Angioimmunoblastic T-cell Lymphoma: Case Report and Review of the Literature

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Introduction

Angioimmunoblastic T-cell lymphoma (AITL) is a rare, aggressive form of peripheral T-cell lymphoma that is characterized by lymphadenopathy, night sweats, fever, weight loss, and autoimmune phenomena. Cutaneous manifestations are present in up to 50% of cases. AITL comprises 1-2% of all non-Hodgkin’s lymphomas. It typically presents in the seventh decade, however cases have been reported in adults ranging from 20 to 91 years of age.

History

A 72-year-old Caucasian woman presented with a pruritic, erythematous eruption around the neck of three weeks duration (Fig A and B). Her past medical history was significant for diffuse large B-cell lymphoma diagnosed three months prior by a right cervical lymph node biopsy. She was being treated with bendamustine and rituximab.

Physical Examination

Physical examination revealed erythematous, edematous papules coalescing into indurated plaques around the neck.

Histopathology

Punch biopsy demonstrated a scant, atypical lymphoid infiltrate focally involving the deep dermis. The cells were medium to large in size and contained hyperchromatic/pleomorphic nuclei (Fig C and D). They were positive for CD3 and CD4, a feature which was concerning for T-cell lymphoma. Further staining of skin and previous cervical lymph node specimens revealed positivity for CD10, Bcl-2, Bcl-6, and PD-1. Polymerase chain reaction demonstrated a T-cell population with clonally rearranged T-cell receptor genes. Primers for immunoglobulin heavy and light chains showed no evidence of a clonal B-cell population.

Clinical Course

The patient declined further treatment and chose hospice care.

Discussion

AITL is characterized by lymphadenopathy, night sweats, fever, weight loss, and autoimmune phenomena. Cutaneous manifestations occur in up to half of patients, the presence of which portends a poor prognosis. Often the rash is a nonspecific, erythematous macular and papular eruption mimicking a morbilliform viral exanthem or drug eruption. Urticarial, nodular, papular, purpuric, eczematous, erythodermic, and vesiculobullous presentations have also been described. In up to one-third of cases, the eruption occurs in association with a new medication, often leading to an initial misdiagnosis of drug hypersensitivity reaction.

Five histological patterns have been described with cutaneous AITL: (1) superficial perivascular infiltrate of eosinophils and lymphocytes that lack atypia, (2) sparse perivascular infiltrate with atypical lymphocytes, (3) dense dermal infiltrate of pleomorphic lymphocytes, (4) leukocytoclastic vasculitis without atypical lymphocytes, and (5) necrotizing vasculitis.

Conclusion

AITL is difficult to diagnose due to nonspecific clinical and histological findings. Cutaneous manifestations are seen in AITL in up to half of cases, which may occur early or in advanced disease. Like all cutaneous metastases, the appearance of the lesions can vary greatly. Our case demonstrates that dermatologists and dermatopathologists can make this diagnosis in the appropriate clinicopathologic context utilizing appropriate immunohistochemical staining and gene rearrangement studies.

References