Generalised pruritus and lymphocytosis in skin of colour

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Case History

A 59-year-old male, originally from Cote d'Ivoire, presented with an incidental new lymphocytosis (>20x10^9/L), no cytopenias and a three year history of generalised pruritus. He reported light patches of skin on his torso, arms and face. He remained systemically well with no B-symptoms.

Medical history included chronic hepatitis B and there were no regular medications.

Examination findings

- Generalised mottled hyper and hypopigmented patches and xerosis on type VI skin **(Figure.1).**
- Scaling on the torso, well-demarcated patches on the abdomen but no papules or plaques **(Figure. 2).**
- Palpable cervical and axillary lymphadenopathy.
- No ectropion, nail changes or alopecia.

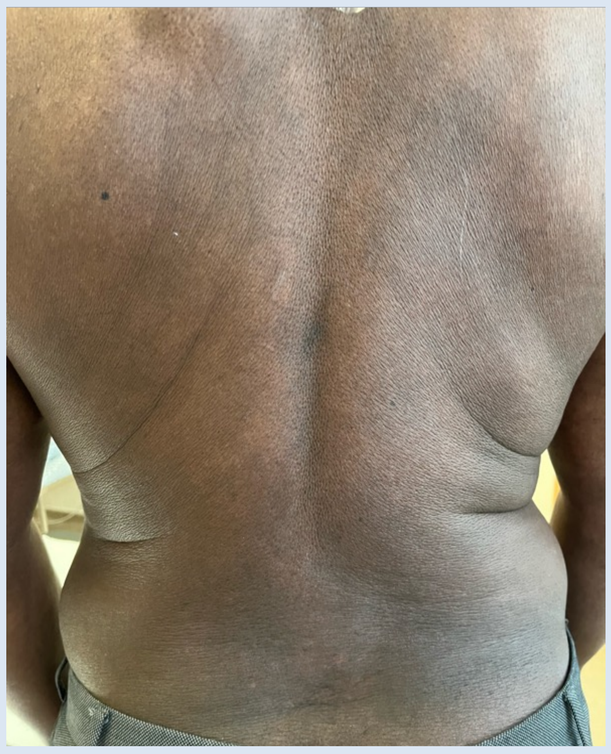

Figure 1

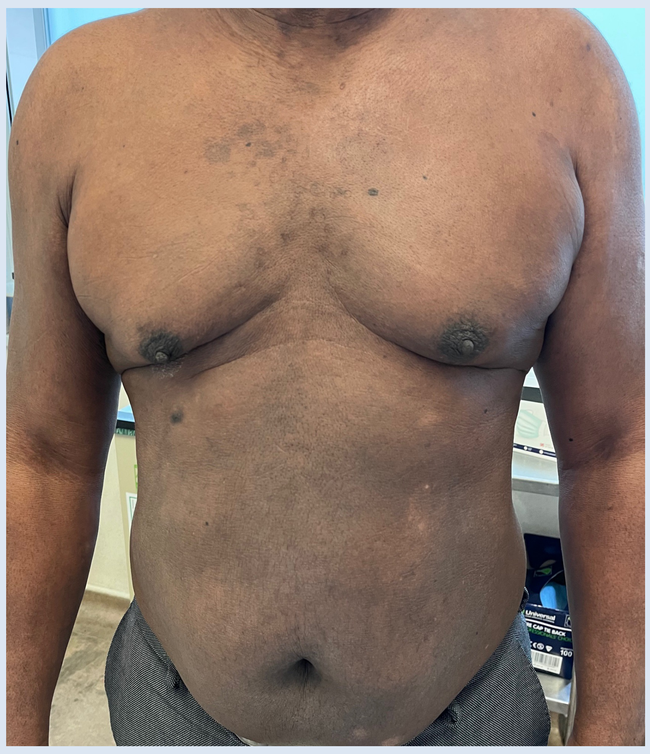

Figure 2

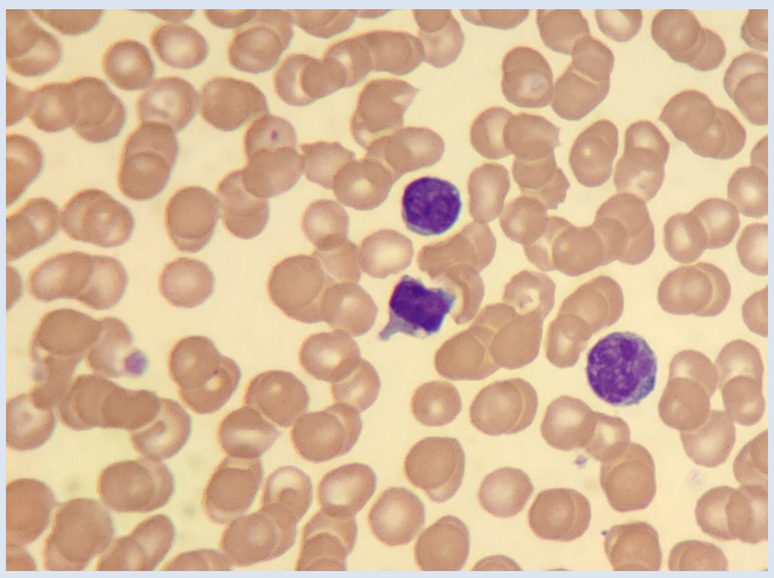

Figure 3

Investigations

Haematology

Blood film

- small lymphocytes with mature, clumped chromatin and a thin rim of basophilic cytoplasm with conspicuous blebbing.
- Occasional forms with cerebriform nuclei were noted (Sezary-like cells) **(Figure. 3).**

Peripheral blood immunophenotyping

- expanded population of CD3+/CD4+ T-cells displaying low CD45 expression and downregulated Pan-T-cells markers.
- HTLV1/2 serology was negative.

Bone marrow immunophenotyping

- expanded population of CD45dim/CD3+dim/CD4+ T-cells that co-expressed Pan-T-cells markers CD2, CD5, CD7.
- CD26 was negative.

Bone marrow trephine

- hypercellularity and increased number of CD4+ T-cells.

TCR rearrangement studies confirmed T-cell clonality.

Histopathology

Skin biopsies demonstrated lymphocytes with irregular nuclear enlargement in the epidermis and formation of Pautrier micro-abscesses. A lymphocytic infiltrate with atypical and irregular lymphoid cells was apparent in the dermis with immunohistochemistry revealing a pan-T-helper phenotype (CD2, CD3, CD4 and CD5 positive). ~1% of the cells in the infiltrate were CD30 positive.

Radiology

A PET-CT revealed focal cutaneous activity in the abdominal wall and avidity in bilateral axillary, distal external iliac and inguinal nodes.

Diagnosis

Presumed diagnosis is Sezary syndrome with leukaemic involvement

Management

Topical mometasone improved pruritus and Tenofovir was commenced for Hepatitis B prior to systemic treatment for presumed Sezary syndrome, pending staging with ultrasound of the PET-avid nodes

Conclusions

- The cutaneous signs were subtle in our case, however clinical and dermatopathological correlation were instrumental in narrowing a wide differential of lymphoproliferative disorders from haematological investigations.
- The lack of erythroderma, as well as CD7 positivity, are atypical.
- Challenges in recognising erythroderma in darker skin, coupled with unfamiliarity of varied cutaneous T-cell lymphoma presentations in skin of colour, may have contributed to a delayed presentation.
- Mycosis fungoides/Sezary syndrome has a higher incidence in Black patients and is associated with more heterogenous clinical signs and increased risk of progression to advanced disease.[1] Increasing awareness of the diverse cutaneous presentations in darker skin is paramount as it can have implications for prognosis.