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LETTER - CLINICAL

Acquired lymphangiectasia of the lip in a patient with Crohn's disease☆

Dear Editor,

Acquired lymphangiectasia is a rare complication of lymphatic obstruction. It has been described in association with malignancy and granulomatous diseases, namely orofacial granulomatosis, Crohn's Disease (CD), and tuberculosis. It must be distinguished from a congenital malformation, generally referred to as lymphangioma.[1] Herein we describe a case of acquired lymphangiectasia of the lip in a girl with CD, to our knowledge the first case of this association in pediatric age.

An 11-year-old girl was referred to our Dermatology Department due to persistent cheilitis and gingivitis since she was 4 years old. A therapeutic attempt with gingivoplasty had been performed at the age of 8 without success. No histology was available from the intervention. Later, at the age of 9, she had been diagnosed with Crohn's disease after intestinal biopsy and was treated with azathioprine and infliximab. However, the cheilitis persisted.

Physical examination showed crusty lesions and flaccid blisters on both lips, as well as erythema with fissures on the gums and jugal mucosa (Fig. 1 A and B). A biopsy of the labial mucosa was performed.

Histology showed epidermal hyperplasia without atypia with lichenoid pattern inflammation and lymphangiectasia (Fig. 2A). The presence of numerous podoplanin-positive subepithelial dilated vessels with a lymphatic appearance is noteworthy (Fig. 2B). Further investigations, including patch testing with the European standard and cosmetics series, and a facial MRI were normal. Topical corticosteroid treatment was prescribed with the intention of reducing the associated inflammation, followed by topical rapamycin 0.2% to decrease the lymphatic component, with no clear improvement after two months.

Granulomatous cheilitis presents as orofacial inflammation secondary to non-necrotizing granulomas. It can present isolated or in the context of granulomatous systemic disease. In children, granulomatous cheilitis is more often associated with CD than in adults, being the first manifestation in 5%–10% of cases of inflammatory bowel disease.[2,3]

Acquired lymphangiectasia in CD is thought to develop as a consequence of chronic granulomatous inflammation. It is described most often in the genital area. In a literature review, we found four cases of acquired orofacial lymphangiectasia associated with CD (Table 1).[1,2,4] As in the case presented here, in two of these patients no granulomas were observed on histology, and in another case, they were only observed in the second biopsy. The medical treatment could have reduced the inflammation, leaving only the residual lymphangiectasia.[1,2,4]

In the absence of granulomatous inflammation, lymphangiectasia can be treated with cryotherapy, sclerotherapy, surgical excision, laser ablation, or photocoagulation.[1] In our case, topical rapamycin was attempted with no results at 2 months. No cases of acquired lymphangiectasia have been successfully treated with topical rapamycin, but 0.1% topical sirolimus has been effective in cutaneous microcystic lymphatic malformations in children and adults.[5]

In conclusion, the presence of orofacial acquired lymphangiectasia could be an early marker of inflammatory bowel disease even in the absence of granulomatous cheilitis or digestive symptoms. Its diagnosis could help in the early detection of CD.

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Authors' contributions

Ana Llull-Ramos: Concepcion; design; acquisition of data; drafting the article; analysis of data; final approval of the version to be published.

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Inés Gracia-Darder: Acquisition of data; analysis of data; final approval of the version to be published.

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Conflicts of interest

None declared.

☆ Study conducted at the Department of Dermatology, Son Espases University Hospital, Palma de Mallorca, Spain.

Table 1 Summary of cases of acquired lymphangiectasia of the lip associated with Crohn's disease.

N (Ref)	Year	Age/Sex	LIP histological findings	Treatment	Of outcome
1[1]	2017	18/F	Widely dilated superficial lymphatic channels	Azathioprine, mesalazine, infliximab, 0.03% topical tacrolimus and lip cryotherapy	Good cosmetic result
2[1]	2017	33/M	Superficial dilated lymphatics and patchy inflammation	Triamcinolone injections, 1% topical hydrocortisone cream, oral mesalazine	No treatment needed
3[2]	2020	29/F	1st biopsy: superficial dilated vessels and dense inflammatory infiltrate with a tiny granuloma 2nd biopsy: superficial lymphangiectasia, inflammatory infiltrate and small granuloma	Infliximab	Improvement
4[4]	2021	30/F	1st biopsy: proliferation of ectatic lymphatic channels 2nd biopsy: granulomata adjacent to dilated lymphatic channels	Methotrexate and infliximab	Improvement
5 (present case)	2021	11/F	Lichenoid pattern inflammation and lymphangiectasia	Azathioprine, infliximab, topical corticosteroids and 0.2% topical rapamycin	No clear improvement

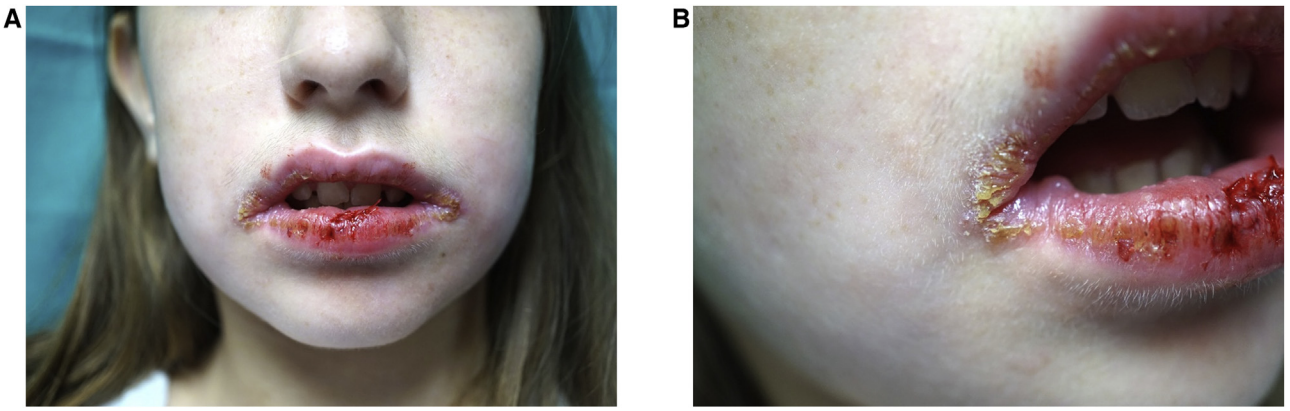

Figure 1 Clinical appearance of the girl's lips (A and B).

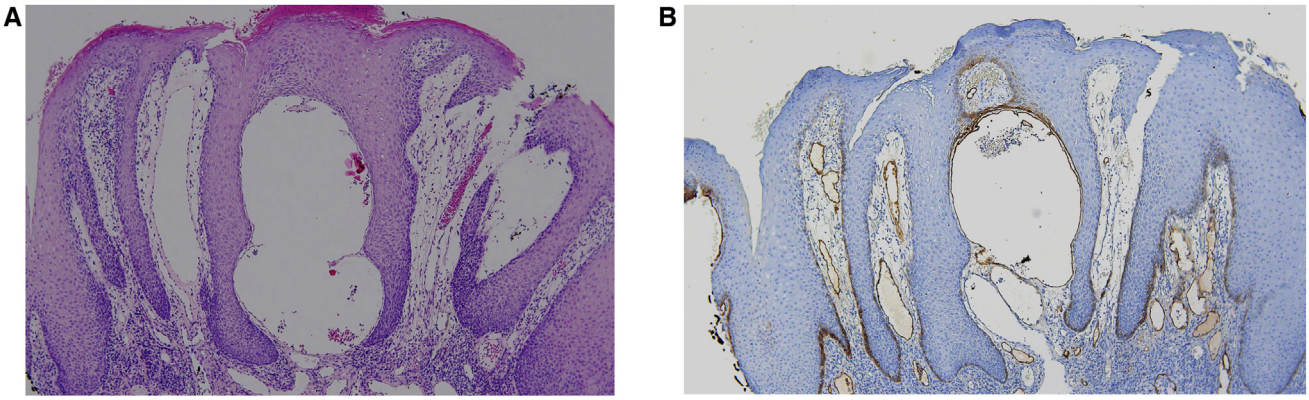

Figure 2 (A) Labial mucosa fragment lined by squamous epithelium with marked acanthosis, spongiosis, hyperkeratosis and no keratinocyte atypia, with a dense lymphoplasmacytic infiltrate in the deeper areas without granulomas, with focal lichenoid change (Hematoxylin & eosin, ×100). (B) Positive immunostaining for podoplanin in subepithelial lymphatic vessels (Hematoxylin & eosin, ×100).

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Ana Llull-Ramos *, Juan Garcías-Ladaria, Inés Gracia-Darder, Aniza Giacaman

Department of Dermatology, Son Espases University Hospital, Palma de Mallorca, Spain

* Corresponding author.
E-mail: anallullramos@gmail.com (A. Llull-Ramos).

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Dermoscopy of pityriasis lichenoides et varioliformis acuta (PLEVA)☆

A 24-year-old woman without significant past medical history, presented with a 12-month history of pruriginous erythematous-violaceous desquamative papules and vesicles, which appeared in crops, initially on her thighs and legs, with latter involvement of arms, forearms, and torso. Some lesions suffered central necrosis, and disappeared from 2 to 4 weeks, leaving depressed scars, with the continuous development of new crops. She referred a vitamin B-complex intramuscular injection 2 weeks preceding the start of the clinical picture, and 6 months previously she had started consuming combined oral contraceptives.

Upon clinical examination she presented several erythematous-violaceous small papules, some with a necrotic central crust, and multiple varioliform scars, with involvement of lower extremities, proximal upper extremities, chest, and abdomen (Fig. 1). A skin biopsy was performed, which showed typical findings of PLEVA, confirming the diagnosis (Fig. 2).

A dermoscopic evaluation of 14 active lesions was made, with contact and no-contact polarized light dermoscopy. Vessels were found in 100% of them, with diverse morphologies (Fig. 3): all the evaluated lesions had dotted vessels, and some had linear irregular and/or glomerular vessels. Non-blanchable reddish globules were not detected. Combination of these morphologies was frequent, polymorphous vessels were present in 79% of the lesions, with dotted and irregular linear vessels being the most common combination. Vessel arrangement was peripherical in most of the lesions, although some showed uniform, clustered, or central distribution. Scales were found in most of them, white in color, with variable arrangement: the majority showed a focal location disposed in a ring-like or targetoid fashion (43%), being located between a central red-brownish clod, and a peripheral ring of vascular structures immersed in a background which generally was pink or purple, and less frequently orange or salmon-colored.

Two main patterns were identified: a typical target pattern, where the disposition of structures gave an image that resembled a typical target or iris lesion of erythema multiforme, consisting of three concentrical zones: central clod, intermediate ring of white scale, and peripheral vascular ring (Fig. 4). An atypical target pattern was found in other lesions, which had only two concentrical zones: a central clod, white scale or structureless area, surrounded by a vascular ring (Fig. 5).

Additionally, two varioliform scars were evaluated with dermoscopy, both showing light brown peripheral structureless areas with a central hypopigmented structureless area, also with a targetoid appearance, but without any vascular structures nor scales.

Pityriasis lichenoides refers to a spectrum of disease that includes three main variants: Pityriasis Lichenoides et Varioliformis Acuta (PLEVA), Pityriasis Lichenoides Chronica (PLC) and febrile ulceronecrotic Mucha-Habermann disease.[1] As a spectrum, these conditions may sometimes overlap even though they usually have distinct clinical, histological, and dermoscopic features.[2] PLEVA usually presents as asymptomatic erythematous macules that evolve into polymorphous erythematous papules with a necrotic center, and then disappear, leaving varioliform scars and dyschromic areas.[1,3]

On dermoscopy, some authors have described a well-defined peripheral ring of vessels with a targetoid appearance, with polymorphous vessels, mainly dotted, glomerular, and/or linear irregular, as were seen in the patient. Ankad et al.[3] proposed that these findings may correlate to blood vessels dilation and microhemorrhages in the papillary dermis.[1,2,4] In two of five articles that reviewed dermoscopy of PLEVA, the targetoid pattern of vessels was not observed, though in one of them both peripheral dotted and glomerular vessels were reported.[4,5] Other authors have described non-blanchable reddish globules, which were not identified in the present case.[4]

☆ Study conducted at the Specialized Diagnostic Clinic VID, Medellin, Colombia.