Poster ID 37

Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma: A rare cutaneous T-cell lymphoma

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Introduction

- Primary cutaneous CD8+ positive aggressive epidermotropic T- cell lymphoma is a rare and poorly characterized variant of cutaneous T-cell lymphoma.
- Historically, known as generalized pagetoid reticulosis or Ketron-Goodman disease, is characterized by rapidly evolving and extensively ulcerated annular plaques.
- Prognosis is poor with rapid disease progression, poor response to chemotherapy and death often associated with complications of extensive epidermal necrosis and only rarely nodal or other extracutaneous involvement.

Discussion

- Primary cutaneous aggressive epidermotropic cytotoxic T-cell lymphoma should be differentiated from mycosis fungoides, which also shows epidermotropism, but has an indolent course..
- It is defined clinically by the rapid onset of erosive or ulcerated plaques usually without preceding skin lesions (patch stage).
- There is immunophenotypic expression of CD8 and the αβ T–cell receptor heterodimer, along with an atypical pagetoid T-cell infiltrate with cytotoxic granules and absence of CD4 expression.
- Clinical course is aggressive, and the response to chemotherapy is often disappointing with median survival of less than two years.

Conclusion

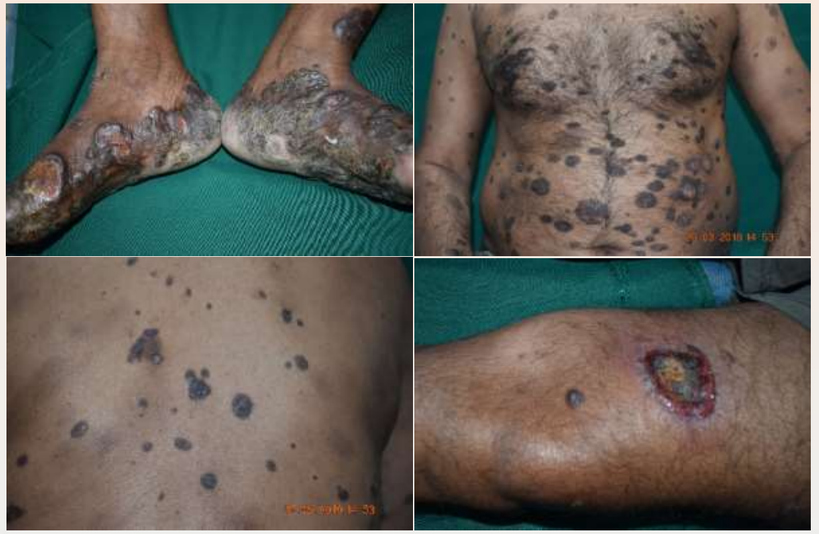
- This rare cutaneous T-cell lymphoma, though has been reported in case reports and case series, no definite diagnostic criteria have been established, and an optimum treatment is still awaiting.

References

- Swerdlow SH, Campo E, Pileri SA, et al. The 2016 revision of the World Health Organization classification of lymphoid neoplasms. Blood. 2016;127:2375–2390.
- Berti E, Tomasini D, Vermeer MH, et al. Primary cutaneous CD8-positive epidermotropic cytotoxic T cell lymphomas. A distinct clinicopathological entity with an aggressive clinical behavior. Am J Pathol. 1999;155:483–492.
- Nofal A, Abdel-Mawla MY, Assaf M, et al. Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma: proposed diagnostic criteria and therapeutic evaluation. J Am Acad Dermatol. 2012;67:748–759.

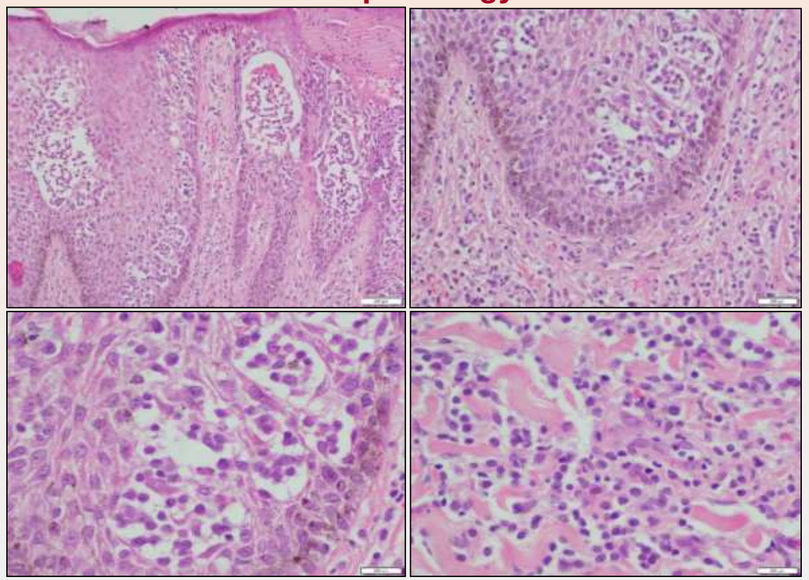
Case Report

Clinical

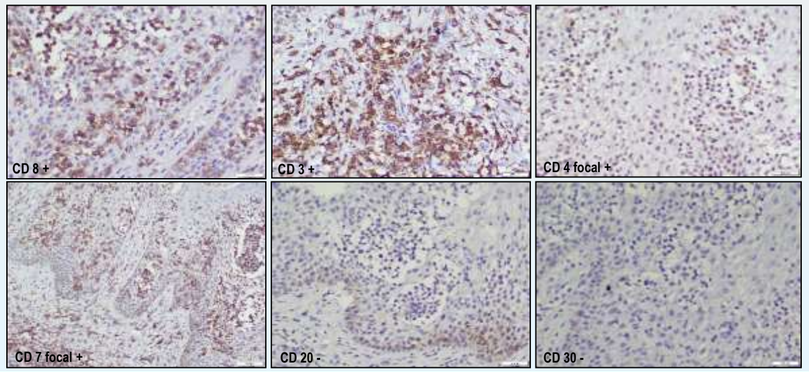


- A 47-year old man presented with multiple brown-colored plaques over his trunk and extremities.
- Onset was abrupt with rapid progression over 5-months.
- No history of systemic features, fever or weight-loss.
- Clinical examination- multiple dusky-erythematous to hyperpigmented plaques, few showing ulceration. Soles were predominantly involved with coalescent palques and ulceration.
- There were no skin nodules, patches, lymphadenopathy or hepatospleenomegaly.
- Patient was treated with methotrexate (15-30 mg/week) for 2 months without much improvement, and was referred to oncology for further management.

Histopathology



Irregular acanthosis with necrotic keratinocytes in upper layers of epidermis.
Multiple collections of large lymphocytes with prominent nucleoli in the epidermis, forming abcess at few foci, suggestive of epidermotropism.



Laboratory Investigations

- CBC, LFT, RFT, LDH- normal
- Bone marrow examination - normal
- 18F-FDG whole body PET-CT- metabolically active cutaneous soft tissue nodules