Netherlands
The Journal of Medicine

PHOTO QUIZ

A fatal rash

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CASE REPORT

A 45-year-old south-Indian male presented with a new-onset 'rash' and 'skin tightness' that initially appeared on his face and progressed to involve his neck, trunk and extremities over a period of two months. He was referred to our institution with a presumed diagnosis of cutaneous T-cell lymphoma after skin biopsy. On physical examination, there was generalised lymphadenopathy, erythematous scaly papules, plaques and nodulo-tumoral lesions over the face, neck, trunk and bilateral extremities (*figure 1*).

The haemogram revealed leucocytosis (29,400/mm$^{3)}$ and the peripheral smear showed 28% atypical cells with convoluted/ indented nuclei. Biochemical panel showed hypercalcaemia (19 mg/dl (normal range: 8.4-10.2)), abnormal renal function (serum creatinine: 1.8 mg/dl (normal range: 0.8-1.5)) and elevated lactate dehydrogenase (LDH) [1154 IU/l (normal range: 300-600)]. Skin biopsy revealed dermal lymphoid infiltrates with extensive epidermotropism and epidermal destruction (*figure 2*). On immunohistochemistry, tumour cells showed positivity for CD3, CD5 and CD25, loss of CD7 and scattered large cells expressed CD30.

Figure 1. *Diffuse erythematous scaly papules and plaques over the neck, upper extremities and trunk and nodulo-tumoral lesions over the face*

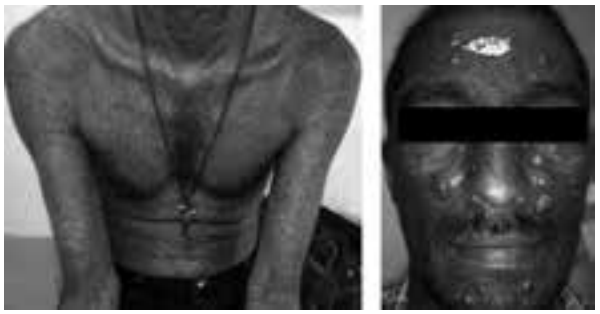
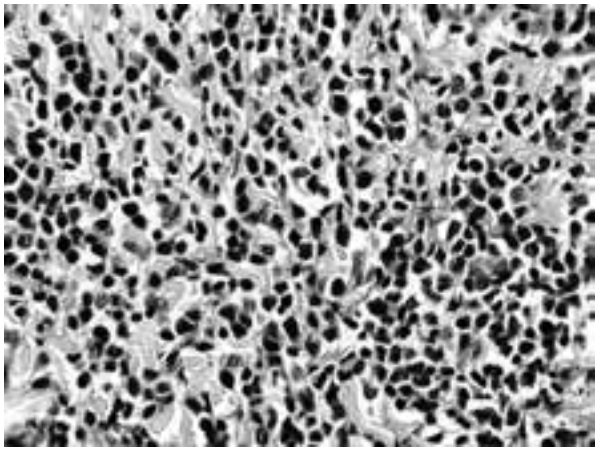


Figure 2. *H & E (x400): Section from skin, dermis shows diffuse infiltration by atypical lymphoid cells with moderate cytoplasm and convoluted nuclei*



WHAT IS YOUR DIAGNOSIS?

See page 264 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 260)

A FATAL RASH

The diagnosis of adult T-cell leukaemia/ lymphoma (ATL) was confirmed by serological analysis demonstrating antibodies to human T-lymphotropic virus 1 (HTLV-I). The patient was started on combination chemotherapy but eventually died from progressive disease with fatal pulmonary haemorrhage.

ATL is a peripheral mature T cell neoplasm caused by the retrovirus HTLV-I. HTLV-I is transmitted vertically, sexually or parenterally. The epidemiology of ATL corresponds to HTLV-I endemic zones such as southwest Japan, the Caribbean basin, north-east Iran, Central and South America and Africa. There have been only few reports from the Indian subcontinent. Close differential diagnoses include cutaneous T cell lymphoma, anaplastic large cell lymphoma and T-prolymphocytic leukaemia.[1,2]

ATL is subdivided according to the Shimoyama classification into four major clinical variants: acute (leukaemic) and lymphomatous (aggressive ATL) and the chronic and smouldering (indolent ATL). The acute variant accounts for up to 60% of cases and has a poor prognosis with median survival from six months to a year. It usually presents with generalised lymphadenopathy, hepatosplenomegaly, bone marrow involvement, hypercalcaemia, lytic bone lesions and skin manifestations.[1]

The 'flower' or 'clover-leaf' cells in the peripheral smear are considered pathognomonic. Tumour cells are positive for T cell markers such as CD2, CD4, CD5, CD45RO, CD29, T cell receptor αβ and usually lack CD7, CD8 and have reduced CD3 expression. CD25 and CD52 are expressed in most cases. Poor prognostic markers include poor performance status, age >40 years, hepatosplenomegaly, more than three involved lesions, elevated LDH and hypercalcaemia. The type of skin eruption is an independent prognostic marker and the worst prognosis is associated with the erythrodermic followed by nodulo-tumoral and multipapular eruptions.[1,3]

Aggressive ATL presents a therapeutic challenge due to intrinsic chemoresistance, large tumour burden and severe immunocompromise. Treatment options include chemotherapy, antiviral regimens, haematopoietic stem cell transplantation and investigational clinical trials. However, most patients with aggressive ATL relapse despite current day therapy.[4]

In conclusion: a fatal presentation of acute adult T-cell leukaemia/lymphoma posing a diagnostic challenge in a non-endemic region.

ACKNOWLEDGEMENTS

We would like to thank Dr. Sujith V. Cherian for his valuable suggestions in preparing the manuscript.

REFERENCES

1. Ratner L. Adult T cell leukemia lymphoma [review]. Front Biosci. 2004;9:2852-9.
2. Karthik U, Ganesan P, Sagar TG, Cyriac S, Majhi U. Adult T-cell leukemia in India: report of two cases and review of literature. J Cancer Res Ther. 2011;7:338-40.
3. Sawada Y, Hino R, Hama K, et al. Type of skin eruption is an independent prognostic indicator for adult T-cell leukemia/lymphoma. Blood. 2011;117:3961-7.
4. Bazarbachi A, Suarez F, Fields P, Hermine O. How I treat adult T-cell leukemia/lymphoma. Blood. 2011;118:1736-45.