ACQUIRED PERIORAL CUTANEOUS LANGERHANS CELL HISTIOCYTOSIS

Khedim N., Hali F., El Fatoiki F.Z., Chiheb S.
Department of Dermatology's Diseases, CHU Ibn Rochd Casablanca, Morocco

Keywords Histiocytosis, Langerhans cells, skin, mouth.

Abbreviation **LCH** = Langerhans cell histiocytosis.

Case report. An 11-year-old girl was first observed for the presence of asymptomatic nodular lesions of the perioral region dating back a few weeks. Family and personal history did not highlight relevant episodes.

Dermatological examination (Fig. 1) highlighted papulo-nodular lesions mainly on the upper lip, but also on the lower lip and in the vicinity of the left naso-labial groove. The largest lesion on the left upper lip had a maximum diameter of 2 cm; the lesions were flesh-colored, eroded and crusted in some places, had an irregular surface, in some cases warty; on palpation the lesions had a hard consistency, were movable on the deep planes and not painful. A 1.5 cm in diameter adenopathy was present anterior to the left submaxillary gland. There were no alterations of the other organs. Because a clinical diagnosis was not easy, a biopsy of a skin lesion and lymph node was performed.

The histological examination of the skin lesion showed an ulcerated nodule (Fig. 3) consisting mainly of lymphocytes and large histiocytic cells with a lax chromatin reniform nucleus and abundant eosinophilic cytoplasm (Fig. 3, inset). On immunohistochemistry, histiocyte cells were positive for S-100 and CD1a (Fig. 4). Lymph node biopsy showed nonspecific reactive lymphadenitis with no signs of malignancy. Clinical and histological data led to the final diagnosis of cutaneous acquired Langerhans cell histiocytosis. Pediatric onco-hematological examination confirmed exclusive skin involvement. The skin lesions were treated with topical corticosteroids and in the following months regressed leaving scarring (Fig. 2).

After one year from the first observation there were no new skin lesions and no other organs were involved. Further dermatological and oncohematological monitoring was planned.

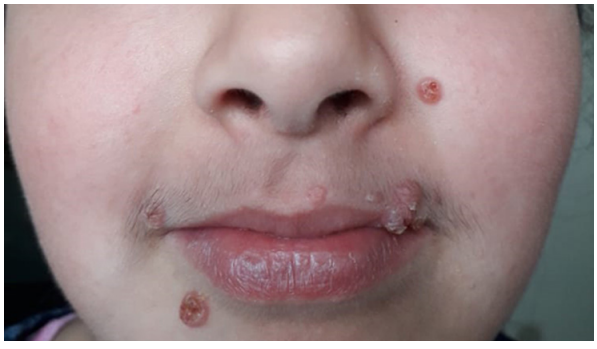

Fig. 1

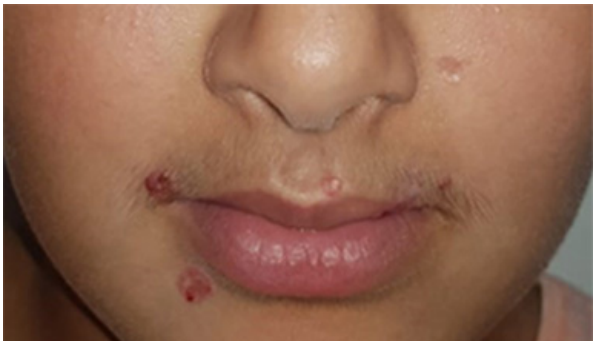

Fig. 2

Fig. 1, 2: Acquired cutaneous LCH with papulo-nodular perioral lesions (Fig. 1). In Fig. 2 scarring outcomes of the previous lesions.

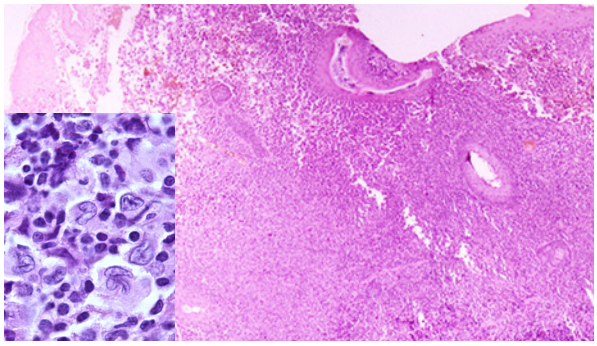

Fig. 3

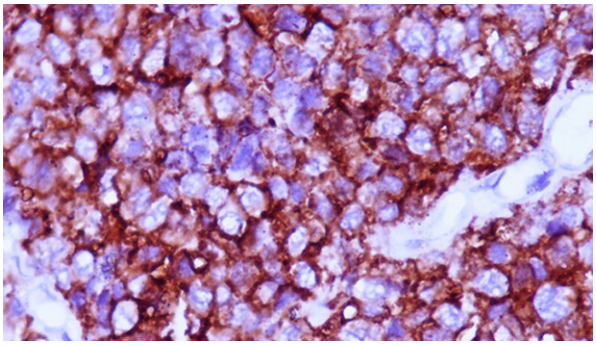

Fig. 4

Fig. 3, 4: In fig. 3 (H&E, 10x) eroded infiltrate with lymphocytes and Langerhans cells (Fig. 3, inset, H&E, 100x). The latter were CD1a+ (Fig. 4).

Discussion. LCH is an inflammatory neoplasm consisting of cells derived from medullary precursors, which usually have mutations of the MAPK signaling pathway (1). The spectrum of LCH is extremely large ranging from a single lesion affecting a single organ, generally bone or skin, to generalized multisystem lesions. The prognosis of the disease is also extremely variable, ranging from spontaneous resolution in a few months in the forms affecting only one organ to death in a few weeks in the multisystem forms with organ dysfunction.

According to some Authors (1) the high-risk forms of LCH originate from a somatic mutation of a hematopoietic progenitor, a mutation that is therefore also present in blood and bone marrow cells, while the low-risk forms originate from a somatic mutation of a dendritic precursor present in a peripheral tissue, for example the skin, and therefore the mutation would be present only in that peripheral tissue.

Skin lesions, which are frequently present in LCH, were in the past considered a sign of poor prognosis because they usually added to the visceral involvement and meant a greater extension of the disease. The discovery of exclusively cutaneous forms of LCH (3) has changed the prognostic significance of skin lesions in LCH. The concept has thus been established that, like LCHs that only affect the bone, even those that only affect the skin can remain localized in the starting tissue and can spontaneously regress. In the presence of LCH skin lesions, neither histology nor immunohistochemistry are able to give indications on the benignity or malignancy of that LCH. The lack of symptoms affecting other organs and the negativity of laboratory investigations will speak in favor of a favorable prognosis. However, in case of skin lesions even the dermatologist is able to predict a favorable prognosis. Indicating a favorable prognosis will be the low number of lesions, their distribution in non-seborrheic sites, their polymorphism and also, from an anamnestic point of view, their presence at birth; the lack of ulceration and the small size of the lesions are not indicative of benignity (2). Based on these criteria, the most benign form is certainly congenital isolated cutaneous Langerhans cell histiocytoma; the congenital Hashimoto-Pritzker form almost always has a benign prognosis; acquired LCH that begins only with skin lesions has a less predictable prognosis: some of these forms regress spontaneously, whereas others move towards the multisystem form.

In the current case in favor of a favorable prognosis is the low number of lesions that are not distributed in seborrheic sites, but the onset at 11 years must recommend careful monitoring. On the other hand, pure cutaneous self-healing forms have been described, albeit exceptionally, in adults and in the elderly, in the latter sometimes periorally (4) as in the current case.

Conclusion. The actual case was described to underline the difficulties in establishing the prognosis of acquired pure cutaneous Langerhans cell histiocytosis and also for the rarity of the perioral localization.

Conflicts of interest

The Authors declare that they have no conflicts of interest.

Address to: Nesrine Khedim
Department of Dermatology'sDiseases
CHU Ibn Rochd Casablanca
1, Rue des Hôpitaux Casablanca
e-mail: khedimnesrine@gmail.com

References

1)Berres M-L., Lim K.P.H., Peters T. et Al. 2014. BRAF-V600E expression in precursor versus differentiated dendritic cells defines clinically distinct LCH risk groups. *J. Exp. Med.* 211 (4): 669-83.

2) Bonifazi E., Milano A. 2015. Histiocytoses. *Eur. J. Pediat. Dermatol.* 25 (1): 27-52. 10.26326/2281-9649.25.1.1088.

3) Hashimoto K., Pritzker M.G. 1973. Electron microscopic study of reticulohistiocytoma. *Arch. Dermatol.* 107 (2): 263-70.

4) Subramaniyan R., Ramachandran R., Gnanasekaran Rajangam G., Donaparthi N. 2015. Purely cutaneous Langerhans cell histiocytosis presenting as an ulcer on the chin in an elderly man successfully treated with thalidomide. *Indian Dermatol. Online J.* 6 (6): 407-9.