Eur. J. Pediat. Dermatol.
15, 113-6, 2005

Atypical Cutaneous Lymphoid Hyperplasia of mixed B/T type in a 15-year-old girl[Δ].

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Summary A case of atypical cutaneous lymphoid hyperplasia of mixed B/T type in a 15-year-old girl is reported. The clinical features led to diagnose a reactive disorder to exogenous stimuli. However, some pathological findings were suggestive of malignancy. Five years after the excisional removal the teenager was in good health, thus confirming the diagnosis of cutaneous lymphoid hyperplasia.

Key words Cutaneous lymphoid hyperplasia, pseudolymphoma.

Cutaneous lymphoid hyperplasia clinically can be indistinguishable from cutaneous lymphomas. Architectural features and cytologic features help in differentiating pseudolymphomas from true lymphomas, but some reactive disorders may present malignant histologic features. We report an unusual case of cutaneous lymphoid hyperplasia with atypical histologic features.

Case report

An asymptomatic 15-year-old girl in good health was seen because of a rapidly enlarging nodule of the right supraclavicular region. The latter had occurred suddenly three months before. There was no history of drug intake and throughout the previous months the patient had reported no systemic symptoms such as fever, arthralgias or neurologic problems. Clinical examination showed a 2 cm in diameter, well defined, hard, subcutaneous, erythematous, nodule, (Fig.1). Biochemical investigations showed gamma globulins 10.1% (normal values -n.v.-12-20%), IgG 591 mg/dl (n.v. 726-1085 mg/dl), IgM 97.9 mg/dl (n.v. 35-72 mg/dl). Work up for *Borrelia burgdorferi*, *Epstein-Barr virus* and *HIV* infection was negative. Chest X-ray examination and abdominal ultrasonography were normal. The nodule underwent excisional biopsy. Histologic examination showed a dense and diffuse mixed inflammatory infiltrate, which only spared the upper dermis and the epidermis (Fig. 2). Although focally, the infiltrate formed relatively well-defined foci centered on blood vessels and appendages, whereas the collagen network was diffusely disrupted (Fig. 2, 3). The infiltrate was composed by small irregular lymphocytes, occasionally larger lymphocytes with clear cytoplasm that were consistent with marginal cells (Fig. 4), extraordinarily abundant plasmacytoid monocytes characterized by roundish nucleus, fine chromatin and numerous apoptotic figures and, finally, ill-defined aggregates of larger histiocyte-like cells also associated to apoptotic

[Δ]*Poster presented at the 10th World Congress of Pediatric Dermatology, Rome (Italy) July, 7-10, 2004*

Fig. 1

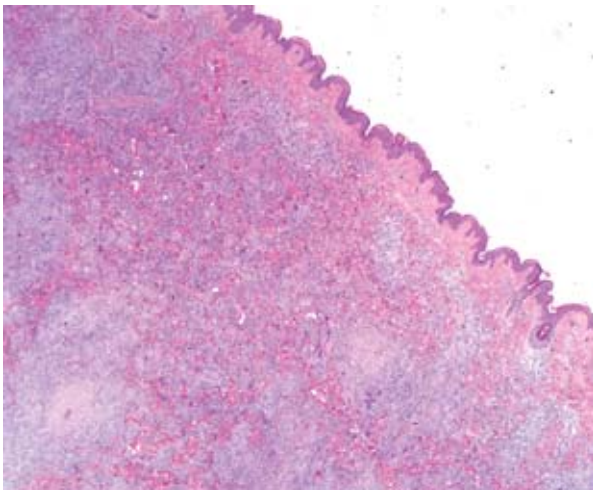

Fig. 2

Fig. 1, 2, 3, 4: Erythematous nodule on the right supraclavicular region (Fig. 1). In Fig. 2 (HE, 25x) skin biopsy specimen showing a diffuse, pandermal inflammatory infiltrate which spares the epidermis. The collagen network appears diffusely disrupted. In Fig. 3 (HE, 100x) lymphoid cells and histiocyte-like cells aggregating to form a granuloma-like structure. In Fig. 4 (HE, 400x) small and larger lymphocytes with clear cytoplasm.

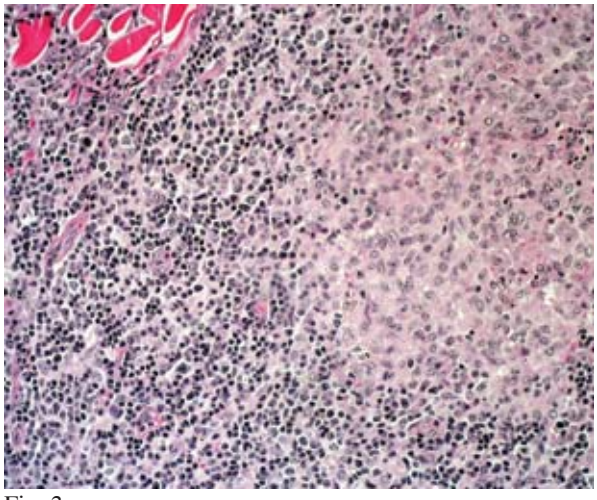

Fig. 3

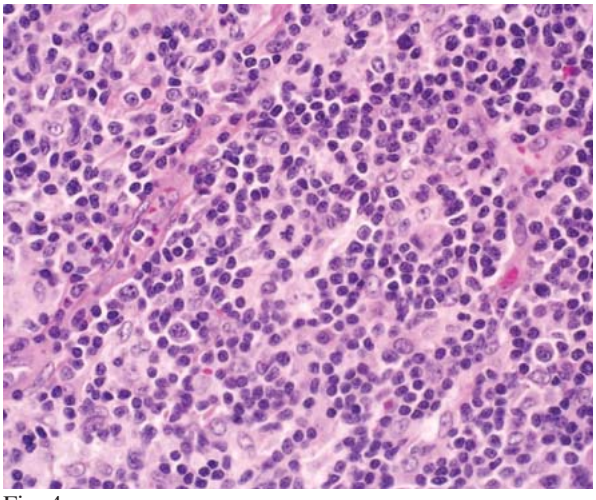

Fig. 4

figures. The latter cells, which were reminiscent of interdigitating cells, had a smaller amount of cytoplasm than expected in true histiocytes. They could represent an interdigitating cell differentiation of plasmacytoid monocytes. Some giant cells containing foreign bodies were also seen.

Immunohistochemistry on paraffin-embedded tissue sections detected a predominance of small and large CD3+ T cells and a minor population of CD20+ B cells, both of which formed a few follicles or were scattered in interfullicular location as small and large cells. The plasma cell component was polyclonal. A few loose meshwork of CD21+ follicular dendritic cells were seen corresponding to the B-cell follicles. The stain with CD68 (KP-1) demonstrated many positive cells including macrophages, histiocytic dendritic cells and clusters of plasmacytoid monocytes. The stain for S-100 protein showed a very large number of reactive interdigitating cells both scattered and forming prominent clusters, corresponding to the aggregates of histiocyte-like cells.

Five years after surgical excision no recurrence was noted and actually the girl is in good health.

Discussion

Differential diagnosis of cutaneous lymphoproliferative disorders represents one of the most challenging problems in dermatology and dermatopathology. Applications as immunohistochemical and molecular techniques have provided useful criteria for distinguishing between the two conditions, although only long term outcome records permit sometimes to confirm the correctness of the diagnosis (1).

This case of atypical lymphoid hyerplasia was unusual because, despite the collagen disruption was strongly suggestive of a cutaneous lymphoma, the polymorphic findings of the infiltrating cells favored a reactive process. The latter interpretation was supported by the immunophenotypic findings of a mixed proliferation of B and T cells (2), histiocytes and S-100 interdigitating cells.

Due to the negative Lyme serology and the absence of any systemic symptomatology, a solitary borrelia lymphocitoma cutis was excluded. However, we were not able to perform a PCR analysis. As the girl remembered having seen the nodule early in the morning during the first days of September, it is possible to hypothesize that this lesion was due to a persistent nodular arthropod-bite reaction. The absence of CD30+ T cells has been claimed as a possible clue to differentiate persistent arthropod-bite reactions from CD30+ lymphoproliferative disorders (3), although this feature cannot be considered pathognomonic.

ACKNOWLEDGEMENT. The Authors thank Glauco Frizzera, M.D., New York Hospital-Cornell Clinical Center, who provided a second opinion pathology slide review.

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