Case Report Open Access

A Case of a Rare Skin Manifestation of Celiac Disease

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Case description: A 67 years old man presented to the clinic complaining of an itchy blistering skin rash over elbows, knees and buttocks for the past 2 months that is not responding to antihistamines. Itching has no diurnal variation. Patient has no chronic comorbidities and is not on regular medications. On examination, symmetrical vesicular lesions with erythema, crusting and itch marks are seen over both elbows, knees and back with -ve Nicolsky's sign. General examination reveals pallor, patient BMI 17.5, abdominal distention, and peripheral neuropathy. Investigations showed microcytic hypochromic anemia with HGB 9.8 mg/dl, MCV 68, HCT 29.8, WBCs and platelets are normal. K 3.1mg/dl, Na 135 meq/dl, Ca 8.5mg/dl, albumin 3g/dl. Skin biopsy and direct immunofluorescence of the skin around the lesions was taken that was diagnosed as dermatitis herpetiformis. Endoscopy was done and it confirmed the diagnosis of celiac disease.Treatment: Patient was advised to follow a gluten free diet plan and dapsone was started at a dose of 100 mg/day. Patient rash resolved gradually over a period of 3 weeks.

Keywords: Dermatitis Herpetiformis, Atypical Presentation, Celiac Disease, Gluten Enteropathy

Background

Celiac disease (CD) was originally considered a rare disorder that presents in childhood, but now it is recognized as a common disorder that may present at any age [1].
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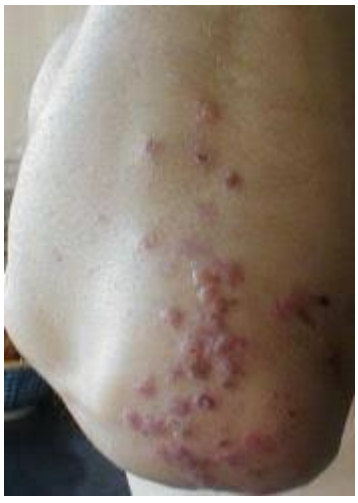
Celiac disease presentation is usually classic (diarrhea) in 62-85% of the cases but atypical presentations are seen in many cases like anemia or decreased bone density (15%), screening first-degree relatives (13%), and incidental diagnosis at endoscopy performed for such indications as reflux or dyspepsia (8%) [2-3].

Less common presentations include abdominal pain, constipation, weight loss, neurologic symptoms, dermatitis herpetiformis, hypoproteinemia, hypocalcemia, and elevated liver enzyme levels [2].

Prevalence of dermatitis herpetiformis (DH) ranges from 1.2 to 39.2 per 100,000 people, although no recent studies are available [4-9] .
Although CD is more predominant among females with a female to male ratio of about 2:1 to 4:1, yet DH is more common among males with a ratio of 1.5:1 to 2:1 [10-11].
Dermatitis herpetiformis usually presents in the third decade, although individuals of any age can be affected[12].
Direct immunofluorescence (DIF) of uninvolved skin remains the gold standard for establishing dermatitis herpetiformis diagnosis [13-14.]
Although most patients with DH have the histologic changes suggestive of gluten hypersensitivity, only 10-15% have clinical symptoms of CD [15]



Report

A 67 years old man presented to the clinic complaining of an itchy blistering skin rash on elbows, knees and buttocks for the past 2 months that is not responding to over the counter antihistamines. Itching has no diurnal variation. Patient has no chronic comorbidities and is not on regular medications. Patient had no constitutional symptoms no had he any history of recent drug intake before the onset of the rash. On examination, symmetrical grouped vesicular lesions with erythema, crusting, excoriations, hyperpigmentation and itch marks are seen over the extensor surfaces of elbows & knees and buttocks with -ve Nicolsky's sign. General examination reveals pallor, patient's BMI 17.5, abdominal distention, and peripheral polyneuropathy.

Investigations showed microcytic hypochromic anemia with HGB 9.8 mg/dl, MCV 68, HCT 29.8, WBCs and platelets are normal. K^+ 3.1 mg/dl, Na^+ 135meq/dl, Ca 8.5 mg/dl, albumin 3 g/dl. Skin punch biopsy from the lesions revealed subepidermal blister with neutrophilic predominance and direct immunofluorescence of the skin around the lesions was done showing positive granular IgA deposits at the tips of the dermal papillae that is consistent of DH. Endoscopy was done and biopsies from the duodenum were taken for histopathology. Duodenal biopsy showed intraepithelial lymphocytosis, crypt hyperplasia, and villous atrophy confirming the diagnosis of celiac disease.

Treatment: Patient was advised to follow a gluten free diet plan. But since response to diet control is slow, basic investigations to check if dapsone was suitable for the patient management was done (CBC, reticulocyte count, Glucose-6-phosphate dehydrogenase level and liver &renal function tests). Patient started dapsone at a dose of 100 mg/day. Patient rash resolved gradually over a period of 3 weeks and the dose was gradually tapered to 25mg/day after that. Follow up of patient CBC and reticulocytes every 2 weeks and liver and kidney functions were followed up after 3 months as advised by Mayo Clinic [16]. Fortunately, the patient experienced no side effects. Patient rash resolved gradually over a period of 3 weeks.

Discussion:

Dermatitis herpetiformis is common among Northern European ancestry but is rarely seen among Africans and Asians [16].

A variety of more common skin disorders that have a similar pattern to DH make it challenging for non-dermatologist to clinically differentiate. This includes scabies, atopic eczema, contact eczema, chronic prurigo, urticaria, popular urticaria as well as other autoimmune blistering diseases as bullous pemphigoid (BP), pemphigus vulgaris (PV), linear IgA bullous dermatosis (LABD) and epidermolysis bullosa acquisita (EBA) [17].

Screening patients with DH for autoimmune diseases especially antithyroid peroxidase (in almost 20% of patients), antigastric parietal cells (in 10–25% of patients), antinuclear and anti-Ro/SSA antibodies, should be tested. Also, screening for thyroid disorders and diabetes should be done. Screening for CD among symptomatic first degree relatives of patients with DH is indicated [12].

Conclusion:

Clinicians should be aware of different skin lesions that present as a manifestation of systemic diseases, such as in DH and CD.

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