Gluten-sensitive enteropathy associated with genital lichen simplex chronicus

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A novel presentation of coeliac disease with lichen simplex chronicus that questions the extent to which localized pruritis is investigated.

Case report
A 49-year-old woman presented to the dermatology department with a 10-year history of anogenital itching. The pruritus was often so severe that she would scratch until she drew blood. Her symptoms had been partially controlled with Dermovate NN® cream during the year prior to her presentation but were still impacting heavily on her quality of life. She denied the use of wet wipes, soap or pant liners and had no history or family history of atopy or any other skin disease.

Examination showed shiny, slightly erythematous, heavily excoriated, thickened skin of the perianal area extending onto vulval skin around the posterior fourchette (Figure 1). The architecture of the area was well-preserved. Skin biopsy from the affected skin showed acanthosis of the keratinized skin, focal parakeratosis and mild to moderate chronic inflammation of the superficial dermis with occasional eosinophils and neutrophils in the epidermis. There was no evidence of a lichenoid change or blistering, thus supporting the clinical diagnosis of lichen simplex chronicus.

Further investigations showed a ferritin of 9 ug/L (10–150 ug/L) and haemoglobin of 11 g/dL (12–15 g/dL). Her MCV, blood glucose and thyroid function tests were normal. Despite a three-month course of iron replacement therapy her ferritin level did not improve. Further investigations showed serum tissue transglutaminase antibodies raised at 110 kU/L and subsequent duodenal biopsy revealed subtotal villous atrophy consistent with gluten-sensitive enteropathy.

The patient was commenced on a gluten-free diet and five months later her ferritin level had risen to 32 ug/L. At one-year follow-up her pruritus had resolved and examination of her perineum and vulva revealed normal skin.

Discussion
Gluten-sensitive enteropathy, known as coeliac disease, is thought to affect 1 in 100 Caucasians. Some studies suggest that between 10–15% of cases remain undiagnosed and only 40% are symptomatic.1 The symptoms are often non-specific and include fatigue, weight loss, bloating, abdominal pain, diarrhoea and steatorrhoea. The resultant malabsorption can in turn lead to metabolic derangement, anaemia and vitamin deficiencies.2

Lichen simplex chronicus is a form of neurodermatitis. It arises in apparently normal skin which is repetitively scratched until there is scaling and reactive localized thickening known as lichenification. An itch–scratch cycle of emotional aetiology is thought to underlie the pathophysiology of lichen simplex chronicus.3

Our case suggests an association of genital lichen simplex chronicus with iron deficiency secondary to coeliac disease. This is credible because the introduction of a gluten-free diet combined with iron replacement therapy coincided with the complete resolution of her lichen simplex chronicus.

A number of skin disorders are seen in association with coeliac disease. Dermatitis herpetiformis is the most common but others include diseases such as alopecia, atopic eczema and vitiligo.4 There have also been reported cases of prurigo nodularis in association with coeliac disease.5–9 This is a chronic inflammatory
dermatosis of unknown aetiology, in which a persistent itch and repetitive scratching of the skin leads to the development of excoriated thickened papules and nodules. It has been suggested that malabsorption in untreated coeliac disease leads to the development of prurigo nodularis lesions as they seem to resolve on a gluten-free diet. The presence of an ‘itch–scratch’ cycle in both prurigo nodularis and lichen simplex chronicus makes the two conditions similar and both problematic to treat.3

We report a case of longstanding genital lichen simplex chronicus associated with iron deficiency anaemia and gluten-sensitive enteropathy that resolved completely following treatment with a combination of gluten-free diet and iron supplementation. The patient gave no history of typical gastrointestinal symptoms suggestive of coeliac disease. Her iron deficiency anaemia did not respond to isolated iron replacement therapy and her lichen simplex chronicus was resistant to conventional treatment with potent topical steroids. It is, therefore, very plausible that lichen simplex chronicus was a cutaneous presentation of untreated coeliac disease. To our knowledge this is the first record of genital lichen simplex chronicus in association with untreated coeliac disease. This is an uncommon or possibly under-recognized presentation of coeliac disease, which often results in non-specific symptoms and can remain undiagnosed for many years.1,2

This finding has also potential future implications on the approach of cases with localized pruritus which are not usually investigated to the same extent as patients with generalized itching. Patients suffering from generalized pruritus in the absence of primary skin disease will be investigated for the underlying systemic diseases with renal, liver and thyroid function tests, full blood count, iron studies, serum immunoglobulins and electrophoresis, serum glucose and chest X-ray. In contrast, the localized pruritus which can result in lichen simplex chronicus is rarely investigated. We would, therefore, advocate a similar approach in cases of localized lichen simplex chronicus that proves resistant to conventional treatments.

Summary

Gluten-sensitive enteropathy (coeliac disease) affects around 1% of the population, but due to the heterogeneity of clinical presentation often remains undiagnosed for a long time. We report the case of a 49-year-old woman with genital lichen simplex chronicus that led to the diagnosis of coeliac disease. As her lichen simplex chronicus resolved with treatment of her coeliac disease it is plausible that her lichen simplex chronicus was a cutaneous presentation of untreated coeliac disease. This case highlights a rare or under-recognized presentation of coeliac disease and suggests that in cases of lichen simplex chronicus resistant to conventional treatment, it may be of value to look for underlying causes of pruritus.

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