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CASE REPORT

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Atypical erythema *annulare centrifugum* in a child with celiac disease

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Abstract

Herpetiformis dermatitis is the best characterized extraintestinal manifestation of celiac disease (CD). However, other chronic heterogeneous skin lesions have been associated with CD and should be considered in the differential diagnosis.

KEYWORDS

celiac disease, erythema *annulare centrifugum*, gluten-free diet, pediatrics, skin biopsy

1 | CLINICAL CASE

We report the case of atypical erythema *annulare centrifugum* (EAC) in a child with celiac disease (CD), which entirely resolved with the gluten-free diet. Atypical CD may appear with herpetiformis dermatitis, which is the best characterized and established extraintestinal manifestation. However, different heterogeneous skin diseases are associated with CD.

A 9-year-old Indian boy was admitted to our pediatric clinic with a history of chronic, recurrent skin lesions that appeared in the first year of life. He was evaluated for atopic dermatitis in his country, and topical corticosteroids have been prescribed without any clinical improvement. His clinical history was negative for growth failure, gastrointestinal symptoms, malabsorption, and infectious, autoimmune, or chronic diseases. Moreover, his familiar history was negative for noticeable systemic, autoimmune, or skin diseases.

The physical examination showed diffuse, nonpruritic, annular, cutaneous erythematous lesions with scaly borders and central clearings, mainly located on the trunk, abdomen, neck, and extremities (Figure 1A–C). We performed thyroid function tests, autoimmune screening, and serum immunoglobulins, which were normal. Complete blood count revealed a moderate iron deficiency anemia (Hb 9 g/dl, transferrin saturation 4%), while the screening for celiac disease (CD) tested positive with high levels of transglutaminase antibodies (TTGA, 156 UI/ml) and positive anti-endomysial antibodies. According to ESPGHAN 2012 guidelines for CD,[1] the patient underwent upper gastrointestinal endoscopy with biopsies. The histology confirmed the diagnosis of CD (Marsh score III). The skin biopsy demonstrated superficial perivascular dermal lymphohistiocytic infiltrate, hyperkeratosis, and focal epidermal spongiosis without IgA deposits identified at the

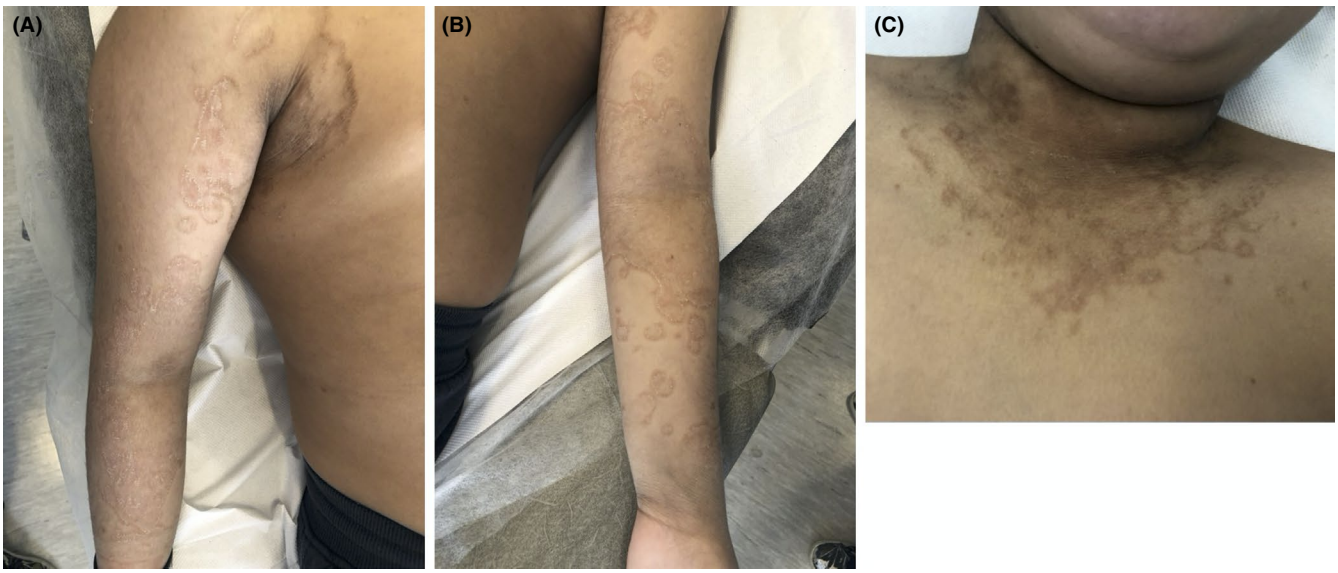

FIGURE 1 Nonpruritic, annular, erythematous lesions with scaly borders and central clearings, located on the patient's arms and axilla (A, B) and neck (C)

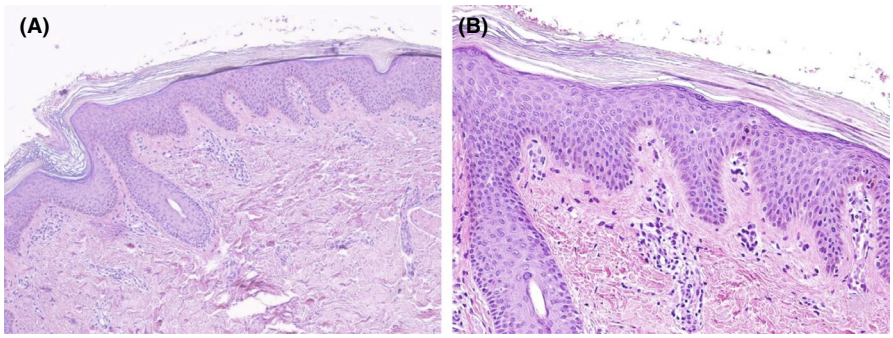

FIGURE 2 Histopathological findings show superficial perivascular dermal lymphohistiocytic infiltrate (A) with hyperkeratosis and focal epidermal spongiosis (B) (hematoxylin and eosin, magnification 20×)

direct immunofluorescence (Figure 2A,B). The exclusion of other dermatoses and the supportive history of chronic annular desquamative lesions allow us to diagnose erythema *annulare centrifugum* (EAC) associated with CD. A gluten-free diet (GFD) was started, and after three months, the child showed complete remission of his skin lesions with minimal postinflammatory pigmentation after three months of GFD (Figure 3A–C).

2 | DISCUSSION

Erythema *annulare centrifugum* is an uncommon inflammatory skin lesion associated with a wide variety of etiologies, such as infections, malignancies, autoimmune diseases, drugs, and foods.[2] However, in many patients, the exact etiology cannot always be identified. EAC typically appears with erythematous polycyclic skin lesions or urticarial papules that progressively expand centrifugally with a central clearing, forming annular or arcuate lesions.[2] Pruritus may also be present. EAC can be histologically classified as superficial or deep and generally shows perivascular infiltrates with variable changes in the papillary dermis and epidermis.[2,3] The differential diagnosis of EAC is broad and is based on the clinical appearance, histological findings, and the exclusion of other dermatoses (Table 1).[4] In our case, the histopathological and microbiological results allow us to exclude infections and inflammatory skin diseases.

Celiac disease is an autoimmune enteropathy elicited by gluten that affects the small intestine, characterized by the variable combination of gluten-dependent symptoms, specific antibodies, and genetic predisposition.[5] Herpetiformis dermatitis is the first well-established extraintestinal manifestations of CD, but other skin diseases, such as psoriasis linear IgA bullous dermatosis, hereditary angioneurotic edema, atopic dermatitis, cutaneous vasculitis, and necrolytic migratory erythema, are also described in CD patients.[5] Most of the skin manifestations of CD improve following a gluten-free diet.[6]

We described a rare pediatric case of EAC associated with CD that surprisingly improved with GFD, suggesting

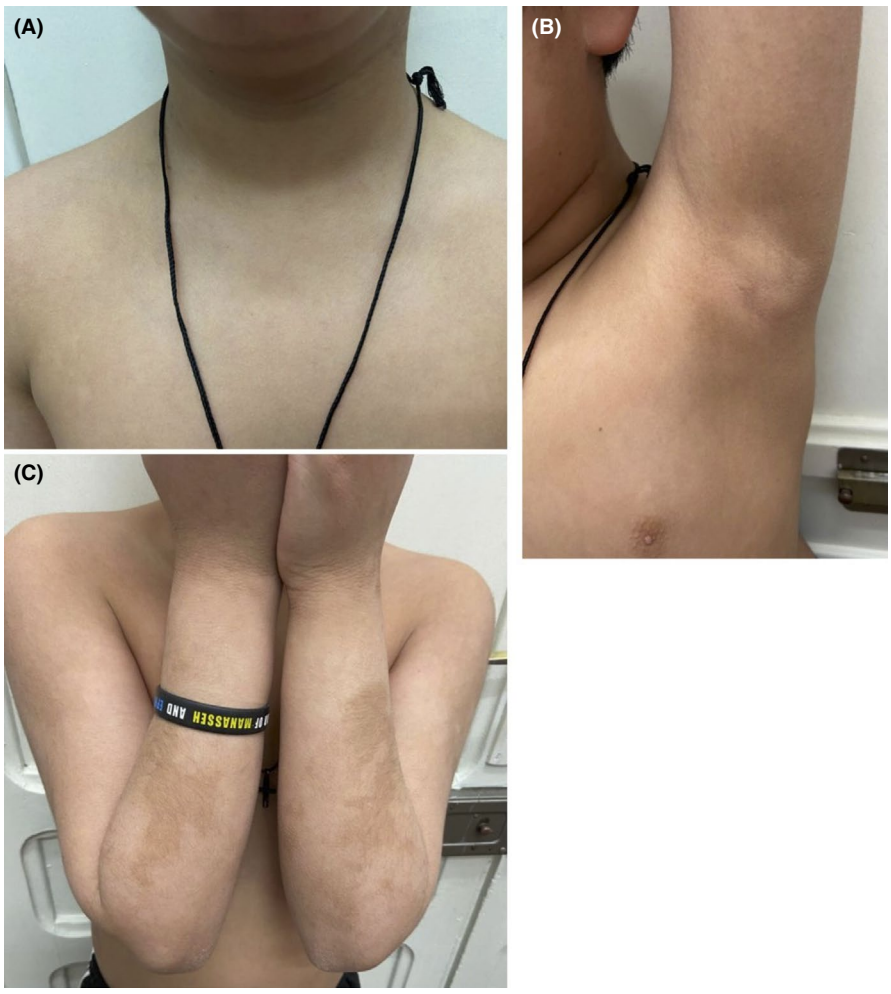

FIGURE 3 Complete remission of the lesions on the neck (A) and arms (B, C)

TABLE 1 Differential diagnoses of annular skin lesions

Differential diagnosis of annular lesions	Differential diagnosis of CD-associated annular skin lesions
Tinea corporis	Annular psoriasis
Annular urticaria	*Rosacea*
Allergic contact dermatitis	Skin malignancies
Erythema *chronicum migrans*	Atopic dermatitis
Erythema *marginatum*	
Erythema *gyratum repens*	
Granuloma annulare	

that the differential diagnosis of atypical, chronic dermatitis unresponsive to conventional therapies should also include the exclusion of CD. Although autoimmunity might be considered a putative common pathogenetic mechanism, the association between CD and EAC is currently limited to a few case reports.

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None. Published with written consent of the patient.

CONFLICT OF INTEREST

The authors declared no conflicts of interest.

AUTHOR CONTRIBUTIONS

MV and MDF: wrote the entire manuscript. DS and SI: reviewed the literature and collaborated to writing of the manuscript. AL and VB: supervised the manuscript drafting. All authors: approved the final version of the manuscript and agreed to be accountable for all aspects of the work. Questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

ETHICAL APPROVAL

Informed consent was obtained from the patient to publish this case report.

DATA AVAILABILITY STATEMENT

Not applicable.

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