Dermatoscopy and Histology of Primary Cutaneous Aggressive Epidermotropic CD8+ T-Cell Lymphoma

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Case Presentation

A 65-year-old Caucasian man presented with an exophytic ulcerated tumor on the left thigh, along with pruritic skin papules and nodules, initially on the lower limbs, thereafter, spreading to the trunk, upper limbs, and intertriginous folds, over a period of 5 months (Figure 1, A and B).

Dermatoscopic examination of the papules (early lesions) revealed clustered dotted and very few short linear vessels and shiny white lines in a pink background. As the lesions progressed and expanded, coiled and hairpin vessels, shiny white lines and yellow scales predominated. The tumor of the thigh was mainly characterized by coiled and a few, short linear vessels (Figure 1, C, E and G).

Histologic examination showed diffuse dermal infiltration by a lymphoid proliferation associated with epidermal necrosis, angiocentricity and angioinvasion with fibrinoid necrosis of small vessels (Figure 1, D, F and H). Immunohistochemical findings were diagnostic of primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma [1]. The histological findings, together with the detected phenotype of CD7-/CD2+, have been associated with high invasive potential. Therefore, further biopsies were obtained. Left femoral lymph node biopsy was positive, whereas bone marrow biopsy was negative.

Teaching Point

Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma is a subtype of cutaneous lymphoma. Dermoscopy could facilitate clinical suspicion and early diagnosis of this rare neoplasm [2]. The evolution of vessels from short linear, to coiled and hairpin vessels, probably correspond to histological angiocentricity and angioinvasion during disease progression, contributing to its invasiveness to extra-cutaneous sites, unfavorable prognosis, and unclear optimal treatment.

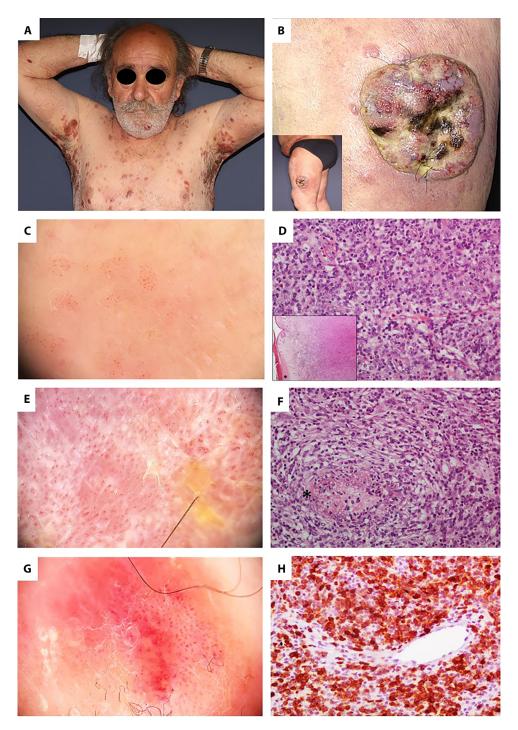


Figure 1. (A,B) Clinical presentation of primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma). (B) Papules, nodules, and ulcerating tumor on the left thigh. Papule (C), nodule (E) and tumor (G) dermoscopy revealed: clustered dotted and very few short linear vessels (C), shiny white lines in pink background (E) and coiled and hairpin vessels, shiny white lines, and yellow scales (G). Histologically, diffuse infiltrates of lymphoid proliferation associated with epidermal necrosis (*) of medium-sized and large neoplastic cells with distinct nucleoli (D) were observed. (H) Angioinvasion with fibrinoid necrosis (*) of small vessels were noted and associated to the dermoscopic features. Phenotypically, they were positive for CD8+), CD3+, perforin+ and negative for CD5-, CD7-.

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