 1.5:1 male predominance.

Case Report

A 70-year old Iranian caucasian male agricultural worker presented to the dermatology clinic with generalized erythroderma associated with severe painful pruritus and with several palpable masses in the axillae and in the cervical region. He had been applying clobetasol daily since the condition began six months before. No one else in the family was affected.

The patient was well nourished with a temperature of 37°C and blood pressure 120/80 mm Hg. He had diffuse scaling and pronounced erythema of the skin with scratch marks due to the severe recalcitrant pruritus especially on the arms, with milder lesions on the head, face and trunk (Figs. 1, 2, 3,4).

The skin of his extremities was bronzed with elastosis and cutis rhomboidalis of the face, hands, and neck suggesting prolonged exposure to sunlight. Lichenified fiery erythematous rashes were present on the proximal parts of his lower limbs while the distal parts showed fiery erythema with fissuring and severe exfoliative scaling. Bilaterally there were firm,

palpable, mobile, painless lymph nodes in the axillae, inguinal and cervical regions especially the left epitrochlear areas. The liver was not palpable below the costal margin, the spleen was not enlarged but there was periumbilical tenderness. Abdominal sonography showed no abnormalities of liver, spleen or pancreas.

Routine laboratory tests showed WBC 20,400,000/ul; lymphocytes 66%, polymorphonuclear granulocytes 26%, monocytes 1%, eosinophils 3%, basophils 1%; Hb 14.9 gm/l, HCT 44.6% and platelets 186,000,000/ul. Examination of stained peripheral blood films showed 16% of atypical cerebriform lymphoid cells (Sézary cells) plus Lutzner cells (Fig.4).

The diagnosis of Sézary syndrome was confirmed by a skin biopsy which showed a band-like infiltrate in the upper dermis composed of mixed accumulations of histiocytes, lymphocytes and atypical hyperchromatic lymphoid cells some of which had serpiginated nuclei with a cerebriform appearance. The epidermis also was infiltrated by these cells singly and as small aggregates or Pautrier microabscesses. A diagnosis of mycosis fungoides was considered. Histopathology of lymph nodes revealed complete effacement of the nodular architecture by diffuse proliferation of the mature lymphocytes and subcapsular sinuses filled with lymphocytes. There was very little mitotic activity and mycosis fungoides cells were not seen. These findings corresponded to Sézary syndrome, a type of lymphocytic lymphoma (Fig. 5 & 6).

Discussion

Most patients with Sézary's syndrome are elderly males who develop the syndrome either ab initio or following lesions considered to be classical mycosis fungoides⁽¹⁾. Our patient was an elderly farmer with prolonged exposure to the sun. In his case the first signs were generalized fiery erythema with severe pruritus and cervical and axillar lymphadenopathy. Abnormal lymphoid (Sézary) cells were present in his peripheral blood and in three skin biopsies.

T-cell lymphomas are relatively rare neoplasms comprising 5% of non-hodgkin's lymphomas although these are fairly common in Japan {2}. The relationship of Sézary's syndrome to other actinic disorders such as actinic reticuloid and mycosis fungoides was reviewed and it was found that all lympho-proliferative disorders involving the skin and characterized by cells with deeply convoluted nuclei on electron microscopy (EM) are grouped as cutaneous T-cell lymphomas (CTCL). These include mycosis fungoides, Sézary's syndrome, actinic

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Fig. 1



Fig. 2



Fig. 3



Fig. 4

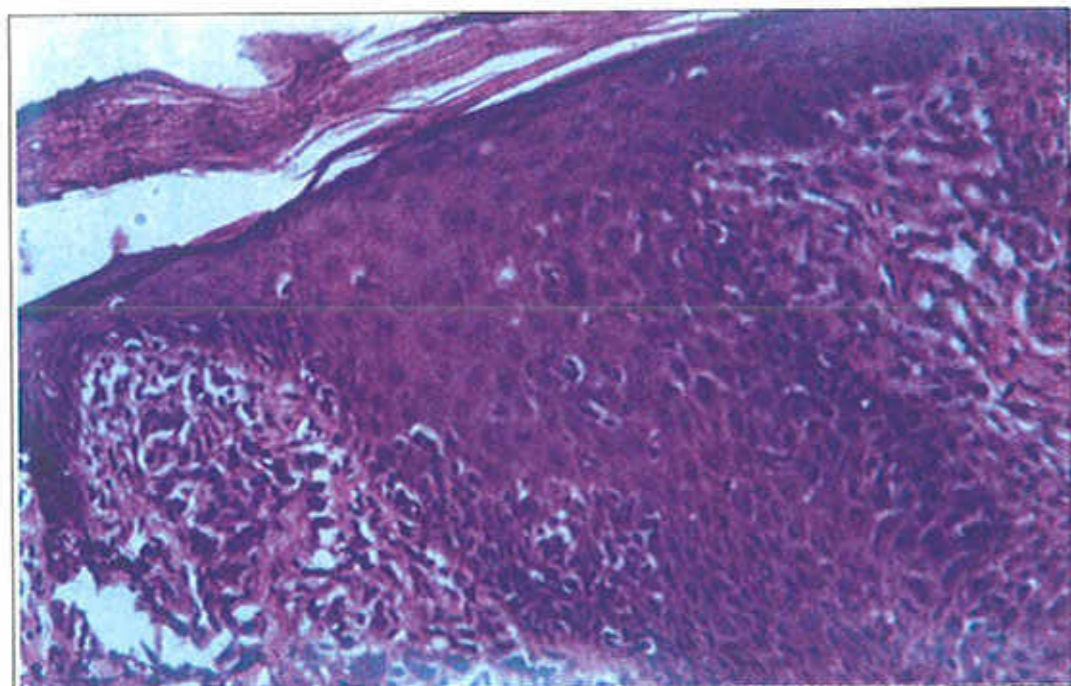


Fig. 5 Cut section of SS skin

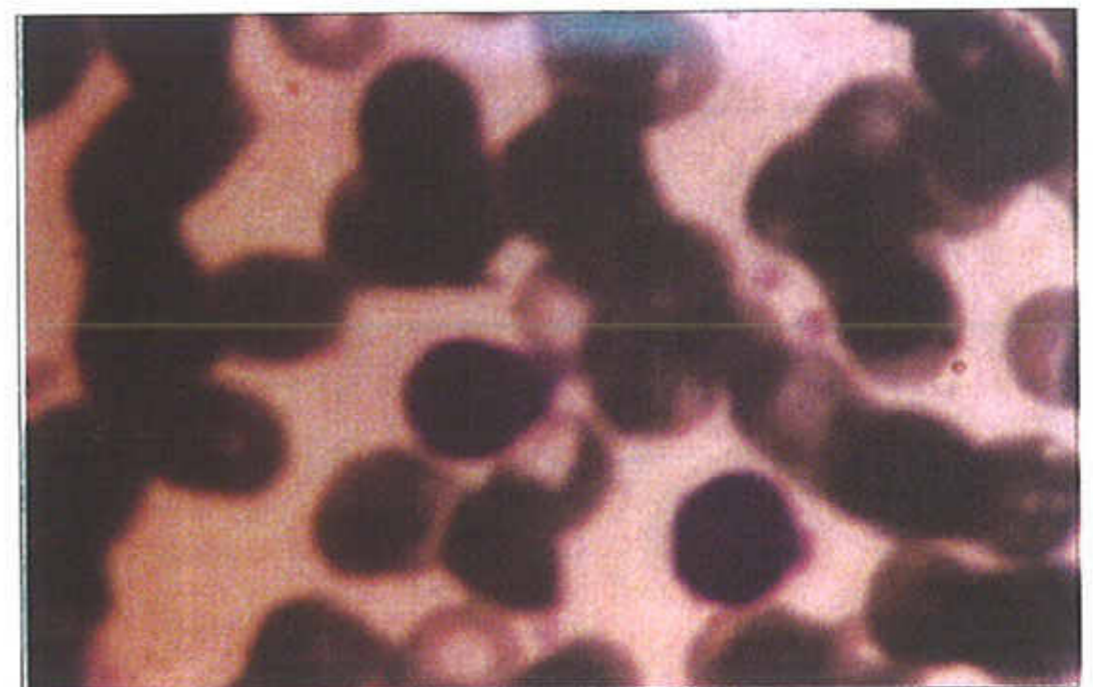


Fig. 6 Sezary cell in SS

reticuloid pagetoid reticulosis and lymphomatoid papulosis¹¹⁻³¹. Only a few T-cell lymphomas have been recognized as clinico-pathologic entities: mycosis fungoides, Sézary's syndrome, and T-cell lymphoblastic 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 of these syndromes. Myelodysplastic syndrome has been reported in association with mycosis fungoides and Sézary's syndrome^{14, 5, 61}. Although they usually present with lymphadenopathy they are commonly grouped together as peripheral T-cell lymphomas. Extranodal associations are common, the skin being the most frequent (75%) site of involvement.

Prognosis is poor in most of the cases although some long survivals have been reported. Mycosis fungoides, a chronic lymphoma of the skin, usually evolves through three stages: a premycotic 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 Sézary's cells) and a nodular or tumor stage associated with a deeper invasion by the tumor and infiltration of lymph nodes and other organs¹⁷¹.

Sézary'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 atypical cells in the circulating blood, producing a cutaneous infiltrate as well as being present in the bone marrow.

Although the epidemiology of Sézary's syndrome is not well known, most patients with CTCL are between 15 and 70 years of age at the time of diagnosis (median age 63 years) and there is a 1.5:1 male predominance. Exposure to various physical agents, toxins, and chemicals has been proposed as respon-

sible for the development of CTCL and presumably Sézary's syndrome. A causative role for retroviruses has also been postulated¹⁸¹.

In our case the hyperkeratosis and fissuring of the palms and soles were severe but splenomegaly and hepatomegaly were not present. The peripheral blood revealed leukocytes and atypical lymphoid cells with the distinctive deeply convoluted nuclei (Sézary's cell). (Fig.4). Histopathology of the skin revealed a subepidermal band of lymphocytic cells mixed with histiocytes and some Sézary's cells. These cells also were infiltrating the epidermis and producing microabscesses of Pautrier.

Morphologically, Sézary's cells are characterized by a high nucleocytoplasmic ratio, cerebriform nuclei with a fine chromatin pattern and scanty cytoplasm and they are present usually in the skin, circulating blood and sometimes in the lymph nodes and bone marrow. In our case these cells were seen in the skin, peripheral blood and the lymph nodes. Sézary's cells in the blood and in the cells of the skin have surface markers for T-cells, usually CD4 subtype.

A wide variety of methods are available for the treatment of Sézary's syndrome including electron beam irradiation, chemotherapy, PUVA, leukopheresis, antithymocyte globulin, monoclonal antibodies or other immune stimulants, retinoides, cyclosporine, interferon and extracorporeal photopheresis¹⁹¹. Our patient was treated initially with 50-mg prednisone daily and cyclophosphamide with a moderate response. Local corticosteroids (clobetasol) with 5% salicylic acid in "Vaseline" oil were used for the severe exfoliation and fissuring of palms and soles.

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References:

- 1- MacKie R.M. *Lymphomas and Leukemias (Sézary's syndrome)*. RH Champion JL Burton; FJG Ebling in text book of dermatology 5 the edition Blackwell Scientific Publication vol 3 1992; 2131-33.
- 2- Shimizu M., Miura M., Kitahara M., Katayama I., Non Hodgkin, S lymphoma. Analysis of 109 Japanese cases with the use of LSGJ classification. *AM J Pathol* 1982; 106:30-39.
- 3- Weis JW, Wintner MW, philyky RL et al. Peripheral T-cell lymphoma. Histologic, immunohistologic and clinical characterization. *Mayo clinic Proc.* 1986; 61:411-426.
- 4- Shirley JA, Sparrow GP. Sézary syndrome associated with sideroblastic anemia. *Scand J Haematol* 1979; 23:373-7.
- 5- Rostoker G., Raphael M., Boissnick S., et al. Coexistence of Sézary syndrome and dysmyelopoiesis with an excess of myeloblasts [letter]. *J Am Acad Dermatol* 1986; 15:1296-8.
- 6- Pablo F. Penad, MD, Maria Jones-Calallero, MD, Esteban Dauden MD, Javier Fraga, MD and Amaro Garcia-Diez MD. Cutaneous T-cell lymphoma and myelodysplastic syndrome *Am Acad Dermatol* 1994 vol 31-6 1065-7.
- 7- Mohammed Afzal, MD, FCAP, Adnan S. Abdolkareem, MD, Kaniz Rasul, MBBS M. Phil, Atilio Baez-Giangreco, MD FCAP. Sézary's syndrome a case report. *Saudi Med. J* 1997; vol.18(5):519-20.
- 8- Wieselthier JS, Koh HK. Sézary's syndrome. Diagnosis, prognosis and critical review of treatment options. *J AM Acad Dermatol* 1990; 22:381-401.
- 9- Arnold Harry L., Odom Richard B., James William D., *Mycosis fungoides. Other malignant lymphomas and allied diseases in Andrews, diseases of the skin clinical dermatology 8 edition* 1990; 863-64.