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IMAGES IN CLINICAL PRACTICE

SCRATCHING THE SURFACE - WHEN SKIN LESIONS ARE MORE THAN THEY APPEAR

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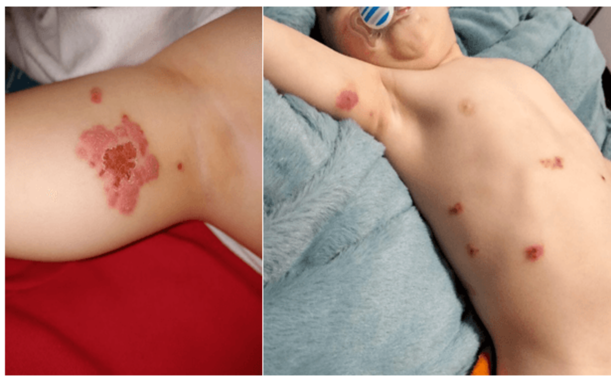
A previously healthy 17-month-old boy was referred for pediatric consultation due to recurrent skin lesions. Five months earlier, he presented a pruritic vesicular rash in the thorax, abdomen and arm, which resolved spontaneously. Four months later, he developed a similar rash. For suspected herpes zoster infection, his family doctor started treatment with acyclovir without any improvement. At the time, there was no weight loss, restricted growth or gastrointestinal manifestations. There was also no history of autoimmune disease in the family. At the pediatric consultation, the child presented the skin lesions demonstrated in Image 1. His mother also reported diarrhea in the previous week, with no other associated symptoms. Blood tests were performed, including total IgA (69.3 mg/dL, normal rate 17-137 mg/dL), anti-tissue transglutaminase IgA (>128 U/mL), anti-gliadine IgG and IgA (188 U/mL, 142 U/mL) and anti-endomysium antibodies (1/320, normal rate < 1/10), which were positive. After discussing diagnostic pathways with the family, the patient underwent upper endoscopy with biopsies of the intestinal mucosa (bulb and distal duodenum) that presented atrophy of intestinal villi, crypt hyperplasia and inflammatory infiltrate, with type 3B Marsh-Oberhuber classification. The patient started a gluten-free diet with regression of the cutaneous manifestations and normalization of the antibodies.

Figure 1. Skin lesions observed on the chest, abdomen and upper arm of the patient.



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What is the diagnosis?

Celiac disease is an immune-mediated enteropathy, precipitated by the ingestion of cereals containing gluten in their constitution by genetically susceptible individuals.[1] It is characterized by a state of chronic inflammation of the intestinal mucosa, with the first line of treatment being a gluten-free diet.[1] The clinical manifestations of celiac disease are very variable, including typical symptoms - such as abdominal pain, diarrhea, growth retardation or weight loss - and atypical symptoms - such as alopecia, anemia, neurologic manifestations (including epilepsy), arthritis and dermatitis herpetiformis.[1,2,3] Dermatitis herpetiformis usually manifests itself in the form of an itchy papulovesicular rash that mainly affects the extensor surfaces of the limbs, scalp, neck and buttocks.[4,5] Although it can appear in any age group, it is diagnosed mainly in men aged between 30 and 50 years, being a rare symptom in pediatric age.[6] Its diagnosis becomes particularly challenging due to the similarities it shares with other exanthematous diseases, such as herpes zoster.[3,4] Although celiac disease has an estimated prevalence of 1% in the general population, with a progressive increase in recent decades, this pathology remains underdiagnosed, especially when it manifests itself in an atypical or low-symptomatic form.[3] The diagnostic method of choice for celiac disease is intestinal biopsy, with the predominance of intraepithelial lymphocytes, villous atrophy and crypt hyperplasia being typical histological findings of the disease.[2] On another hand, the diagnose of dermatitis herpetiformis is confirmed by perilesional skin biopsy typically showing neutrophilic micro-abscesses in dermal papillae and granular immunoglobulin A deposits on immunofluorescence examination.[6] Several studies demonstrate that anti-TG2 IgA antibodies, used in screening for celiac disease, are also frequently positive in individuals with dermatitis herpetiformis. Anti-TG3 IgA antibodies are also positive in most dermatitis herpetiformis patients but their applicability in the diagnosis of dermatitis herpetiformis is not completely understood.[6] There is also an important relationship between the diagnosis of dermatitis herpetiformis and the presence of HLA-DQ2 or DQ8 haplotypes, suggesting that HLA testing could play an important role in excluding DH if these haplotypes are not present, preventing unnecessary further investigation.[7] We intend, by sharing the clinical case described, to raise awareness of the possibility of dermatitis herpetiformis as the first clinical manifestation of celiac

disease in pediatric age, even though it is an uncommon manifestation in this age group. The recognition of skin lesions suggestive of dermatitis herpetiformis is essential for a timely diagnosis and appropriate treatment in these cases.

Compliance with Ethical Standards
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Conflict of Interest: None

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