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Dermatitis Herpetiformis

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Signs and Symptoms

The main sign that accompanies DH is the eruption of intensely pruritic papulovesical lesions that typically present bilaterally on the elbows, knees, buttocks, neck, and scalp (Criado et al., 2012). They may appear on the upper back, abdomen, groin, and face as well. The lesions are small blisters that resemble those that are caused by the herpes simplex virus. By the time a patient seeks evaluation by a care provider, the lesions have often been scratched so much as to cause erosions, excoriation, and or crusted papules (Junkins-Hopkins, 2010). The surrounding area may have erythema and or urticarial plaques. Patients may have areas of hyperpigmentation from previous outbreaks (Bonciani et al., 2012). The breakouts are of a chronic nature and will have periods of flare ups and remission. The periods of remission do not typically last long than several weeks (Criado et al., 2012). The hallmark symptom that patients with DH report is extremely intense itching. They may report experiencing the itching, a burning and or a stinging sensation up to 12 hours prior to eruptions on the skin.

AdditionalResources

Dermatitis herpetiformis lesions [Online image]. Retrieved July 28, 2015 from http://www.usmle-forums.com/usmle-step-1-forum/36692-treatment-dermatologic-problem.html

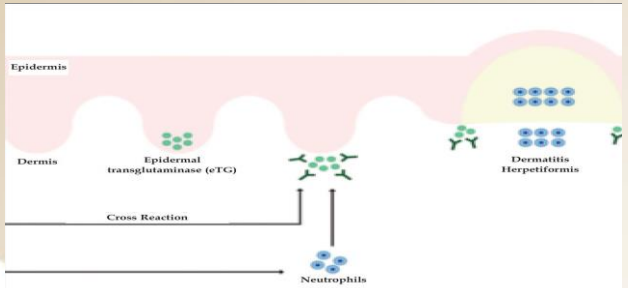
Gluten free [Online image]. Retrieved July 28, 2015 from http://www.medicalnewstoday.com/articles/288406.php

Skin [Online image]. Retrieved July 28, 2015 from http://www.scielo.br/scielo.php?pid=S0365-05962014000600865&script=sci_arttext

Pathophysiology

The pathophysiology of DH involves genetics, environment, and an immune response. Nearly all patients with DH are carriers of either the human leukocyte antigen (HLA) DQ2 or HLA DQ8 alleles (Bonciani et el., 2012). These are the same alleles the are present in patients with CD. The HLA DQ2 is the more commonly carried allele. The presences of one of these alleles is so strongly associated with DH and CD that the sensitivity of the test is nearly 100%. The absence of either allele nearly excludes a diagnosis of either disease. Studies performed using monozygotic twins and analysis of the incidence of the alleles among first-degree relatives shows the strong genetics factors are at play in the development of DH and CD (Bolotin & Petronic-Rosic, 2010).

The environment required to induce the reaction involved with DH is a diet containing gluten. Gluten is a protein composite found in wheat, rye, and barley (Bonciani et al., 2012). Gliadin, a component of gluten, is indigestible, and an alcohol-soluble protein that leads to the immune reaction involved in DH and CD.



Skin
(Dermatitis Herpetiformis)

The immune response in DH is seen in biopsies of active lesions, as well as perilesional skin (Bonciani et al., 2012). A biopsy positive for granular IgA in the dermal papillae and may also be present in the basement membrane (Junkins-Hopkins, 2010). It is believed to be IgA that helps neutrophils infiltrate the papillae and the basement membrane (Nakajima, 2012). Immunofluorescence studies are used for definitive diagnosis and to rule out similar presenting disorders of the skin such as bullous dermatosis (Bolotin & Petronic-Rosic, 2011).

Serum studies can be used to help in the diagnostic process. They evaluate for the presence of circulating IgA antibodies to endomysium (Bolotin & Petronic-Rosic, 2011). The IgA antibodies found in DH patients are specific for epidermal-specific transglutaminase (TG), known as anti-epidermal transglutaminase (eTG). Serum studies measuring anti-eTG are 95% sensitive for DH.

The immune response to gluten in the DH patient is evidenced by the inflammatory infiltrate composed of neutrophils on the dermal papillary tips seen on biopsy, as well as IgA deposits (Bonciani et al., 2012). Lesions are composed are also composed of eosinophils and fibrin which help to form microabscesses. The inflammation triggering T cells are also present on the lesions and perilesional skin, further evidence of immune response.

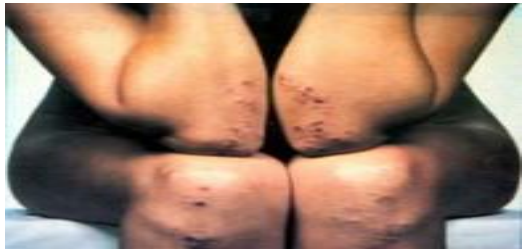
Dermatitis Herpetiformis: An Introduction

Dermatitis herpetiformis (DH) is a chronic autoimmune disease that manifests on the skin (Bolotin & Petronic-Rosic, 2011, p. 1017). DH presents with erythematous papules that often have vesicles (p. 1020). The lesions are intensely pruritic, so much so that they are often excoriated by the time a patient is evaluated by a physician. They have a distinct presentation erupting symmetrically on the extensor surfaces of the upper and lower extremities. Locations include the elbows, knees, scalp, and buttocks (p. 1020).

DH is considered an extra-intestinal manifestation of celiac disease (CD) (Krishnareddy, 2013). DH and CD are both autoimmune gluten sensitivities that are mediated by immunoglobulin A (IgA) (Bolotin & Petronic-Rosic, 2011, p. 1017). Key differences include the main autoantigens involved. DH involves epidermal transglutaminase (eTG) while CD involves tissue transglutaminase (tTG). A definitive diagnosis of DH is made with a biopsy taken from perilesional skin that is positive for granular deposition of IgA in the dermal papillae and the basement membrane under direct immunofluorescence (p. 1020). A diagnosis of CD begins with serologic testing (Autodore, Verma, & Gupta, 2012, p. 272). . If those tests are positive for CD related immunoglobulins, a endoscopic biopsy of the duodenum is in order. If CD is present, the biopsies will show villous atrophy. Both diseases are associated with human leukocyte antigen (HLA) (Bolotin & Petronic-Rosic, 2011, p. 1019). Genetic tests show HLA-DQ2 and HLA-DQ8 in effected carriers. Both diseases are treated with a life-long gluten free diet (GFD). DH treatment may include the use of the antibiotic dapsone for an extended period of time.

DH is of particular importance to myself because I have struggled with finding a diagnosis for the symptoms I have been experiencing over the past few years. DH was first considered a possibility for me after I presented to a dermatologist for the second time in a matter of months with incredibly pruritic lesions. No matter what I did they would not go away. I ended up seeing a gastroenterologist, but my neurologist has been the most helpful in my journey. She has celiac disease and completed the serologic testing needed for diagnosis. I tested positive for the genes involved with CD, but the immunologic testing was negative. She recommended I start a GFD to alleviate they various symptoms I was having that included others that just the skin presentation. After going back and forth with the necessity of a GFD when I did not have positive serologic studies, I realized that my thinking was much clearer when following the diet. I committed to following it. I did however continue to break out with the pruritic lesions. After multiple courses of prednisone I again returned to the dermatologist. He said that he believed DH is the correct diagnosis. After some more research I found that serologic studies are not always positive in those with DH, but it is still a manifestation of CD.

There is an abundance of literature on CD. It is my hope that through this project others practitioners will be provided with information on a disease they might not have otherwise heard of.



Nursing Implications

The pathophysiology of DH has significant implications for nurses that diagnose and care for these patients. Being able to provide an accurate diagnosis is the first step. Prescribing treatment and educating patients on the intricacies of a gluten free diet and the medication options available can be a involved process. The importance of a life-long gluten free diet must be communicated. Poor compliance has been associate with low levels of information on the GFD (Fasano, 2012). Even if remission is seen with the adjunct of medication, the diet is the only way to keep the flares from returning.

Currently the main pharmacologic treatment of CD is dapsone. Even following a strict GFD, IgA deposits within the skin are seen up to several years after its initiation (Bolotin & Petronic-Rosic, 2011). Dapsone has both anti-inflammatory and antibacterial properties that inhibit the recruitment of neutrophils. Dapsone therapy helps to resolve the active lesions within days of beginning treatment. There are drawbacks to its use, including methemoglobinemia and agranulocytosis. These are serious conditions that require frequent monitoring. If the patient is unable to tolerate dapsone therapy, sulfasalzine or sulfamethoxypridazine may be used.

DH is a disease that can present like many other skin diseases. To be able to accurately diagnose it can make an incredible difference in a patient's life.

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