Circulating Sezary cells in subacute spongiotic dermatitis

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ABSTRACT

Sezary syndrome comprises a triad of fiery red erythroderma, generalized lymphadenopathy, and circulating atypical cells with cerebriform nuclei (Sezary cells) exceeding an absolute value of 1000/cumm or exceeding 10% of circulating cells. Sezary cell is not pathognomonic of Sezary syndrome and can be seen in many benign dermatoses such as chronic eczema, psoriasis, lupus erythematosus, parapsoriasis, atopic dermatitis, and vasculitis. We report circulating Sezary cells in a patient of subacute spongiotic dermatitis, one of the rare causes for Sezary cells in peripheral blood.

Key words: *Peripheral blood, Sezary syndrome, Subacute spongiotic dermatitis*

Since its first description by Lutzner and Jordan in 1968,[1] Sezary cell has been the center of an ongoing controversy concerning the specificity of its association with cutaneous T-cell lymphoma. Subsequent studies have identified Sezary cell as a thymus-derived T- lymphocyte.[2] Sezary cell is not pathognomonic of Sezary syndrome and can be seen in many benign dermatoses such as chronic eczema, psoriasis, lupus erythematosus, parapsoriasis, atopic dermatitis, and vasculitis.[3]

We report circulating Sezary cells in a patient of subacute spongiotic dermatitis, one of the rare causes for Sezary cells in peripheral blood to create awareness that a physician and pathologist should maintain an index of suspicion for the disease, by performing a skin biopsy, and vigilantly following up the patient to effectively treat these patients.

CASE REPORT

A 60-year-old male presented to the medicine OPD of Indira Gandhi Medical College, Shimla, with complaints of itching, redness, and scaling all over the body for 4–5 months, which gradually spread to the trunk and upper limbs.

On the examination, vitals were within normal range. There was diffuse erythema, hyperpigmentation, increased skin marking with large yellowish scales crusting all over the trunk, along with red, round, itchy, and non-oozy painless scaly lesions over both legs (Fig. 1). Axillary and inguinal lymphadenopathy were noted, the largest inguinal lymph node being 1 × 1 cm. No hepatosplenomegaly was seen.

The liver function tests and blood glucose levels were within normal limits. A complete hemogram showed total leukocyte count 13,500/mm^3 while differential count revealed eosinophilia. The buffy coat smears revealed the presence of atypical mononuclear cells (Sezary cells) 35/100 lymphocytes (Fig. 2). Flow cytometry showed the CD4:CD8 ratio to be 3.5:1. Fine-needle aspiration cytology was performed from the enlarged inguinal lymph nodes. Cytomorphology revealed non-specific reactive hyperplasia. Biopsy from the axillary lymph nodes showed features of dermatopathic lymphadenitis. The skin biopsy was done which revealed hyperkeratosis, parakeratosis containing plasma cells, acanthosis, and lymphocytic infiltrates in the lower epidermis. Papillary dermis showed edema, lymphohistiocytic aggregates, melanophages, and fibrosis (Fig. 3). The final diagnosis of subacute spongiotic dermatitis with circulating Sezary cells was thus rendered.

The patient was treated with methylprednisolone 16 mg orally for 10 days followed by 8 mg orally for 10 days and 4 mg orally for the next few days. The improvement was noted in the patient; lately, dermatitis was cleared and these cells were no longer seen in the peripheral blood smear.

DISCUSSION

Sezary syndrome is characterized by the triad of erythroderma, lymphadenopathy, and circulating atypical mononuclear cells (Sezary cells).[4] Sezary cells are lymphocytes with hyperconvulated or cerebriform nuclear contours. Morphologically, they are characterized by high N:C ratio, deep, and narrow nuclear indentation, condensed chromatin at the nuclear membranes and cytoplasm poor in organelles.[5]

The involvement of the peripheral blood in mycosis fungoides has been shown to be a significant prognostic factor.[6] However, morphologically, similar cells can be found in the peripheral

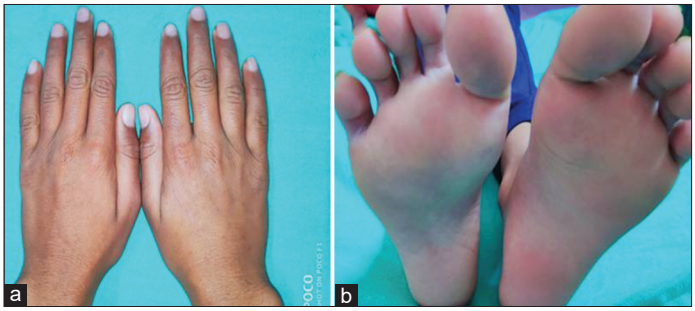

Figure 1: Photograph of the patient showing erythromyelgia on the (a) hands and (b) and feet

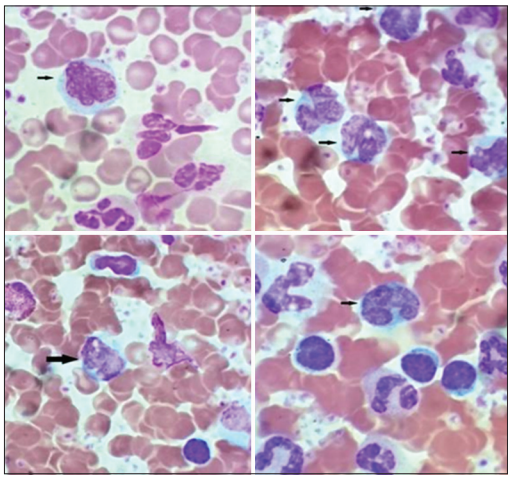

Figure 2: Photomicrograph showing Sezary cells (black arrows) with eosinophils. (×100; Giemsa)

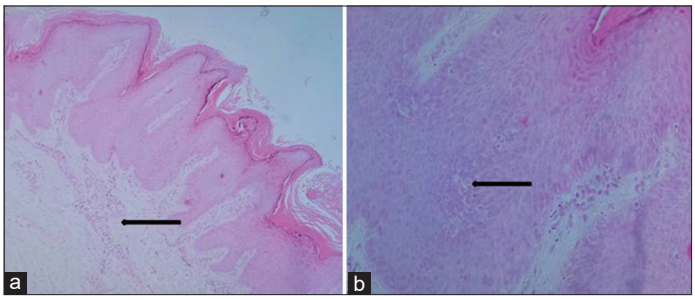

Figure 3: Microphotograph from the skin biopsy showing (a) hyperkeratotic, acanthotic, and stratified squamous epithelium with mild lymphocytic infiltrate (black arrow) (H&E; ×100); (b) hyperkeratotic, acanthotic, and focal spongiotic changes (black arrow) stratified squamous epithelium (H&E; ×400)

blood of patients with a variety of non-neoplastic dermatosis,[7,8] indicating the reactive T-lymphocytes can sometimes closely resemble their malignant counterparts.

The non-neoplastic conditions, in which peripheral blood mononuclear like Sezary cells are reported to, include actinic reticuloid, actinic keratosis, Dilantin, hypersensitivity syndrome, discoid lupus erythematosus, erythrodermic follicular mucinosis, lichen planus, psoriasis, and vasculitis. Duncan and Winkelman[9] reported Sezary cells in circulation in 14 hospitalized dermatology patients: Six with contact dermatitis and three with exfoliative psoriasis, with an absolute count of Sezary cells being more than 1000/mm^3.

The question of whether the Sezary cell is a neoplastic cell, or merely, a reactive cell has not been settled. The appearance of this cell in patients with the non-neoplastic disease, in cultures of normal skin and pokeweed or phytohemagglutinin-stimulated lymphocyte cultures from healthy human beings points to a non-neoplastic etiology.[10] This is also supported by the presence of circulating lymphokines in patients with Sezary syndrome. Flaxman *et al*. reported[11] the presence of Sezary cells in cutaneous lesions of a variety of inflammatory dermatoses, solar-induced skin disorders (e.g., solar keratoses and basal cell carcinoma), and vasculitis.

Furthermore, the independent observations by Yeckley *et al*.[12] and Van-Leuwen *et al*.[13] have suggested an alternative nature of Sezary like cells. Using phytohemagglutinin, a plant mitogen which stimulates T-cells, these groups showed a morphologic change from normal mature lymphocytes to cells with Sezary like morphology in peripheral blood lymphocytes, produced by *in vitro* stimulation. This suggested that Sezary like cells found in patients of contact dermatitis, in cutaneous infiltrates in a reactive lymph node, normal spleen, and even in normal blood, are merely reactive T-cells, and do not represent a morphologically distinct skin-associated T-cell subpopulation.

Winkelman and Hoagland,[14] in a study on 96 patients with an original diagnosis of mycosis fungoides, revealed 12 patients with circulating Sezary cells, only three had more than 15% of such cells. These 12 patients had benign and malignant skin diseases. The Sezary cell does not seem to have any importance in the pathogenesis of mycosis fungoides.

The clinical presentation of Sezary syndrome is non-specific erythroderma, described as diffuse, bright red with scaling which may be accompanied by fever chills, loss of weight, malaise, insomnia secondary to overwhelming pruritis, and poor temperature hemostasis. There may be scaling, nail dystrophy, and ankle edema.[15] The case under study also had long-standing erythroderma, itching, redness, and scaling all over the body.

The diagnosis of Sezary syndrome rests on the characteristic hematologic picture. There is, usually, a moderate leukocytosis ranging between 10,000 and 30,000/mm^3 with a marked lymphocytosis and sometimes eosinophilia.[16] Our case showed evidence of leukocytosis with eosinophilia and approximately 35% cerebriform cells in the peripheral blood.

It would thus appear that the Sezary cell is not specific for any single disease process. Sezary's cells are characteristically found in the peripheral blood smear of patients with Sezary's syndrome and in malignant, premalignant, and benign dermatoses. Patients may give a history of pruritic, inflammatory patches, or plaques present for several years, even decades. The biopsy in these cases is performed to further define the process if it is the early manifestation of Sezary syndrome or Sezary cells associated with benign dermatosis.

CONCLUSION

Sezary's cells are characteristically found in the peripheral blood smear of patients with Sezary's syndrome and in malignant,

premalignant, and benign dermatoses. In our patient, we found that Sezary's cells were no longer demonstrable when the dermatitis was cleared. The treating physician should maintain an index of suspicion for the disease, by performing a skin biopsy and vigilantly following up the patient to effectively treat them.

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