A Case of an Indolent CD8-Positive Lymphoid Proliferation of the Ear
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Abstract
A lady presented with indolent slowly spreading erythematous nodule on the left external ear which on histopathology showed dense monomorphic lymphoid cells in the dermis. No epidermotropism or angioinvasion was seen. Immunohistochemistry showed that the infiltrating lymphoid cells were CD8+ but CD4-. Majority of the cases of cutaneous T-cell lymphomas have a CD4+, CD8− T-cell expression. Few cases have been reported with similar CD8-positive lymphoid proliferation with a curious ear tropism.

Key Words: CD8+, ear, T-cell lymphoma

Introduction
Cutaneous T-cell lymphomas usually have a CD4+, CD8− T-cell phenotype. A CD8+ T-cell phenotype may be rarely found in cases of mycosis fungoides, uncommon cases of primary cutaneous CD30+ lymphoproliferative disorder, around half of the cases of pagetoid reticulosis, and cases of subcutaneous panniculitis-like T-cell lymphoma with an alpha/beta T-cell phenotype, and in approximately, one-fifth of the cases of peripheral T-cell lymphoma (PTL), unspecified. We report a case of a CD8+ lymphoid proliferation presenting on the ear with characteristic clinical, morphologic, immunohistochemical features and may represent a new clinical condition.

Case Report
A 38-year-old female patient came with complaints of a single nodular swelling over the left ear which was associated with mild itching. The patient gave a history of noticing a swelling over the left ear 3 years back. It was a small, single lesion that gradually grew in size over 3 months [Figure 1]. The patient noticed improvement of lesion with reduction in size on treatment with topical medication and recurrence of lesions on stopping treatment. There was no history of change in the size of lesion over the past 1 year. The patient had complained of pain associated with lesion for 4 months. There was no associated history of weight loss or any loss of appetite. There was no history of generalized weakness, hemoptysis, hematemesis, or melena; no history suggestive of chronic cough or evening rise of temperature.

On examination, her vitals were stable with no significant systemic findings. She was moderately built and moderately nourished. There was no peripheral lymph node enlargement. Cutaneous examination showed an erythematous well-defined nodule on the left helix which was 2 cm × 1.5 cm in diameter. The lesion was firm in consistency and nontender. There were no other significant skin lesions.

The routine investigations were within normal limits. Clinical differential diagnosis included Jessner’s lymphocytic infiltrate, lupus vulgaris, leishmaniasis, and discoid lupus erythematosus.

A skin biopsy for histopathological evaluation revealed the epidermis to be atrophied with mild hyperkeratosis and dermis showed dense infiltrate of medium-sized monomorphic lymphoid cells with round to irregular
nuclear margin and blastoid chromatin [Figures 2 and 3]. The infiltrate extended into the subcutaneous adipose tissue. In the fat tissue, no rimming of adipocytes was seen. Epidermotropism and angioinvasion were not evident.

Immunohistochemistry of the lesional lymphoid cells was immunopositive for CD3, CD5, CD8, and TIA-1 and immunonegative for CD20, CD30, CD56, CD4, Granzyme B [Figures 4, 5 and 6]. Mib-1 showed a small growth fraction labeling <5% of cells. Immunohistochemical findings are summarized in Table 1. Polymerase chain reaction for T-cell clonality was negative.

Discussion
Our patient presented with a slowly growing erythematous nodule limited to the external ear, with no signs of systemic involvement and followed a benign clinical course. Histologically, the case showed a dense and diffuse dermal proliferation of monomorphic lymphoid cells with round to irregular nuclear margin and blastoid chromatin. In the fat tissue, no rimming was seen. Epidermotropism and angioinvasion were not evident.

The histopathologic features clearly suggested an aggressive lymphoma; however, the proliferation index, in this case, was remarkably low. The indolent clinical presentation and behavior of the lesion with the histologically aggressive lymphoma caused difficulty in classification. Such cases cannot be readily classified according to the new WHO-EORTC classification. The

| CD20 | CD3 | CD4 | CD8 | CD2 | CD5 | CD7 | CD30 | TIA-1 | Granzyme-B | Mib-1 |
|------|-----|-----|-----|-----|-----|-----|------|------|-----------|-------|
| -    | +   | -   | +   | -   | +   | -   | -    | +    | -         | <5%   |

+: Denotes immunopositive, -: Denotes immunonegative
provisional entity of CD8+ cutaneous T-cell lymphoma shows marked epidermotropism, ulceration, necrosis, and an aggressive course which is quite distinct from our case. Mycosis fungoides is rarely CD8+ and can be excluded on the basis of absence of papules and plaques in our case. Pagetoid reticulosis is CD8+ in 50% of cases; however, CD30 is usually positive which is absent here. Subcutaneous panniculitis-like T-cell lymphoma is usually CD8+ and shows a preferential subcutis involvement. Hence, it was ruled out.

Based on histopathological features alone the diagnosis of our case can best be classified as Peripheral T cell lymphoma (PTL) according to the present EORTC classification. In a study by Bekkenk et al., approximately 15% of PTL, unspecified with primary cutaneous disease had a CD8+ T-cell phenotype. These have an aggressive course and a poor prognosis. We observed an indolent course in our case, and so we propose our case to be classified as an indolent CD8-positive T cell lymphoid proliferation of the ear.

Most of the cases of cutaneous lymphoid proliferations presenting on the ear represent B-cell proliferation including cutaneous lymphoid hyperplasia caused by a Borrelia burgdorferi infection or cutaneous lesions of B-cell lymphocytic leukemia. These may be identified on the basis of distinct histologic and immunohistochemistry features.

After a review of literature, we were able find a few case reports with similar clinical, histologic, and immunophenotypical features as our case. To date, only 22 single lesions of indolent CD8+ lymphoid proliferation have been described. Twelve cases with CD8-positive T-cell lymphoid proliferation presenting primarily as ear lesion and showing an indolent course were reported. Beltraminelli et al. had proposed that on the basis of all the features, this entity best fits into the WHO/EORTC group of CD4-positive small/medium-sized pleomorphic T-cell lymphomas (SMPTCL) and that it should be considered a variant of SMPTCL. However, Greenblatt et al. argued that the expression of follicular helper T-cell markers was absent in some cases, and the tumor cell morphology was quite different from SMPTCL. Hence, the need for a separate entity of cutaneous indolent CD8-positive T-cell lymphoid proliferation among the existing classification system. Better recognition of this condition is necessary to prevent unwarranted aggressive treatment if classified according to the existing system of classification.

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Conflicts of interest
There are no conflicts of interest.

What is new?
Indolent CD8 positive T-cell lymphoid proliferation has distinct clinical, histopathological and immunohistochemistry features and warrants a distinct place in the existing classification system.

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