Primary cutaneous aggressive epidermotropic CD8+ cytotoxic T-cell lymphoma: A case report and significance of timely ancillary testing

Editor,
A 70-year-old man was admitted to the Medical Intensive Care Unit with respiratory tract infection, sepsis, and acute kidney injury; not responding to treatment. Six months back, he had presented with maculopapular skin lesions on the upper and the lower limbs. A skin biopsy had raised the possibility of chronic lymphocytic vasculitis in the primary laboratory. No immunohistochemistry (IHC) was done at that time. As the patient deteriorated, a paraffin block of the same biopsy was submitted to our laboratory for a lymphoma IHC panel 6 months later.

Morphology on routine H and E microscopic examination showed a cutaneous lymphoproliferative lesion displaying dermal nodular aggregates of atypical lymphoid cells surrounding the adnexal structures, and neurovascular bundles along with pagetoid type of epidermotropism [Figure 1a and b].

On IHC the lesional cells were positive for CD3 [Figure 1c] with a high Ki-67 index, [Figure 1d] CD8 predominance [Figure 2a] and negative for CD30 [Figure 2b], CD20, and CD4.

T-cell receptor gamma gene rearrangement confirmed T-cell origin and clonality [Figure 2c].

Amalgamating clinical presentation, histomorphology, IHC, and molecular assay results, a conclusive diagnosis of primary cutaneous aggressive epidermotropic CD8+ cytotoxic, T-cell lymphoma was established. This lymphoma represents <1% of all cases of cutaneous T-cell lymphoma (CTCL) that has been classified as a nonspecific, provisional entity according to the WHO/EORTC classification in 2005, and the 4th edition of the WHO classification in 2008.[1,2] Less than 50 cases have been reported worldwide and probably this is the first Indian patient.

It was first reported by Berti et al., in 1999. He described this tumor as a distinct clinicopathological entity with an aggressive clinical behavior. It is characterized by widespread rapidly evolving papules, plaques, and papulonodules, often showing central necrosis and ulceration.[3] Uncommonly, it may present as multiple maculopapular eruptions on the hands, feet, and face.[4]

Patients are usually adults with a slight male predominance. Our patient was an elderly gentleman who presented with maculopapular skin lesions.

Nofal et al.[5] proposed two sets of diagnostic criteria using a combination of clinical, histopathological, and IHC features.

Constant features - all of constant clinical, histopathological, and IHC features must be present to establish a definite diagnosis.
Clinical:
1. History: Short; few weeks or months
2. Course: Aggressive
3. Lesions: Widespread papules, plaques, and tumors often ulcerated, without any precursor lesions.

Histopathological:
1. Epidermotropism, often prominent
2. Nodular or diffuse infiltrates of pleomorphic T-cell.

IHC:
1. CD8+
2. CD4−.

Variable features - helps to avoid missing cases.

Clinical (these cases showed classic constant lesions, in addition to uncommon variable features):
1. Pyoderma gangrenosum: Like lesions
2. Pagetoid reticulosis: Like superficial hyperkeratotic patches and plaques
3. Annular erythematous scaling patches.

Histopathological:
1. Spongiosis, blistering, and necrosis
2. Deep infiltration reaching subcutaneous fat with rimming of adipocytes
3. Invasion and destruction of adnexal skin structures
4. Angiocentricity and angioinvasion.

IHC:
1. Variable expression profiles of CD2, CD5, CD7, CD15, CD30, CD45, and CD56.

Conventional therapies for classic CD4+ CTCL are usually ineffective in this type of lymphoma, which requires multiagent chemotherapy and bone marrow transplantation. Hence, accurate and timely diagnosis is necessary.

The clinical course is usually aggressive with a high tendency for systemic spread to extracutaneous sites such as central nervous system, lung, and testis while lymph nodes are usually spared. The prognosis of the patients is poor with a median survival of 32 months from diagnosis.

As depicted through this case report, it would be logical and wise to conclude that reflex ancillary tests performed on a primary biopsy provide an early and conclusive diagnosis catalyzing timely optimal patient management.

This patient presented with a cutaneous lymphoma, which was not suspected or confirmed in the initial stage. He had multisystem involvement over a period of 6 months, and, unfortunately, succumbed to the disease on the day he received his final diagnosis.

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Conflicts of interest
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