Generalised lichenoid juvenile xanthogranuloma with systemic involvement

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Case

A 5 years-old boy presented multiple asymptomatic yellowish little papules with a predominant flexural distribution in the eyelids, neck (Figure 1), armpits, antecubital and popliteal fossa (Figure 2), groins, and back.

The skin biopsy showed a well-demarcated dermal infiltrate surrounded by a downward extension of the rete ridges: the infiltrate was composed by histiocytes, Touton giant cells and eosinophils (Figure 3). Immunohistochemical staining revealed cell expression for CD68 but not for Langerina, CD1a and S-100 (Figure 4). Transient peripheral eosinophilia was observed. Ophthalmologic and radiologic exams didn't revealed extracutaneous involvement. A generalised lichenoid juvenile xanthogranuloma was diagnosed.

After a follow-up period of two-years, he developed a central diabetes insipidus. Radiologic exams revealed an infiltration of the pituitary stem (Figure 5), nodes and salivary glands.

The biopsy of a salivary gland showed a mixed infiltrate within a fibrous stroma composed by histiocytes with vacuolated cytoplasma, plasmatic cells, lymphocytes, neutrophils, eosinophils and multinucleated giant cells (Figure 6). Emperipolesis was observed at the periphery on the sample.

Immunohistochemical staining revealed cell expression for CD68, S100 and FXIII (Figure 7). No mutations were detected on BRAF gene studies.

Due to the extracutaneous involvement, systemic chemotherapy with the multisystemic Langerhans cell histiocytosis treatment was recommended. A partial central nervous system response was observed but skin lesions had the same aspect.

Figure 1. Skin lesions in the eyelids and neck.

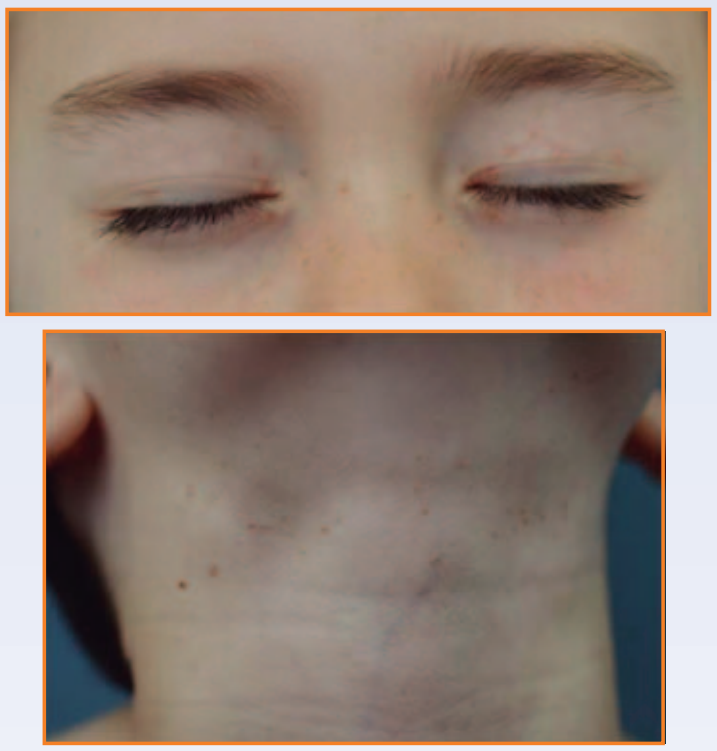


Figure 2. Skin lesions in armpits, antecubital and popliteal fossa.

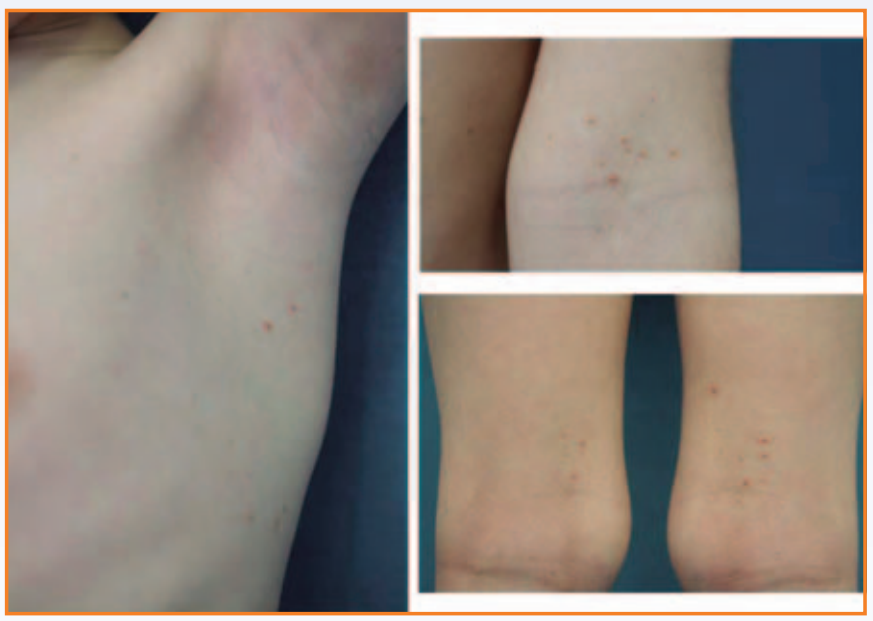


Figure 3. Histological aspect of a skin lesion: dermal infiltrate composed by histiocytes, Touton giant cells and eosinophils surrounded by extension of the rete ridges.

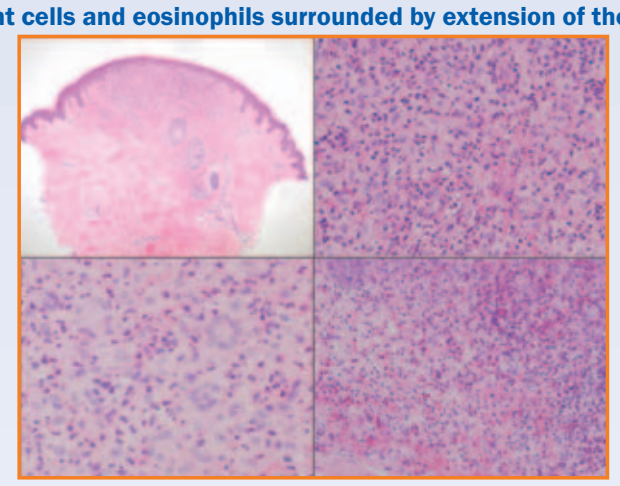


Figure 4. Immunohistochemical features of the dermal infiltrate.

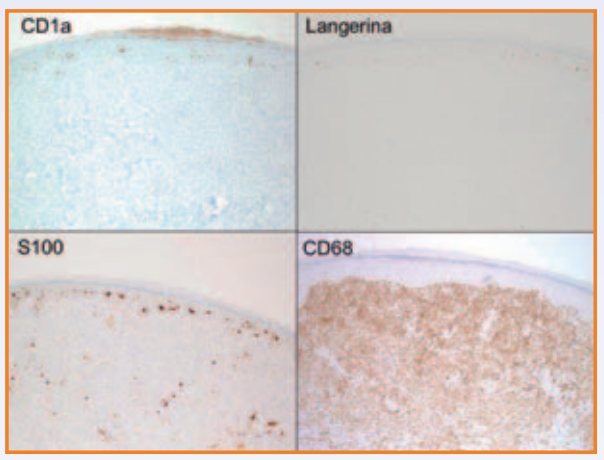


Figure 5. NMR: infiltration of the pituitary stem.

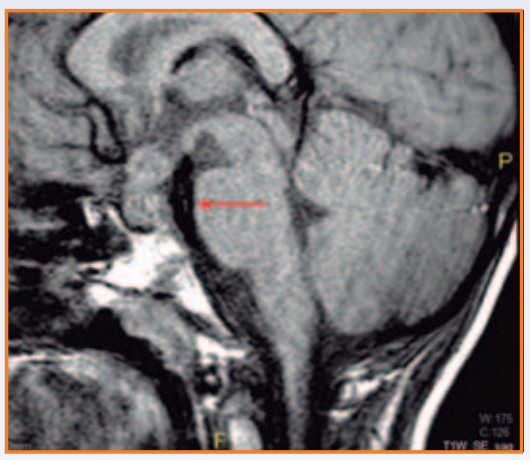


Figure 6. Histological aspect of the salivary gland biopsy.

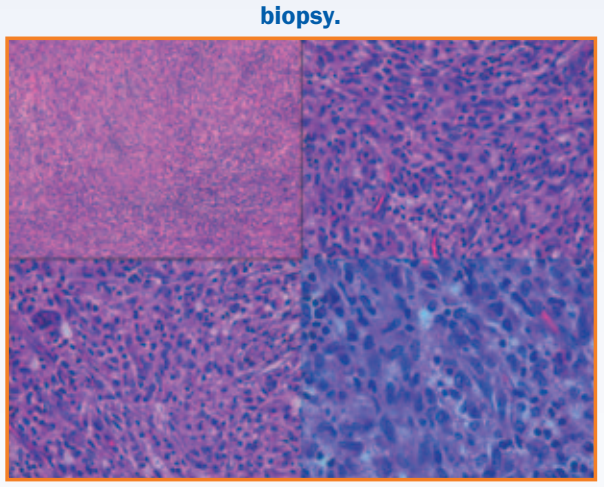
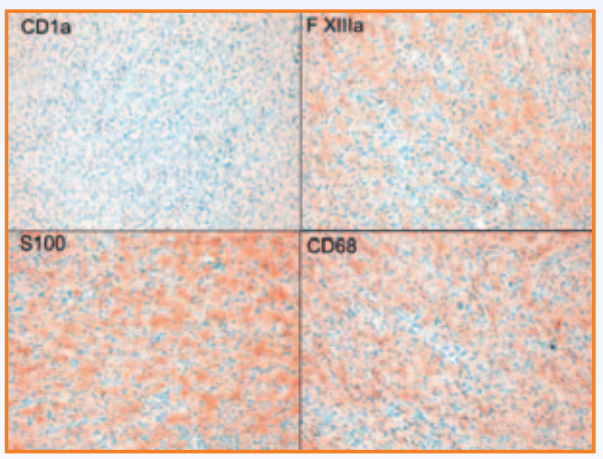


Figure 7. Immunohistochemical features of the salivary gland infiltrate.



Discussion

Generalised lichenoid juvenile xanthogranuloma is an infrequent variant of juvenile xanthogranuloma with a good prognosis. Until now, no previous reports of extracutaneous involvement of generalised lichenoid juvenile xanthogranuloma have been described. In the present case, a Rosai-Dorfman disease was suspected in the salivary gland. Diabetes insipidus is a well-known complication of Langerhans and non-Langerhans histiocytosis. Regarding the multiorgan involvement in this case, a systemic involvement of both Langerhans and non-Langerhans histiocytosis was suspected. This can be explained by a same cell progenitor that can differentiate to different cell lines depending on the environmental stimuli.

In conclusion, we recommend a long period of follow-up in any patient diagnosed of cutaneous histiocytosis, even if a good prognosis is suspected.

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