

Sezary syndrome in a young retropositive male: A rare case report

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ABSTRACT

Sezary syndrome, a rare disease, is the leukemic counterpart of mycosis fungoides accounting for less than 5% of cutaneous lymphomas. Very few case reports have been published of Sezary syndrome/mycosis fungoides presenting in young male and with coexisting HIV. We present a case of a 23-year-old retropositive male presenting with Sezary syndrome which is very rare. The present case highlights the fact that Sezary syndrome can rarely present in young and retropositive patients. It should be kept in differential diagnosis if a patient presents with erythroderma, generalized lymphadenopathy and characteristic peripheral smear findings. A multimodal approach involving flow cytometry, skin biopsy and fine needle aspiration cytology (FNAC) is required for arriving at a definite diagnosis.

Keywords: Erythroderma, HIV, Lymphoma, Mycosis Fungoides, Sezary.

Sezary syndrome includes three signs: erythroderma, generalized lymphadenopathy, and presence of Sezary cells in the skin, lymph nodes and peripheral blood. One of the following is also needed for making a diagnosis: an absolute Sezary cell count >1000/microlitre, CD4: CD8 ratio of >10 and loss of one or more T cell antigens [1]. Cutaneous lymphomas are as such rare, amongst which Sezary syndrome has a very low incidence constituting less than 5% of them. It presents in elderly patients with a median age of 60 years with a male preponderance [1]. Very few case reports of Sezary syndrome/mycosis fungoides presenting in young male and with coexisting HIV have been published [2-5]. It is one of the rare HIV associated neoplasms. We present a young retropositive male who was diagnosed with Sezary syndrome.

CASE REPORT

A 22-year-old retro-positive male was presented with complaints of multiple erythematous papular lesions, first appearing on the trunk and spreading to both upper and lower limbs over a span of seven days. The patient did not have fever, vomiting, photosensitivity or any drug intake.

On examination, numerous elevated reddish papular lesions measuring 5mm were present all over on the trunk, upper and lower extremities. There was widespread scaling which was found in the various sites of the body (Fig. 1) and the patient also had significant lymph nodes (submental, bilateral submandibular, jugular, axillary, inguinal). With the above findings in a retropositive male, a provisional diagnosis of immune response inflammatory syndrome was made.

A routine CBC was done which helped to arrive at the diagnosis. The peripheral smear showed raised WBC count of 30,000/mm³ with increased lymphocytes (80%), an absolute lymphocyte count of 24,000/mm³. Also, 34% of these lymphoid cells (8160/mm³) were found to have characteristic cerebriform nuclei (Fig. 2). To confirm the diagnosis, flow cytometry of peripheral blood was performed which revealed a moderately bright lymphoid cluster on CD45/SSC analysis comprising of 56% of the total acquired events. Total 98% of these cells were found to be of T lymphoid type on CD19/SSC analysis. These cells showed heterogenous positivity for CD5, CD3 (dim to moderate) and were found to be negative for CD7. Also, 97% of these cells were positive for CD4, indicating a significantly raised CD4: CD8 ratio (Fig. 3).

Hence, based on these immunophenotypic findings a diagnosis of T cell lymphoproliferative disorder, possibly Sezary syndrome, was rendered and both skin biopsy and FNAC from lymph node were advised. Skin biopsy showed an infiltrate of atypical lymphoid cells abutting the epidermis having indented nuclei with the presence of characteristic Pautriers microabscess within the epidermis. The mid dermis also had a nodular perivascular infiltrate of similar cells (Fig. 4). The cells showed immunopositivity for CD3, CD4 and were immunonegative for CD8. FNAC smears from both cervical and axillary lymph nodes revealed infiltration by atypical cells, many with characteristic indented cerebriform nuclei.

DISCUSSION

Cutaneous T cell lymphomas are a rare composite group including mycoses fungoides, primary cutaneous CD30 positive T-cell lymphoproliferative disorders, primary cutaneous peripheral T



Figure 1: (1a) Raised popular erythematous lesions on upper limbs and trunk, (1b) Scaling over the lesions

cell lymphomas, rare subtypes, most common being mycoses fungoides [1]. When these cells appear in the blood, it amounts to Sezary syndrome and it can arise de novo too [6]. As stated previously, it is very rare and uncommon in young individuals, as in our case, which was only 23 years old. It is more common in males as was seen in our case too [1]. Sezary syndrome can involve any organ, most common being oropharynx, lungs and CNS [1]. Bone marrow involvement is variable and was uninvolved in our case [1]. In addition to the classical presenting features, there can be pruritus, alopecia, ectropion, palmoplantar hyperkeratosis and onychodystrophy [1] but none of these were seen in our case.

Cutaneous T cell lymphoma is rare in HIV positive patients as compared to B cell Non-Hodgkin lymphomas. In a 3-year prospective study of 1161 HIV positive patients, Munoz MA *et al* found only two cases of mycosis fungoides [7]. Kerschmann *et al* in their study found only 8 cases of mycosis/sezary in HIV positive patients out of 25 cases with non-Hodgkin lymphoma [8]. Esteve *et al* could also found only one case of mycosis amongst

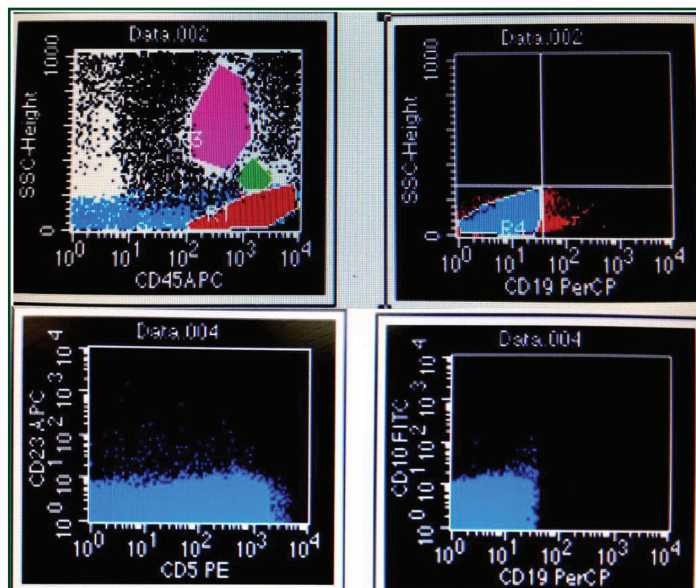


Figure 3a: Flow cytometry showing a bright lymphoid cluster with majority CD19 negative and CD 5 positive

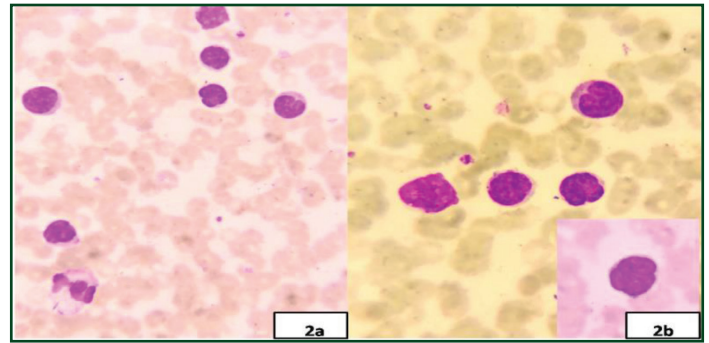


Figure 2: (2a) Peripheral smear showing lymphocytosis, 200X, Giemsa stain, (2b) Peripheral smear showing characteristic sezary cells, 400x, Giemsa stain, Inset: Sezary cell showing cerebriform nucleus, 1000x, Giemsa stain

8 cases of cutaneous lymphomas in retropositive patients [9]. The association of Sezary with HIV is rarer; on an extensive search, we could only find two reported cases of Sezary syndrome associated with HIV [4,10].

Etiopathogenesis of this syndrome remains unsolved. Many viruses like Human T cell Leukemia virus -1, Ebstein Barr virus, have been implicated as causative agents. Some authors have suggested the pathogenic pathway involved in epidermotropic nature of the disease [11]. According to them, there occurs increased production of IL-1 by the keratinocytes, which in turn leads to an elevated expression of intercellular adhesion molecule -1 on both endothelial cells as well as on keratinocytes. This leads to increased binding of the atypical helper T cells to these keratinocytes as well as endothelial cells [11].

Diagnosis of Sezary syndrome rests on identifying the pathognomic sezary cells in the peripheral blood. These cells have a highly indented nucleus and resemble cerebrum hence, called cerebriform. An absolute count of >1000/ mm³ of Sezary cells is required for diagnosis, in our case, it was 8160/microlitre [1].

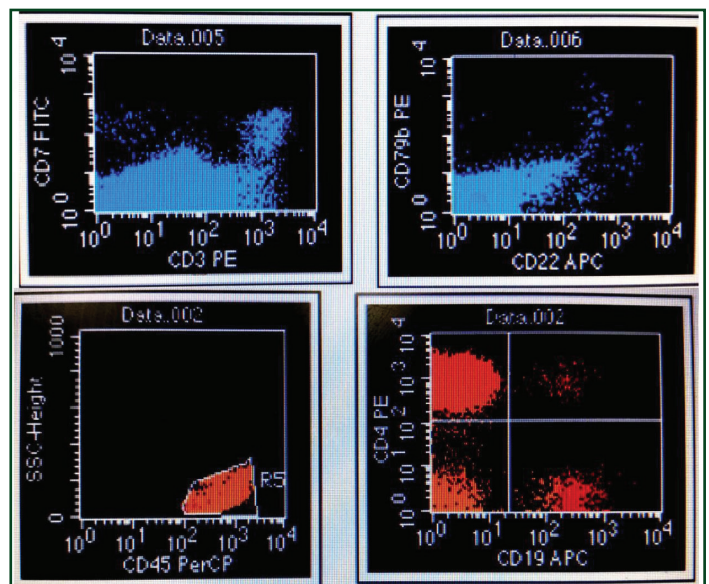


Figure 3b: Flow cytometry showing lymphoid cluster with positivity for CD 3, CD4 and negativity for CD 7

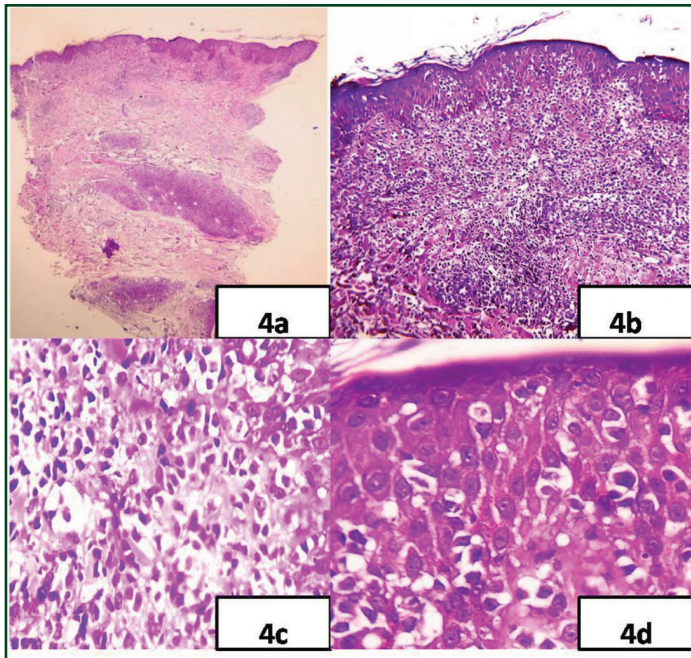


Figure 4: Skin biopsy report (4a) epidermotropic as well as dermal infiltrate, 40x, H&E, (4b) epidermotropic infiltrate of atypical cells, 100x, H&E, (4c) atypical cells in the dermis, 400x, H&E, (4d) pustular microabscess in the epidermis, 400x, H&E

Immunophenotypically, these are helper T cells which show loss of T cell antigens. Characteristically, these are negative for CD7 and CD26 [1]. In our case too, these cells were immunopositive for CD3, CD4, CD5 and immunonegative for CD7.

Skin biopsy is another very important modality with regard to diagnosis and further characterization. As noted in our case, it revealed the characteristic histopathological features of mycosis fungoides. However, in some cases of Sezary syndrome, the features can be very subtle and hence many sequential biopsies may be required to arrive at the diagnosis [12]. At times, the TCR gene rearrangement analysis may be needed to establish the diagnosis.

Treatment of Sezary syndrome is palliative and is stage dependent. Skin-directed therapies, biological response modifiers, and chemotherapy are the mainstays of available options. Biological responses modifiers are under experimental trial are only used along with the topical therapies. Chemotherapy is used for advanced cases, the most common being CHOP regimen [12]. Our patient got discharged to take a referral from another center and hence, could not be followed up. Although mycoses fungoides has an indolent course, the prognosis of Sezary syndrome is worse with a median survival as low as three to four years [11].

CONCLUSION

Sezary syndrome is rare in HIV positive patients and in young males. As highlighted by our case, it should be kept as a differential

diagnosis in retropositive young males who are presented with generalized lymphadenopathy and skin rash. A simple peripheral blood examination can give an early clue to the diagnosis which can be further confirmed by flow cytometry, skin and lymph node biopsy with immunohistochemistry.

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