

Primary cutaneous aggressive epidermotropic CD8 + T-cell lymphoma on upper eyelid

Sir,

Primary cutaneous aggressive epidermotropic CD8 + T-cell lymphoma constitutes less than 1% of all cases of cutaneous T-cell lymphoma (CTCL).^[1] Although common subtypes of CTCL, such as mycosis fungoides (MF), are originated from CD4 + memory T cells, this subtype is derived from a cytotoxic T-cell subset and follows a more aggressive clinical course with poor prognoses.^[2]

An 80-year-old man was referred with the well-demarcated, indurated plaque with central necrotic ulceration on his left upper eyelid [Fig. 1]. He developed a single, erythematous lesion with on left upper eyelid 2 years ago. In a tertiary hospital, he was diagnosed with pseudolymphoma and had been treated with steroid intralesional injection and tacrolimus ointment. During the next year, his disease progressed and the lesion tended to bleed and he had to use an eye patch. On exam, no afferent pupillary defect, diplopia, or proptosis was present. Slit lamp examination was unremarkable. He could not elevate the upper eyelid due to mass. There was no involvement in the conjunctiva, upper fornix, and ocular surface.

A biopsy was performed and histology showed appearances



Figure 1: A slightly indurated well-demarcated ulcer, slightly pearly in periphery

suggestive of a malignant lymphoma [Fig. 2]. Skin biopsy specimen revealed a bandlike, epidermotropic, and perivascular atypical lymphocytic infiltration with large-cell morphology, which extends to the reticular dermis and subcutis. The tumor cells were positive for CD3, CD8, Bcl-2, CD56, and granzyme B (in a few scattered cells only), and negative for CD10, CD25, CD20, CD30. *In situ* hybridization for Epstein-Barr virus was negative. HIV testing and the test for monoclonal rearrangement of the T-cell antigen receptor genes were not performed. Other organ involvement was not found at a computed tomography and PET-CT. He was diagnosed with primary cutaneous aggressive epidermotropic CD8 + T-cell lymphoma and transferred to the oncologist for the combined chemotherapy with CHOP (cyclophosphamide, adriamycin, vincristine, prednisone). However, the treatment was not effective after three cycles of the combined chemotherapy. We planned to perform the radiotherapy and flap surgery, but the patient refused the surgery because of his general medical conditions such as previous myocardial infarction and diabetes.

There are approximately 30 cases of on primary cutaneous aggressive epidermotropic CD8 + cytotoxic T-cell lymphoma reported in the literature, and most of the cases represented multiple or widespread papules, nodules, and tumors, often with hemorrhage, ulceration, and necrosis.^[3] One case of solitary ulcerated lesion at ear pinna similar to this case was reported by Fika *et al.*^[4] The prognosis is very poor, according to studies by Berti *et al.*,^[2] which found median survival to be only 32 months. This case generally follows an indolent clinical course similar to that of the more common, classic CD4 + variants. The other case, reported by Fika *et al.*,^[4] showed solitary lesion, with slow progression, excellent responsiveness to local radiotherapy. The CD8 + immunophenotypic variant of MF, reported by Dummer *et al.*,^[5] is also characterized by

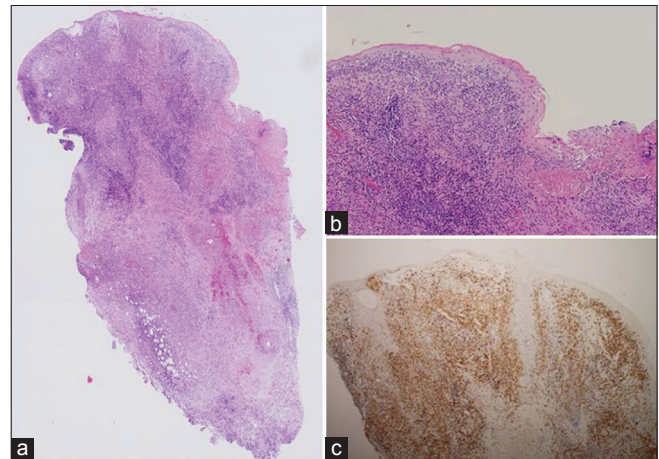


Figure 2: (a) Punch biopsy of upper eyelid lesion showed epidermal ulceration, with underlying pandermal and epidermotropic lymphocytic infiltrate, composed nearly entirely of large atypical lymphocytes, extending to and involving subcutis (H&E, $\times 40$). (b) Composed predominantly of a diffuse monotonous population of large, cytologically atypical mononuclear cells, exhibiting nuclear pleomorphism, with hyperchromatic to vesicular convoluted nuclei, some with prominent nucleoli, and moderate amount of cytoplasm, comprising approximately 80% of total cellularity (H&E, $\times 100$). (c) Immunohistochemically, large neoplastic lymphocytes showed predominantly cytotoxic phenotype with diffuse positivity for CD8, and essential negativity for CD4 (not shown)

a slowly progressing course, responsiveness to conservative therapy. The significant histologic and immunophenotypic overlap between the more indolent and aggressive cases suggest that differential diagnosis must be made based on clinical features and disease course. These indolent variants must be discriminated from the more aggressive CD8 + lymphoma to prevent aggressive treatment.

This case report is additional information to the data collected on primary cutaneous aggressive epidermotropic CD8 + cytotoxic T-cell lymphoma. In this case, the clinical course did not show an aggressive progression and solitary lesion on upper eyelid without extracutaneous manifestation unlike the most of the cases reported as primary cutaneous aggressive epidermotropic CD8 + cytotoxic T-cell lymphoma, which have no effective treatment at present.

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