Volume 20 Number 11
November 2014

Case Presentation

Unusual CD8 positive lymphomatoid papulosis in childhood

Sandra Mateo MD[1], Igor Vázquez Osorio MD[1], María José Seoane PhD[1], José Manuel Súarez-Peñaranda PhD[2], Hugo Vázquez Veiga PhD[1]

Dermatology Online Journal 20 (11): 5

[1]Department of Dermatology. Complejo Hospitalario Universitario, Santiago de Compostela. La Coruña, Spain.

[2]Department of Pathology. Complejo Hospitalario Universitario Santiago de Compostela. La Coruña, Spain.

Correspondence:

Sandra Mateo, MD
Departamento de Dermatología
Hospital Gil Casares
C/. Choupana, s/n
15703 Santiago de Compostela, La Coruña, Spai
Phone: 00 (34) 981956021
Cell phone: 00 (34) 659421959
Fax: 00 (34) 981956021
E-mail: sandramateosuarez@gmail.com

Abstract

Lymphomatoid Papulosis (LyP) is a rare disorder characterized by a self-healing eruption of papules and small nodules with histopathologic features mimicking a cutaneous T-cell lymphoma CD 30+. We report a 15-year-old girl with CD8+ T-cells, an unusual phenotype in this disease. The clinical and pathological differential diagnoses are discussed.

Key words: Lymphomatoid Papulosis, CD 30+, lymphoproliferative disorders, cutaneous T-cell lymphoma

Introduction

Lymphomatoid Papulosis (LyP), a skin disease within the spectrum of the CD30+ cutaneous lymphoproliferative disorders, has been rarely reported in children. Expression of CD8+ in tumor cells is unusual and very few cases have been described in childhood.

Case synopsis

A previously healthy 15-year-old girl presented to our department for a 2-month history of asymptomatic, small, red papules over her trunk and limbs. She did not have any other symptoms and physical examination was otherwise normal.

She had been previously treated with topical corticosteroids without success. Skin biopsy showed an infiltrate of atypical lymphoid cells with epidermotropism displaying the following phenotype CD3+, CD8+/CD4-, CD30+. Cytotoxic markers (TIA-1 and granzyme-B) were also positive.

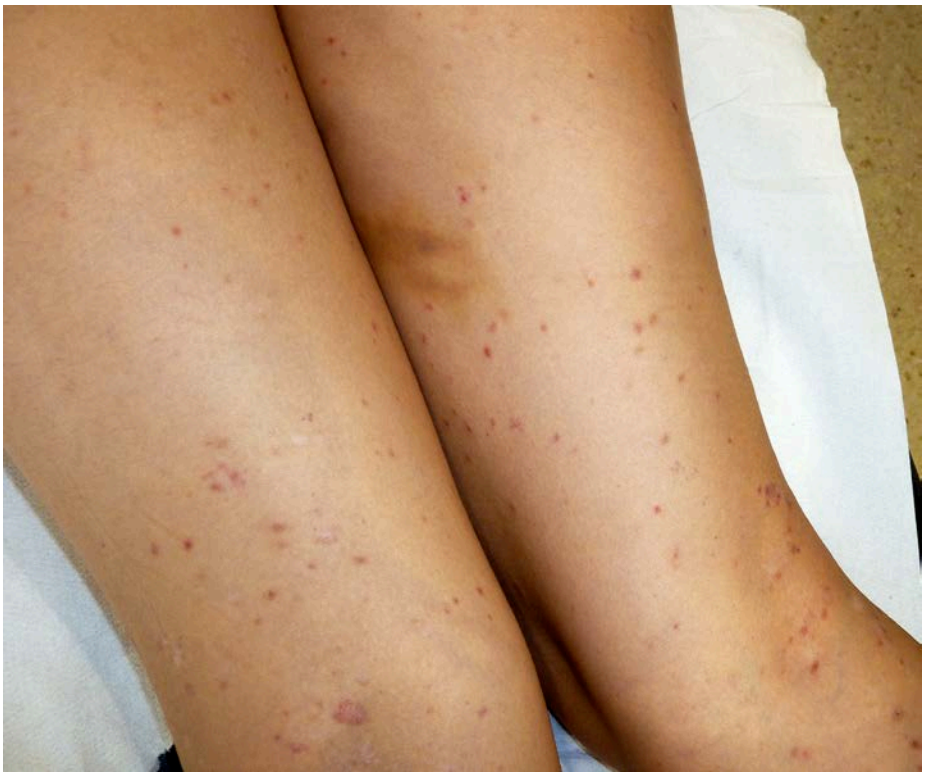

Figure 1. Asymptomatic, small, red papules over her lower extremities

The patient underwent further diagnostic investigations including laboratory blood test and chest X-Ray. No systemic disease was found. Skin lesions regressed without treatment in 4 weeks. No new lesions were identified at a one-year follow-up. The combination of clinical and histological features was consistent with the diagnosis of LyP.

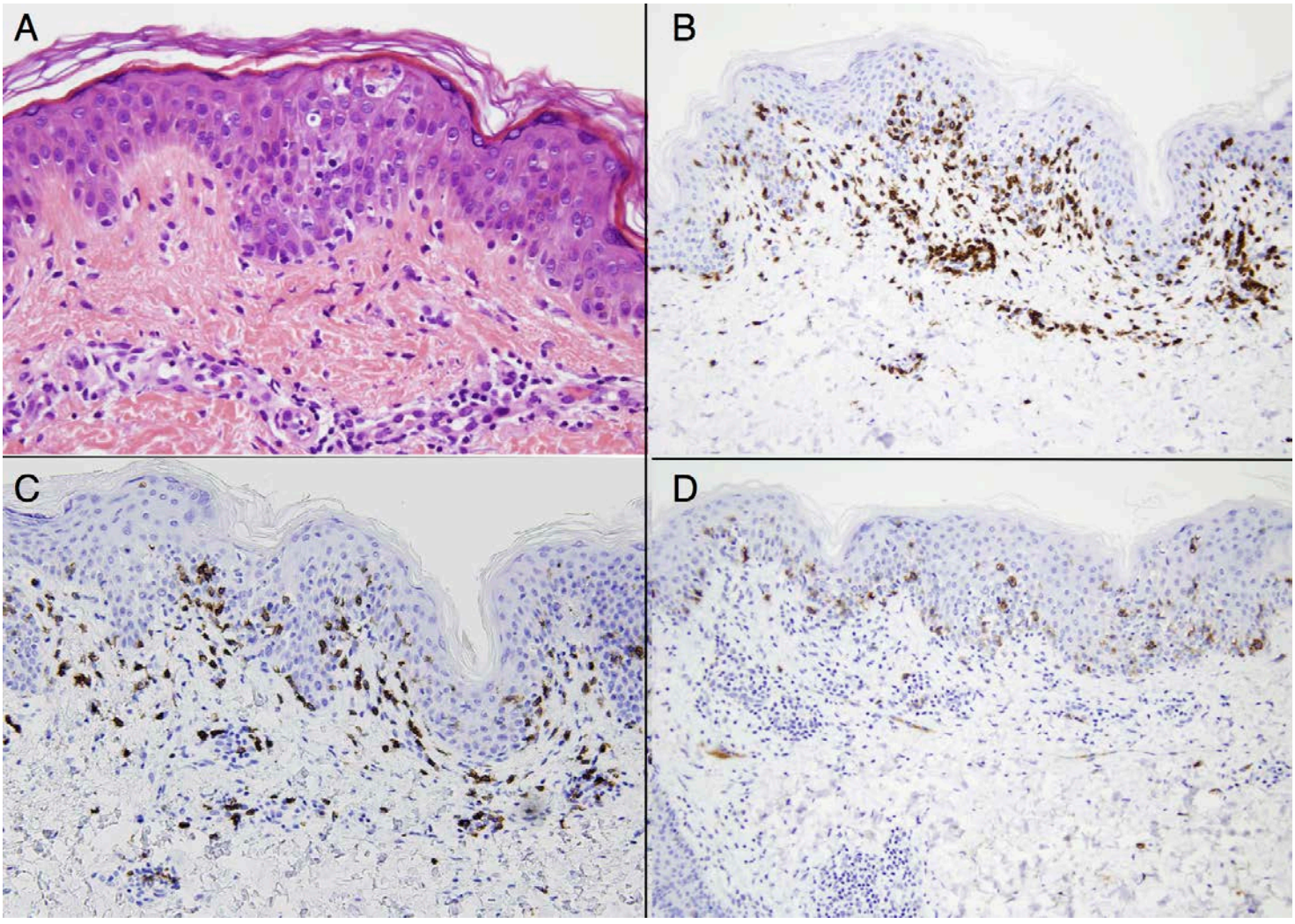

Figure 2. Histopathological examination shows a moderately dense infiltrate of small lymphocytes with striking epidermotropism (A). Immunohistochemical study demonstrates staining with CD3 (B), CD8 (C) and CD30 (D).

Discussion

LyP in children is very rare and it has not been well studied. The clinical and histopathological features are similar to those seen in adults and CD4+ type A LyP is the most frequent variant [1]. On the other hand, cases with CD8+ phenotype are highly unusual, but some examples of the three classic subtypes have been reported [2,3]. Recently, a fourth type has been described, type D LyP, characterized by epidermotropic CD8+ lymphoid cells. Only two cases of the last have been reported in children [4,5].

In our case, although T-cells showed clear epidermotropism, the quantity of T-cells in the epidermis was not enough for establishing a type-D LyP, as it was originally described by Saggini. However, it cannot be completely ruled out as lesions were regressing when biopsied. Alternatively, a type B LyP could be considered as the more appropriate diagnosis [6].

CD8+ LyP must be differenced from other CD8+ lymphomas such as cytotoxic CD8+ lymphoma and a subset of mycosis fungoides because of the different behavior and prognosis of these conditions. This may be difficult based on histopathology alone and clinicopathologic correlation is essential [1].

Given the self-healing character of LyP, treatment is not usually necessary except for cosmetic reasons. Most cases resolve spontaneously, although they can leave some residual scarring. Therapies that have been described in children include topical and systemic steroid, systemic antibiotics, and ultraviolet light [1].

Classical subtypes of LyP are associated with a lifelong increased risk of malignant lymphoma. In adults, 5 to 20 percent of cases of LyP may be preceded, associated with, or followed by a malignant lymphoma. There have been rare cases of lymphoma described in pediatric patients with LyP, but the exact incidence in this age group is unclear. Because of this potential risk, long term observation of patients is required [1].

Conclusions

This case is an example of the uncommon CD8+ LyP in children. This variant of LyP is often misdiagnosed as a aggressive lymphoma. It is important to recognize this subtype of LyP in order to avoid unnecessarily aggressive diagnostic or therapeutic measures.

References

1. Nijsten T, Curiel-Lewandrowski C, Kadin ME. Lymphomatoid papulosis in children: a retrospective cohort study of 35 cases. Arch Dermatol. 2004; 140(3): 306-12. [PMID: 15023774]
2. de Souza A, Camilleri MJ, Wada DA, Appert DL, Gibson LE, el-Azhary RA. Clinical, histopathologic, and immunophenotypic features of lymphomatoid papulosis with CD8 predominance in 14 pediatric patients. J Am Acad Dermatol. 2009; 61(6): 993-1000. [PMID: 19577330]
3. Aoki E, Aoki M, Kono M, Kawana S. Two cases of lymphomatoid papulosis in children. Pediatr Dermatol. 2003; 20(2): 146-9. [PMID: 12657013]
4. Saggini A, Gulia A, Argenyi Z, et al. A variant of lymphomatoid papulosis simulating primary cutaneous aggressive epidermotropic CD8+ cytotoxic T-cell lymphoma. Description of 9 cases. Am J Surg Pathol. 2010; 34(8): 1168-75. [PMID: 20661014]
5. McQuitty E, Curry JL, Tetzlaff MT, Prieto VG, Duvic M, Torres-Cabala C. CD8-positive ("type D") lymphomatoid papulosis and its differential diagnosis. J Cutan Pathol. J Cutan Pathol. 2014; 41(2): 88-100. [PMID: 24118002]
6. Plaza JA1, Feldman AL, Magro C. Cutaneous CD30-positive lymphoproliferative disorders with CD8 expression: a clinicopathologic study of 21 cases. J Cutan Pathol. 2013 Feb; 40(2): 236-47. [PMID: 23189966]