Angioinvasive Lymphomatoid Papulosis type E. Case Report and Review of the Literature


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CASE REPORT

- Previously healthy 19-year-old man with a 4-year history of waxing and waning, well-demarcated erythematous macules, papules and nodules, some of which evolved into necrotic ulcers; predominantly in the trunk and lower extremities (Fig 1,2).
- He had previously been attended elsewhere, where three skin biopsies were performed, two reported as LyP type A and one as Large T-cell CD30 (+) lymphoma; and he received systemic chemotherapy for 2 years, without improvement.
- Upon admission to our hospital, a new biopsy revealed dermal aggregates of large atypical cells, which exhibited distinct angioinvasion along superficial and deep blood vessels, together with a mixed inflammatory infiltrate, composed of small lymphocytes, eosinophils and scant neutrophils. Neoplastic cells were positive for CD30, CD3, CD4 & CD8. The diagnosis of angiocentric (type E) lymphomatoid papulosis (LyP) was thus rendered and the patient was started on methotrexate (10 mg p.o. weekly) plus PUVA therapy.

DISCUSSION

LyP is a rare papulo-nodular skin disorder that belongs to the spectrum of primary cutaneous CD30+ lymphoproliferative disorders. Clinically, LyP is characterized by a variable number of self-healing papulo-nodular lesions, with a typical waxing and waning course. Historically LyP was sub-classified in to three types (A, B, C) based on histopathological findings. Recently two new histopathological variants were described: LyP type D corresponding to an epidermotropic CD8+ & CD30+ infiltrate and LyP type E, which is characterized by oligolesional papules that rapidly ulcerate and evolve into large necrotic eschar-like lesions with a diameter of 1-4 cm and histologically by an angiocentric and angiodestructive infiltrate of small- to medium-sized atypical lymphocytes expressing CD30 and frequently CD8. As in other forms of LyP, lesions exhibit spontaneous regression in weeks, recurrences are common, and the prognosis is excellent with no extracutaneous spread or disease-related deaths.

This case exemplifies the clinicopathologic findings of an exceedingly rare variant of LyP. The histological findings of a dermal infiltrate composed of atypical and pleomorphic angiocentric lymphocytes make this disorder difficult to differentiate from an aggressive primary cutaneous lymphoma such as NK/T-cell lymphoma and gamma/delta T-cell lymphoma. However, the indolent clinical course characterized by waxing and waning papules, nodules and/or ulcers enables the correct diagnosis from a clinical standpoint but only for those familiar with the entity.

We present this case to help increase the awareness amongst dermatologist of this recently described disease which should be kept in the differential diagnosis when dealing with dermal-based angiocentric lymphoid infiltrates.

SELECTED REFERENCES