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Case Report

Herpetiform dermatitis: A case report

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Abstract Herpetiform dermatitis (HD) also known as 'morbus duhring' is a chronic autoimmune skin disease that causes severe itching. This condition is associated with antitransglutaminase IgA autoantibody on the superficial papillary dermis layer that causes neutrophil infiltration and blister formation. Formerly, it was explained by American dermatologist, Louis Adolphus Duhring in 1884. This disease is characterized by papules, vesicles, plaques, urticaria, erythema, and excoriations that vary from 1–3 mm, mainly located on the extensor areas of the upper and lower extremities, shoulders, mid-back, and buttocks that are symmetrically distributed. HD is known to be associated with gluten-sensitive enteropathy (GSE) or celiac disease (CD) which is believed to play an important role in its pathogenesis. More than 90% of HD patients are shown to be gluten-sensitive. This case report investigates HD's clinical characteristics and association with GSE or CD in a defined patient population.

Key words
Herpetiform dermatitis; Morbus duhring; Autoimmune skin disease; IgA autoantibody.

Introduction

Herpetiform dermatitis (HD) also known as 'morbus duhring' is a chronic autoimmune skin disease that causes severe itchiness.[1] HD show increased expression of HLA-DQ2, HLA-DQ8, HLA-A1, HLA-B8, and HLA-DR3.[2,3] HD is managed by medical and non-medical therapy.[3]

Case History

A 54-year-old man came to Dermatology and Venereology Polyclinic, University of Sumatera Utara Hospital with the main complaint of red and severely itchy rashes accompanied by blisters and scabs from scratching on both forearms, elbows, both hands, upper back, back of the waist, both thighs, calves and soles, for 7 years. The disease started as red nodules appeared on both forearms and around the elbows, the size of pins to corn kernels, which were very itchy, within 1-2 days these red nodules turned into water-filled blisters. In the last two years, the rash spread to other parts of the body, such as on both thighs, then extended to the calves and back. The patient likes to consume noodles and bread every day.

During a dermatological examination, multiple erythematous and hyperpigmented papules, ranging in size from 1 to 3 mm, with some areas of confluence, along with crusting, erosion, and excoriation were observed in a generalized distribution. These lesions were present on the anterior and posterior aspects of both forearms, the dorsal surfaces of hands, the lumbar region, the lateral aspects of both thighs, and both shins. Additionally, hyperpigmented plaques with crusting were noted on the anterior surfaces of both hands **(Figure 1)**.

Complete blood examination reveals hemoglobin of 16 g/dL, hematocrit of 47.6%,

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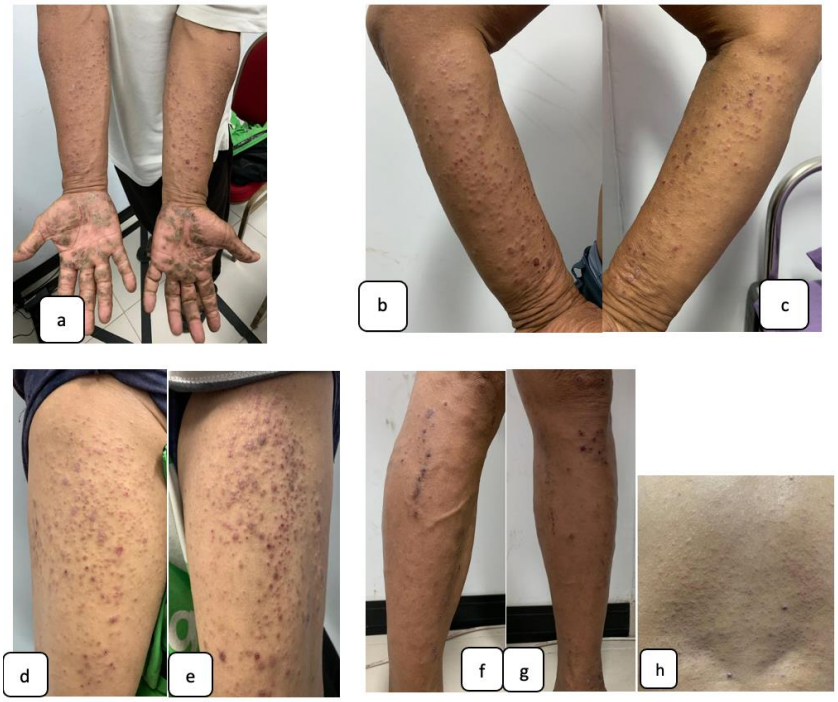

Figure 1 During a dermatological examination, multiple erythematous and hyperpigmented papules, ranging in size from 1 to 3 mm, with some areas of confluence, along with crusting, erosion, and excoriation, were observed in a generalized distribution. These lesions were present on the anterior and posterior aspects of both forearms, the dorsal surfaces of both hands, the lumbar region, the lateral aspects of both thighs, and both shins. Additionally, hyperpigmented plaques with crusting were noted on the anterior surfaces of both hands (a-h).

leukocytes of 7.170/µL, erythrocyte of 516.000/µL, platelets of 287.000/µL. The histopathological examination impression supports a herpetiform dermatitis **(Figure 2)**.

Based on the history and dermatological examination, the differential diagnoses in the patient were dermatitis herpetiformis, linear IgA dermatosis and prurigo nodularis. The patient's provisional diagnosis was dermatitis herpetiformis. The patient was given treatment with desoximetasone 0.25% topical cream 2x a day on papular lesions, fusidic acid applied 2x a day on scratch marks, cetirizine 1x10 mg and dapsone tablets 50 mg/day. Patient was encouraged to start a gluten-free diet.

After 6 weeks of treatment, there was improvement in the patient's skin lesions. Dermatological examination reveals multiple erythematous-hyperpigmented papules in the scapular and lumbar regions, symmetrically distributed bilaterally. Hypopigmented macules are present on the anterior and posterior aspects of both forearms, while hyperpigmented macules are noted on the lateral aspects of both thighs and shins. Additionally, xerosis is observed on the palmar surfaces of both hands.

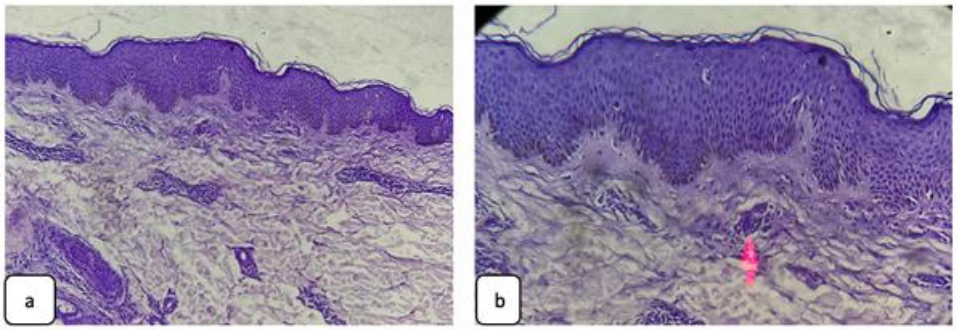

Figure 2 Histopathological examination showed infiltration of mononuclear and polymorphonuclear inflammatory cells forming microabscesses in the papillary dermis. In the subepithelial layer a multifollicular stroma is seen, consisting of fibrocollagenous connective tissue infiltrated with inflammatory lymphocytes and histiocytes;40× magnification; H&E (a) red arrows indicate microabscesses;100 magnification; H&E (b).

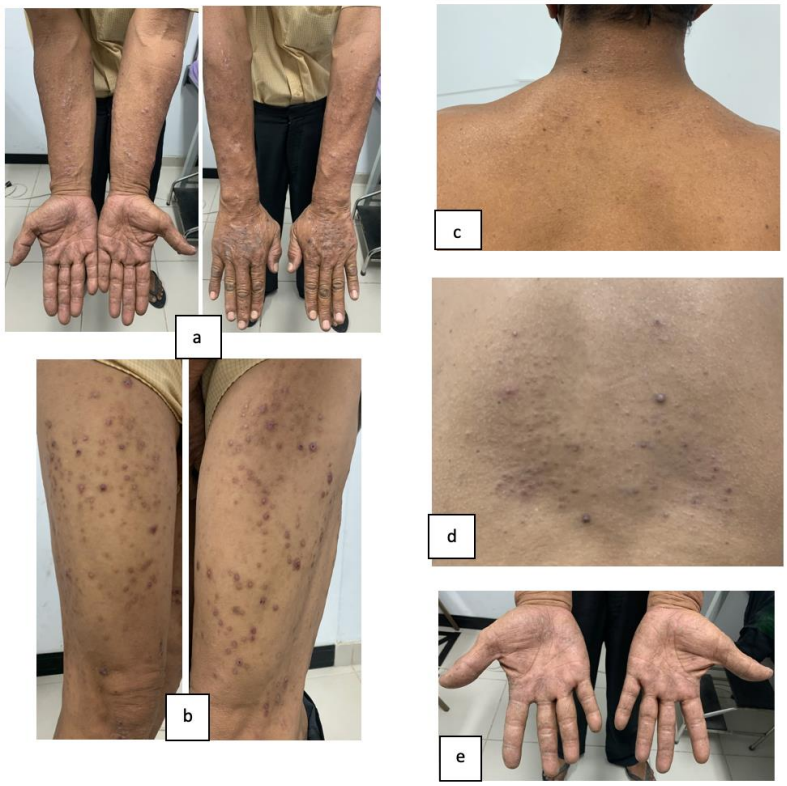

Figure 3 Dermatological examination reveals multiple erythematous-hyperpigmented papules in the scapular and lumbar regions, symmetrically distributed bilaterally. Hypopigmented macules are present on the anterior and posterior aspects of both forearms, while hyperpigmented macules are noted on the lateral aspects of both thighs and shins. Additionally, xerosis is observed on the palmar surfaces of both hands. (a-e).

(Figure 3). Patient management is continued; cetirizine tablet 1x10 mg, cimetidine tablet 1x200mg, dapsone tablet 100 mg/day, sangobion® once a day, topical desoximetasone cream 2x a day on the arms, thighs, calves, and back, desoximetasone 0.25% cream + Soft u derm® topical 2x a day on both palms. We consulted the gastroenterology department and the nutrition department for patient management.

Discussion

In this case, a 54-year-old male patient with the chief complaint of severe itchy-red rash accompanied by blisters and scabs from scratching on both forearms, elbows, both hands, back of the waist, both thighs, calves and feet that has been experienced for the last 7 years. Epidemiologically, HD is a rare disease. Some reports show the ratio of the incidence of HD in men to women is 1.5:1.[4,5] Reasons that explain the low prevalence of HD among this population include the absence of a predisposition to the human leukocyte HLA haplotypes DQ2 and DQ8, which are invariably found in Caucasian HD patients and the low consumption of wheat in this geographic region.[6,7]

The typical lesions of HD are erythematous papules and vesicles which are arranged in groups and are located on the extensor areas. Polymorphism and symmetrical distribution of lesions are the main clinical features of HD. The disease usually presents with grouped erythematous papules and urticarial plaques with profuse vesicles. The vesicles may coalesce into small, tense blisters with a sero-hemorrhagic content, which are characterized by a centrifugal growth pattern. Erosion, excoriations, and crusting may be formed from the rupture of the blisters and from scratching secondary to the associated pruritus. HD lesions are symmetrically localized on the extensor surfaces of the upper and lower limbs, mostly on the elbows and knees, buttocks, and the sacral

region; the abdomen, upper back, shoulders, nuchal area, and scalp may also be affected.[7] Because the condition is intensely itchy, intact vesicles are rarely seen. Lesions tend to be symmetrical and heal without scarring. Purpura punctata can also be seen on the palms and soles.[3] Grouping of herpetiform (herpes-like) lesions is common in several areas, but patients may also have multiple non-grouped lesions. Symptoms vary widely from a burning sensation which is usually severe and itching in most patients.[3]

The patient admits that he eats noodles and bread almost every day. The pathogenesis of HD, which depends on a complex inflammatory network in the skin gut, is currently only partially understood.[7] Gluten has an important role in HD. Gluten is a type of protein found in wheat, oats, and rye.[3] There is growing evidence that HD should be considered as a phenotypic-specific skin expression of gluten-sensitive enteropathy that is indistinguishable from celiac disease.[3,4]

In routine histopathological examination papillary dermis filled with neutrophils, with separation at the lower end of the rete ridges is a feature that can be found in HD.[6,8] DIF with deposits of granular IgA at the tips of the dermal papillae is the gold standard test for the diagnosis of HD.[5]

Dapsone is a therapeutic option for HD; doses of 50-150mg/day can control itching and the formation of new lesions.[5] Dapsone combination with a gluten-free diet will result in more rapid clinical improvement.[9]

The prognosis for this patient is uncertain but generally favorable concerning life expectancy, functional outcome, and potential for recovery. HD is a chronic disease that requires the patient to adopt a long-term gluten-free diet.[7-9]

Conclusion

HD is considered a skin manifestation of CD. Clinical manifestations, biopsy, and DIF examination can establish the diagnosis of HD. Dapsone consumption with a gluten-free diet gives better results.

Declaration of patient consent The authors certify that they have obtained all appropriate patient consent.

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Conflict of interest Authors declared no conflict of interest.

Author's contribution

CPH: Diagnosis & Management of the case, critical review, final approval of the version to be published.

MM: Identification of the case & Management of the case, manuscript writing, final approval of the version to be published.

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