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TYPE B LYMPHOMATOID PAPULOSIS IN A 12-YEAR-OLD CHILD

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Abbreviations **LP** = lymphomatoid papulosis; **PCL** = primary cutaneous lymphomas.

Case report. A 12-year-old boy was first observed for the presence of about fifteen asymptomatic, raised skin lesions, which had started on the left buttock and then appeared on the trunk and limbs. The initially small lesions grew to the size of a chickpea, sometimes ulcerated, but then regressed in 3-8 weeks without usually leaving a scar. Dermatological examination showed red-violet papules and nodules of variable size between 3 and 13 mm (Fig. 1, 2). The sometimes ulcerated lesions were in various evolutionary phases and distributed randomly on the limbs and trunk. The histological examination showed a lymphoid, sometimes epidermotropic infiltrate (Fig. 1, 3), consisting of small and medium sized lymphocytes. The infiltrate affected the full thickness dermis and was distributed around

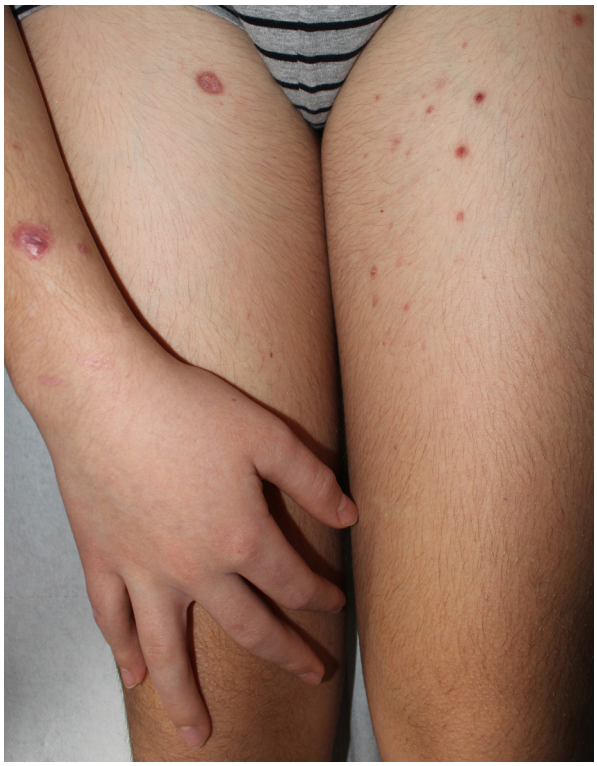

Fig. 1

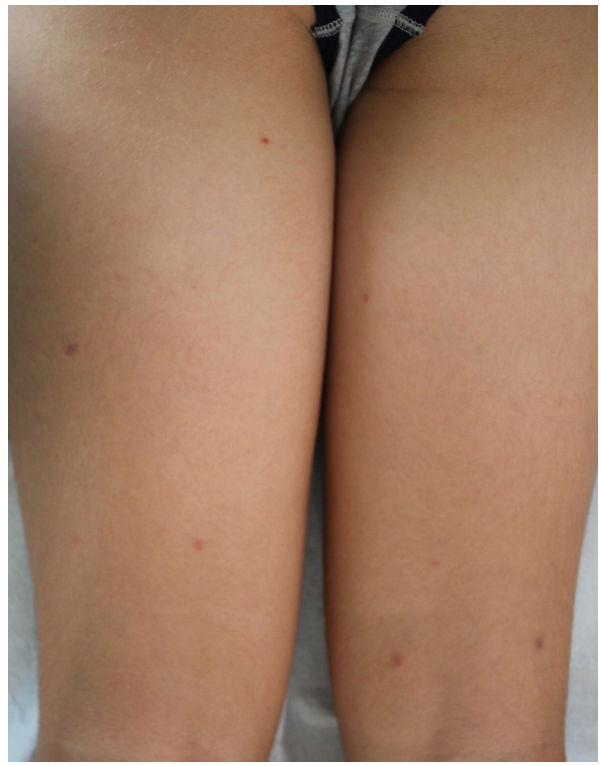

Fig. 2

Fig. 1, 2: 3-13 mm in diameter papules and nodules of the limbs.

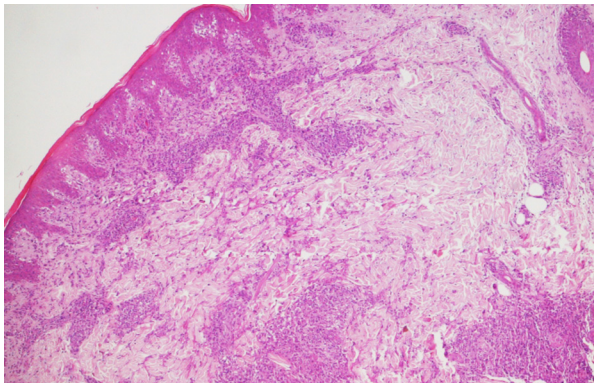

Fig. 3

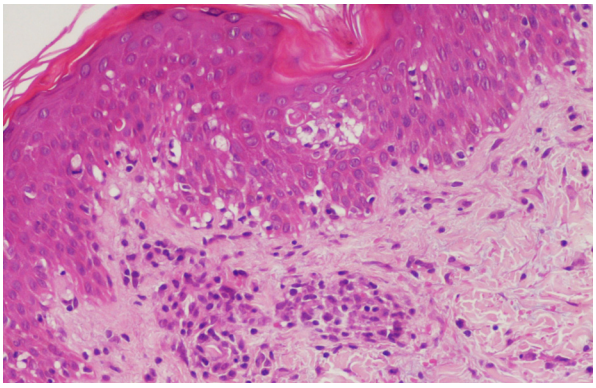

Fig. 4

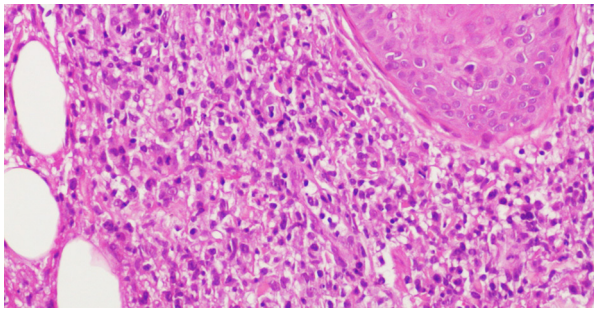

Fig. 5

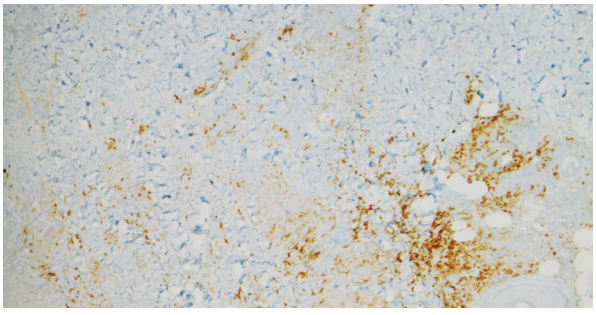

Fig. 6

Fig. 3, 4, 5, 6: Epidermotropic lymphoid infiltrate (H&E, Fig. 3, 40x, Fig. 4, 100x) which in the dermis is arranged around the vessels and appendages (Fig. 5, H&E, 100x). In Fig. 6 (100x) positivity for CD30.

vessels and hair follicles; in depth at the limit between the dermis and hypodermis, the perifollicular infiltrate assumed a massive appearance (Fig. 5). The infiltrate consisted of CD3+ lymphocytes, with CD4+ being more numerous than CD8+; CD20+ were almost lacking; there was a fair amount of CD30+ cells, especially at the level of periadnexal massive infiltrate (Fig. 6).

The pathologist suggested a primary cutaneous T-cell lymphoproliferative disorder that did not allow a clear differential diagnosis between mycosis fungoides, anaplastic large cell lymphoma and lymphomatoid papulosis. Laboratory examinations and abdomen ultrasound were normal.

The presence of papular lesions and small nodules occurring in subintrant crops, not preceded by persistent patches, the less than 2 cm in diameter nodules and the spontaneous regression of all the lesions led us to diagnose **type B lymphomatoid papulosis**. We recommended symptomatic therapy of the ulcerated lesions and periodic clinical-hematological monitoring.

Discussion. In the WHO-EORTC classification (6) lymphomatoid papulosis (LP) is included among the primary cutaneous lymphomas; this term refers to non-Hodgkin's lymphomas which at the time of diagnosis do not present extracutaneous manifestations; after the gastro-enteric system, the skin is the most frequent site of extranodal lymphomas (5).

Primary cutaneous lymphomas (PCL), despite being made up of a cell population comparable to that of lymph node lymphomas, have a clinical behavior significantly different from lymph node lymphomas localized secondarily in the skin, because they remain confined to the skin for a long time or

always and often regress spontaneously; therefore PCLs require a different therapy. PCLs are distinguished in T-cell PCLs, which make up 80%, and B-cell PCLs. Among the T-cell PCLs, after mycosis fungoides, CD30+ lymphoproliferative disorders (6) are most common: the latter include LP and anaplastic large cell lymphoma, that represent parts of the same disease spectrum, are not histologically distinguishable and differ essentially because skin lesions do not heal spontaneously in lymphoma unlike those in lymphomatoid papulosis. The primary lesion of LP is a papule that can enlarge to become a nodule; this usually does not exceed 2 cm in diameter; papules and nodules can become exuding and ulcerate; they usually come in subintrant crops of 1-10 elements and spontaneously regress in 3-6 weeks (1). LP can last from a few months up to 40 years (5).

In contrast to the rather monomorphic and repetitive clinical features, histology shows highly variable patterns that recall different types of primitive cutaneous T lymphomas; in the same patient different histological types can be observed at the same time or at a later time (3).

The medium-term prognosis of LP is good with a 10-year survival close to 100% (5). The risk of developing another lymphoma in adults with lymphomatoid papulosis is 10-20% (2) and mycosis fungoides is the most frequently associated lymphoma. In children, the risk is lower: in a recent meta-analysis of 251 children with lymphomatoid papulosis (4), the incidence of lymphomas is 5.6%, but the most frequent lymphoma is anaplastic large cell primary cutaneous lymphoma.

In most cases, symptomatic therapy is sufficient; topical corticosteroids may be useful in lesions of exposed areas to limit the duration of cosmetic damage.

Conclusion. We presented a rare case of **type B lymphomatoid papulosis** to underline that in these cases the histological examination directs towards the diagnosis of lymphoproliferative disease. However, the clinical features and course decide the prognosis of the disease.

Conflicts of interest

The Author declare that he has no conflicts of interest.

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