Dermatitis herpetiformis (Duhring's disease)
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Saturday, 09 October 2010 06:16 - Last Updated Tuesday, 09 November 2010 05:43
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Dermatitis herpetiformis (Duhring’s disease) is a chronic, intensely itchy skin disease that typically affects the buttocks, thighs, knees, and forearms. The condition is characterized by a distinctive pattern of symmetric, urticarial lesions that can be triggered by dietary factors, particularly gluten. The disease is associated with an increased risk of developing gluten-sensitive enteropathy, also known as celiac disease.

**Clinical Manifestations**
- The usual symmetric distribution of lesions on elbows, knees, buttocks, shoulders, and sacral areas.
- Almost complete lack of symptoms in a rare patient. Most patients usually can predict the eruption of a lesion as much as 8 to 12 hours before its appearance because of localized hypopigmentation.
- Urticarial bullae are present on the skin.

**Pathophysiology**
- The disease is pathologically characterized by subepidermal blister formation, which is pathognomonic. The blister formation is due to the presence of immunoglobulin A (IgA) deposits, which are only occasionally seen by direct immunofluorescence.
- The pathogenesis of dermatitis herpetiformis involves a strong association between susceptibility genes and the disease. HLA-B8, -DR, and -DQ are associated with DH even more frequently than is seen in celiac disease and with a specificity for Tgases.
- Anti-IgA antibodies directed against dietary proteins produced in gut secretions in patients with DH.
- The third component of complement (C3) is frequently found in the same location as IgA deposits, suggesting that the IgA deposit is the actual target for complement activation.

**Diagnosis**
- IgA deposits in normal-appearing skin is the most reliable criterion for the diagnosis of DH.
- The finding that T-cell lines from patients with DH produce significantly more interleukin 4 (IL-4) than control T-cells is consistent with a Th2-type response.

**Treatment**
- A gluten-free diet is the mainstay of treatment for dermatitis herpetiformis. It is effective in alleviating the skin disease within a few weeks. However, it is only the very highly motivated patient who can adhere to the diet, which requires total avoidance of gluten in all forms.
- Anti-inflammatory drugs often exacerbate DH, even in patients taking dapsone.
- Studies using elemental diets in the treatment of DH have shown that a significant proportion of patients experience initial improvement. Patients should be instructed to take the minimal dose required to achieve the desired response.

**Complications**
- There is a marked increase in the incidence of certain major malignancies, especially gastrointestinal lymphomas. Studies have shown that up to 1 percent of patients with DH develop a lymphoma from 2 to 31 years after diagnosis.
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**Differential Diagnosis**
- Patients with DH have an increased incidence of gastrointestinal lymphomas.
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**AT A GLANCE**
- Dermatitis herpetiformis (Duhring's disease) = an intensely itchy, chronic skin disease that typically affects the buttocks, thighs, knees, and forearms.
- The disease is associated with an increased risk of developing gluten-sensitive enteropathy, also known as celiac disease.
- The usual symmetric distribution of lesions on elbows, knees, buttocks, shoulders, and sacral areas.
- Almost complete lack of symptoms in a rare patient. Most patients usually can predict the eruption of a lesion as much as 8 to 12 hours before its appearance because of localized hypopigmentation.
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