CASE 25

Patient: A 75-year-old Thai man from Lamphun
Chief Complaint: 6-month-history of itchy vesicles at both thighs and elbows, upper back and sacral area
Present Illness: The patient presented with 6-month-history of several pruritic vesicular eruption symmetrically distributed on both elbows, anterior aspect of thighs, upper back and sacral areas. Soothing agents such as topical camphor and phenol did not seem to be beneficial. He also had been occasionally suffering from dyspepsia, nausea, vomit and loss weight 4 kilograms in 6 months.
Past History: He had had hypertension, atrial fibrillation and osteoarthritis of knees for twenty years.
Current Medication: Amlodipine 5 mg/day, Atenolol 50 mg/day, Arcroxia 90 mg/day
Family History: Unremarkable
Dermatological Examination (Figure 25.1-5)**:** Symmetrical several clear vesicles with excoriation on both elbows, anterior aspect of both thighs, upper back and sacral area

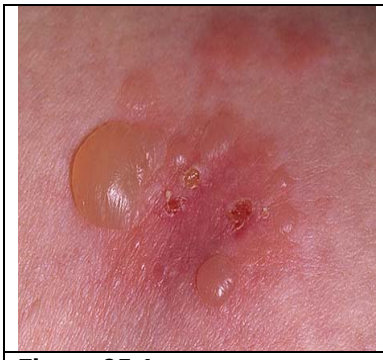

Figure 25.1

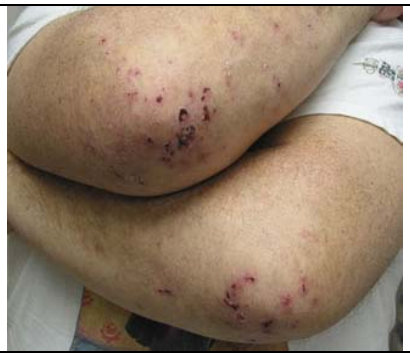

Figure 25.2

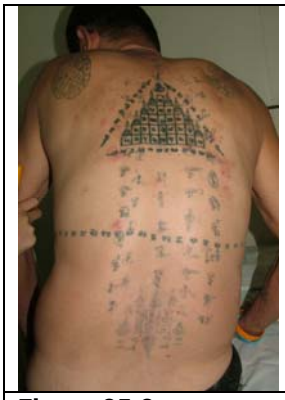

Figure 25.3

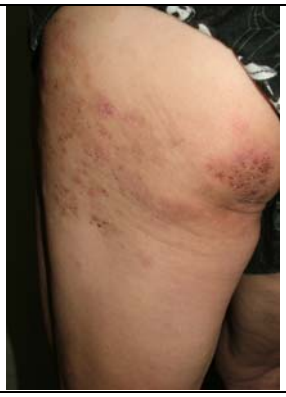

Figure 25.4

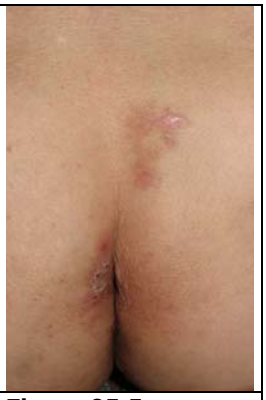

Figure 25.5

Histolopathology (S10-1599) (Figure 25.6): Subepidermal vesicle in association with superficial inflammatory cell infiltrate, predominately neutrophils

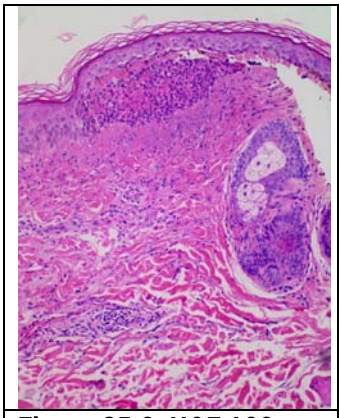

Figure 25.6, H&E 100x

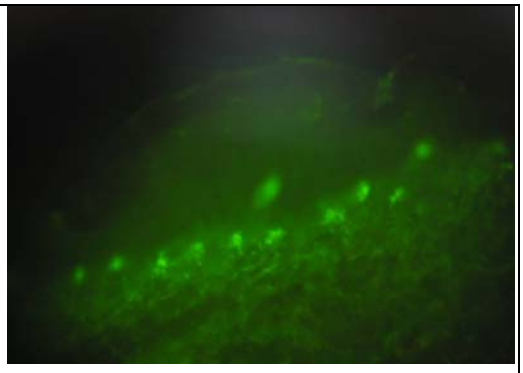

Figure 25.7, DIF

Direct Immunofluorescence (Figure 25.6)**:** granular IgA deposit within the dermal papillae

Investigations:

Ultrasonography of whole abdomen: Diffuse increase liver parenchymatous without focal mass. No IHD nor CBD dilatation. Pancrease and spleen appears normal. No ascites or retroperitoneal lymphadenopathy.

EGD: No typical endoscopic finding in Celiac disease. Randomized biopsy was performed at duodenum and stomach.

Chronic mild to moderate inflammation of duodenum, chronic moderategastritis, moderate tissue eosinophilia, no H. Pyroli was seen.

Direct immunofluorescence revealed 50 percent of plasma cells in lamina propria. Normal villous without intraepithelial lymphatic infiltration. All findings cannot be concluded with celiac disease.

Diagnosis: Dermatitis Herpetiformis

Presenter: Wanjarus Roongpisuthipong

Consultant: Parichart Chalidapongse

Treatment: Dapsone 100mg/day and gluten-free diet

Discussion:

Dermatitis herpetiformis (DH) or Duhring-Brocq disease is an autoimmune subepidermal blistering disease characterized by chronic and recurrent eruption of erythematous, urticarial, papular, vesicular and bullous lesions. Granular IgA deposits at the dermal papillae represent the immunological marker of the disease, which is strictly associated with a gluten-sensitive enteropathy (GSE) or celiac disease.[1]

In a study from Scotland the incidence was found to be 11.5 per 100,000 [2] and ranging from 19.6 to 39.2 per 100,000 in Sweden.[3] The disease occurs mainly the age between 20 and 55, but is occasionally seen in children.[4]

The onset of DH may be acute or gradual, and pruritus usually the first and predominant symptom. Lesions are pleomorphic, consisting of itchy, erythematous papules and urticarial wheals that symmetrically on the extensor surfaces, buttock, back, rarely on the face. Small grouped vesicles progressively developed on plaques of erythema but they rapidly are excoriated, evolving into crusts and erosions.[5] Oral lesions, detectable in about 50% of patients, mainly consist of mucosal redness, ulcerations, atrophy in tongue or oral blisters. Pain or burning sensation may accompany these lesions.[6]

DH shares a strong association with class II histocompatibility locus antigens DR3 and DQW2 and with the alleles DQA1*0501 and B1*0201 of chromosome 6.[7] Granular deposits of IgA at the dermo-epidermal junction (DEJ) in the skin of affected patients. Furthermore circulating IgA subclass1 autoantibodies to endomysium can be detected in both DH and celiac disease; these antibodies are closely linked to the degree of intestinal abnormalities and are direct indicators of intestinal abnormalities and are direct indicators of the patients' adherence to a gluten-free regimen.[8] Sárdy et al. demonstrated that patients with DH have IgA transglutaminase antibodies with higher affinity to epidermal transglutaminase (type III) than tissue transglutaminase (type II).[9] Diagnosis of DH usually requires clinical manifestation and histology that compatible with the disease and also evidence of granular IgA deposits in the dermal papillae by the direct immunofluorescence in the perilesional skin of patients with DH, which is the hallmark of the disease.[10] In pediatric patient, screening of antibodies (IgA and IgG) against tissue transglutaminase (tTG) and deamidated gliadin peptides (DGP) by enzyme immunoassay (EIA) was slightly more sensitive than IgA anti-tTG alone.[11]

Patients with DH have a high incidence of autoimmune disorders, thyroid disease, pernicious anemia, lupus erythromatosus, vitiligo and insulin-dependent diabetes.[12] As with celiac disease, there is an increased incidence of lymphoma and a gluten-free diet appears to protect patients.[13]

The clinical differential diagnosis includes Eczema, Atopic dermatitis, neurotic excoriations, papular urticaria, transient acantholytic dermatosis, pemphigoid, erythema multiforme and scabies. The histopathological differential diagnosis includes bullous pemphigoid, scarring pemphigoid, Henoch-Schonlein purpura, and alcoholic liver disease.

Diaminodiphenyl sulfone (dapsone), sulphapyridine or sulfapyridine have been used to suppress the skin manifestation.[14] There is a strong evidence to recommend a gluten-free diets as a mainstay of treatment in DH for the following reasons: (1) the need for medication is reduced or abolished; (2) there is resolution of enteropathy; (3) a general feeling of well-being; and (4) protective effect against lymphoma development.[15,16] Patients should avoid nonsteroidal anti-inflammatory drugs to decrease incidence of exacerbation.[17]

References

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