


CASE REPORT

An unusual presentation of celiac disease in adult patient

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Abstract

Pyoderma gangrenosum is among the exceptional extra-intestinal manifestations of celiac disease. We report a case of a 52-year-old patient who presented with pyoderma gangrenosum that turned out to be the initial presentation of celiac disease.

KEYWORDS

celiac disease, extra-intestinal manifestations, pyoderma gangrenosum

1 | INTRODUCTION

Celiac disease (CD) is an immune-mediated enteropathy triggered by dietary gluten sensitivity characterized by a large variety of atypical presentations, especially in adults, with several extra-intestinal manifestations (EIM), including skin diseases.^{1,2} Pyoderma gangrenosum (PG) is among the exceptional EIM of CD.^{3,4} We here report a case of a 52-year-old patient who presented with PG that turned out to be the initial presentation of CD.

2 | CASE HISTORY AND EXAMINATION

A 52-year-old patient, with a family history of CD, presented to our dermatology department with painful ulceration of the left leg, evolving in a centrifugal way for 3 months. Skin examination showed large superficial ulceration with pus discharge and active inflammatory borders (Figure 1). Achromic bilateral and symmetrical macules were noted on the legs (Figure 2).

3 | INVESTIGATIONS AND DIAGNOSIS

The skin biopsy showed a neutrophilic dermal infiltrate with signs of vasculitis, in favor of PG. Upper GI endoscopy showed loss of kicking folds in the descending duodenum and the histopathology revealed complete villous atrophy. The serology markers were positive for antigliadin, antireticuline, and anti-endomysial antibodies. Endoscopy, biological, and morphological analyses ruled out systemic diseases frequently associated with PG. Based on the above findings, the diagnosis of PG and vitiligo associated with CD was made.

4 | MANAGEMENT AND FOLLOW-UP

The patient was managed with a gluten-free diet, prednisone 0.5 mg/kg daily, and LED therapy. Complete healing of the skin lesion was obtained after 3 months of treatment.

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FIGURE 1 Large superficial ulceration with pus discharge and active inflammatory borders

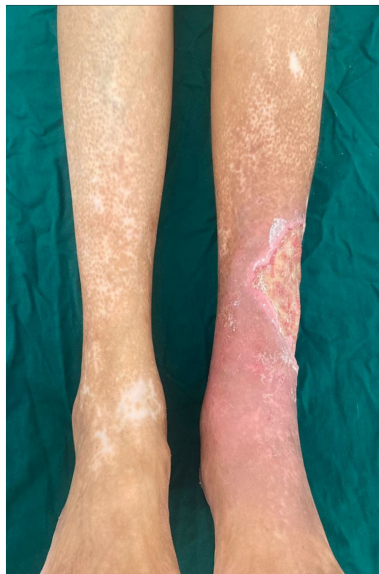


FIGURE 2 Achromic bilateral and symmetrical macules were noted on the legs.

5 | DISCUSSION

The current case is being reported given the clinical presentation of CD could be misleading especially in adults. In fact, our patient did not have any gastrointestinal symptoms; she sought and attracted medical attention due to skin disorders, namely PG and vitiligo. CD is thought to be underdiagnosed in adulthood in part owing to the fact that physicians are not aware of the high prevalence of EIM in this age group while many patients may seek medical advice because of them.

In the last years, growing evidence has documented the involvement of skin diseases among the EIM of CD

which can no longer be considered a simple random.⁴ Besides the well-known association between CD and dermatitis herpetiformis, many other dermatoses are actually more common or show atypical presentation often associated with resistance to standard therapies in those patients.⁴ Among these are autoimmune, allergic, and inflammatory diseases.⁵ Lupus erythematosus, dermatomyositis, Behcet disease, and vitiligo are frequently reported, while prurigo, erythema nodosum, and erythroderma result very rare.^{4,5}

Although rare, the association between CD and vitiligo is well documented. The prevalence of vitiligo in the general population is estimated to be 0.5%–2%. In cohorts of celiac patients, a higher prevalence of 3.8%–9.1% has been noted. CD autoantibodies are more frequent in the serum of vitiligo patients compared with normal individuals. Both of these autoimmune diseases may be stimulated by a common immune system signal triggered by gluten. A convincing argument in favor of this hypothesis was the improvement of vitiligo in CD patients after the introduction of GFD.

In summary, this case report highlighted the importance of considering cutaneous manifestations in CD patients presenting in adulthood. It also adds to the pool of knowledge about the presentation and management of PG in association with CD. Clearly, the CD is probably an underrecognized etiology of PG. The diagnosis of PG should raise the possibility of concomitant CD, since its diagnosis can be challenging and the results of the delay to treat it devastating. On the contrary, the CD should sensitize us to the possible future development of PG.

Pyoderma gangrenosum is an inflammatory neutrophilic dermatosis classically associated with a wide variety of etiologies, including inflammatory bowel disease, rheumatic arthritis, leukemia, and lymphoproliferative disorders.³ In our case, thorough testing excluded concurrent inflammatory diseases frequently associated with PG and leads us to diagnosis an atypical adult CD as PG's etiology.

The association between PG and CD, although unusual, has been previously described.^{3,4,6} Only few occasional observations were described in the literature.^{3,4} CD was often severe, resistant to steroids and immunosuppressive drugs.³ Although autoimmunity might be considered a putative common pathogenetic mechanism, the pathogenic link between the diseases remains unclear. Emiliano Antiga recently proved that IL15 may have an independent role in the pathogenesis of PG as they detected an over-expression of IL15 in the inflammatory infiltrate of PG.⁷ This marker has been long investigated in CD.⁷ Thus, we think that this may suggest a pathophysiological pathway between PG and CD.

6 | DISCUSSION

In summary, this case report highlighted the importance of considering cutaneous manifestations in CD patients presenting in adulthood. It also adds to the pool of knowledge about the presentation and management of PG in association with CD.

AUTHOR CONTRIBUTIONS

Miss. Refka Frioui is the guarantor of the content of the manuscript, included the data and analysis. Dr. Anissa Zaouak contributed to interpretation of data and revision of the manuscript. Dr Wafa Jouini interpreted the histological images. Dr. Samy Fenniche and Dr.Houda Hammami contributed to analysis and interpretation of data, revised it critically for important intellectual content, and final approval of the version to be submitted.

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None.

CONFLICT OF INTEREST

The authors declare that there are no conflicts of interest in this work.

DATA AVAILABILITY STATEMENT

All data generated are included in this published article.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy. The examination of the patient was conducted according to the principles of the Declaration of Helsinki. The authors certify that they have obtained all appropriate patient consent forms, in which the patient gave his consent for images and other clinical information to be included in the journal. The patient understood that

his name and initial will not be published and due effort will be made to conceal his identity, but that anonymity cannot be guaranteed.

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