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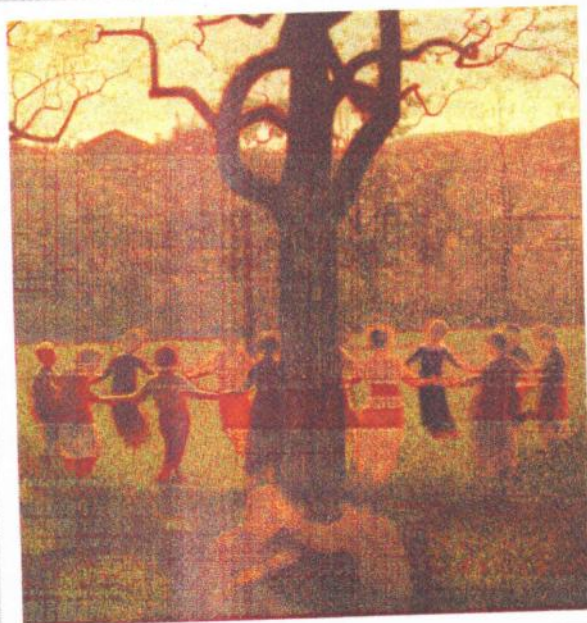
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Acquired self-healing Langerhans cell histiocytosis of the skin in early childhood. Case report.

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Summary

Langerhans cell histiocytosis is a rare idiopathic disorder characterized by proliferation of specialized bone-marrow derived Langerhans cells. A three months breast-fed child presented from the age of two months widespread small red-brownish papules, erosions and crusts on the scalp, face, chest, back and extremities. The initial diagnosis of the disease was seborrheic dermatitis. Hepatomegaly, splenomegaly and lymphadenopathy were absent. On X-ray examination the lungs and bones were not affected. Light microscopy examination of the skin (HE, PAS) showed a histiocytic infiltrate in the papillary dermis with epidermotropism. The immunohistochemical examination demonstrated S100+, lysozyme- and CD68- dendritic cells in the infiltrate. Electron microscopy examination revealed specific Birbeck granules in these cells. After one month treatment with topical corticosteroids and emollients the lesions regressed. Because of the clinical features and the results of the light microscopy, immunohistochemical and electronmicroscopic examinations of the skin, the case here reported should be considered an acquired self-healing Langerhans cell histiocytosis. However, the patient is carefully followed up to detect a possible relapse or progression of the disease.

Key words

Langerhans cells histiocytosis, Birbeck granules, Protein S100.

Histiocytosis is a term, which describes a group of syndromes, characterized by histiocytic cell proliferation in different organs. The disease may affects the skin, bones, lymph nodes and visceral organs such as lungs, liver, spleen.

Three histiocytosis classes were defined by Histiocyte Society: Class I - Langerhans cell histiocytosis; Class II - Histiocytosis of the mononuclear phagocyte system and Class III - Malignant clonal proliferation, such as acute monocytic leukaemia (3, 14).

Langerhans cell histiocytosis (LCH) is a group of idiopathic disorders characterized by proliferation of specialized bone-marrow derived Langerhans cells (5). The Langerhans cells were described for the first time as epidermal

dendritic cells by the German pathologist Paul Langerhans in 1868 (11). Langerhans cells, which are skin histiocytes with a specific antigen presenting function are immunohistochemically different from the macrophages. The latter are protein S100-, lysozyme + and CD 68+ (5). On the other hand, the histiocytes in LCH are CD1a and Protein S100 positive and contain Birbeck granules as normal intraepidermal Langerhans cells (14). The immunomarking with Protein S100 antibodies was accepted as an additional method in the diagnosis of Langerhans cells histiocytosis (7, 17), although anti-CD1a is more specific immunohistochemically (23).

The clinical spectrum and prognosis of LCH significantly vary ranging from benign single