An unusual case of pemphigus erythematosus

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CASE REPORT

An unusual case of pemphigus erythematosus

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ABSTRACT

Pemphigus erythematosus is characterized by fragile vesicles or bullae, erosions, crusts, and scales in seborrheic area. There are several forms of atypical lesions such as erythematous papules and plaques, verrucous plaques, pustules, and lichenificati Here, we report an atypical pemphigus erythematosus with erythematous papules, plaqu and pustules skin lesions. A 52-year-old Indonesian man presented with prominent pruritic erythematous macules, papules, plaques on the scalp, trunk, and extremities, and also a pustule for each on the back and right arm. Clinically, the patient was diagnosed as small-plaque parapsoriasis, but stopathology examination on the pustule revealed a subcorneal acantholysis and direct immunofluorescence staining showed immunoglobulin G and complement C3 on the cell surface of keratinocytes. These result suitable for pemphigus erythematosus. The patient was treated with topical and systemic corticosteroid, and there were significant improvements in the skin lesions. Pemphigus erythematosus may present with prominent erythematous papules, plaques, and a few pustules. A careful assessment of the clinical manifestation and histopathological findings enabled us to make a correct diagnosis and succesfully treat the patient.

Key Words: Pemphigus erythematosus, Plaques, Pustules

1. INTRODUCTION

Pemphigus is an autoimmune chronic bullous disease involving the skin and mucous membranes.[1] The histological characterization of pemphigus is an intraepidermal cleft and acantholysis, immunopathologically, immunoglobulin (Ig) G autoantibodies directed against the cell surface of keratinocytes.[2] There are four clinical variants of pemphigus: pemphigus vulgaris, pemphigus foliaceus, pemphigus paraneoplastic, and pemphigus IgA.[1] Pemphigus erythematosus is a variant of superficial and localized pemphigus foliaceus,[1,2] which has features of both lupus erythematosus and pemphigus.[3,4] Its common manifestation is vesicobullous. There were a few cases of pemphigus erythematosus with clinical manifestations as pustules[5,6] and hyperkeratotic lesions.[7] We present a case of pemphigus erythematosus with prominent erythematous papules, plaques, also a few pustules, and we discuss the associated clinical, histological, and immunological findings.

2. CASE PRESENTATION

A 52-year-old Indonesian man came to our dermatology clinic for a year history of pruritic erythematous macules and multiple papules on both arms. Erythematous papules widened as plaques and spreaded to the chest, back, abdomen, scalp, and lower limbs (see Figure 1A-B). Before his presentation to our clinic, he was treated by two other dermatologists and had several systemic and topical treatments, but the skin lesions appeared after the treatment were stopped.

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Two weeks earlier, patient complained about extensive pruritus with erythematous macules, multiple papules and plaques (see Figure 1C), which spread on the scalp, face (in malar area), mostly in upper trunk, both extremities, and a pustule (see Figure 1C) was found for each on the back and right arm. Patient denial the history of persistent erythematous macules after sun exposured on the face, blisters with clear contents, erosions, and oral ulcerations. There was no history of bullous disorders in patient and his family. Gram staining showed inflammatory cells and was not found bacteria.

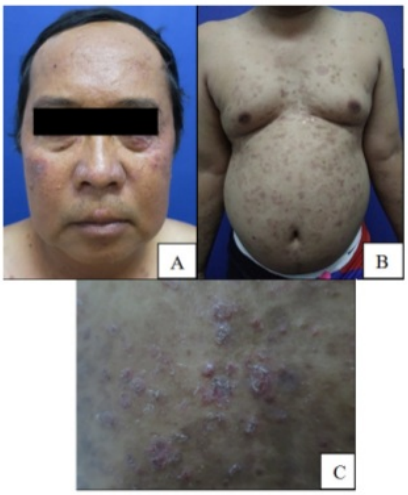

Figure 1. Clinical findings in this patient were erythematous plaques and hyperpigmented macules in the malar area (A), erythematous macules, papules, plaques, and hyperpigmented macules on the trunk (B), erythematous plaques, hyperpigmented macules, scales, and a pustule on the back (C).

According to physical examination and Gram staining, the diagnosed of the patient was small-plaque parapsoriasis. The patient was biopsied and the histopathology examination from the plaque on the back and the pustule on the right arm using hematoxylin and eosin, revealed subcorneal vesicobullous reaction, acantholytic cells in the cleft, parakeratotic, and crust in epidermis (see Figure 2A-B). To ensure the histopathology result, direct immunofluorescence staining was taken from sun unexposed area which the pustule on the back. The tissue sections were stained with IgG, IgA, IgM, C3, fibrinogen, and polyvalent, and showed IgG and complement C3 on the surface of the keratinocytes (see Figure 3A-B). Those results were suitable with pemphigus erythematosus. The patient was treated with 0.25% desoxymethasone cream and received 16 mg methylprednisolone orally per day for two weeks. Significant improvements in the skin lesions were observed within nine days after the start of the treatment. Methylprednisolone were tapping off every two weeks following the improvement of skin lesions. Written informed consent was obtained from the patient for publication of this case report and accompanying images.

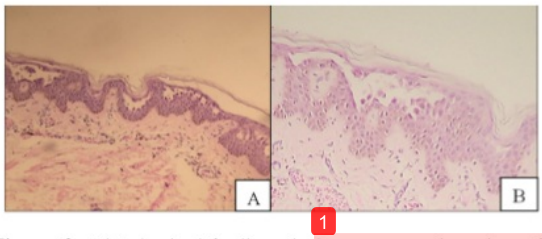

Figure 2. Histological findings in lower power imaging of the pustular lesion reveal subcorneal vesicobullous reaction and acantholytic cells in the cleft, parakeratotic, and crust in epidermis from the pustule on the right arm, hematoxylin and eosin stain, 40x (A). Subcorneal vesicobullous reaction and acantholytic cells in the cleft, hematoxylin and eosin stain, 100x (B).

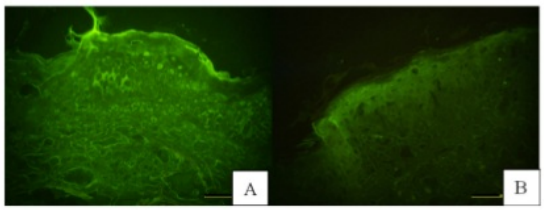

Figure 3. Direct immunofluorescence staining from the pustule on the back showed IgG deposition on the cell surfaces (A). Complement C3 on the cell surfaces (B)

3. Discussion

Pemphigus erythematosus in unusual presentation is infrequent.[5] There were a few case reports of pemphigus erythematosus with atypical lesions. Most of the pemphigus erythematosus with unusual cases showed extensive pustules,[6] vesicopustular,[5] and skin lesions mimicking seborrheic keratosis.[7] In our patient, we observed prominent papules, plaques, and a few pustules accompanied by pruritus, that were different from the vesiculobullous lesion in other pemphigus erythematosus patients. Common distribution of pemphigus erythematosus spread to the scalp, malar area,[8] and trunk,[5] but mainly in seborrheic area.[2] This distribution of skin lesions were found in our patient. We did Gram staining on the pustules to exclude of the secondary bacterial infection and the result showed inflammatory cells, without found any bacteria. Pemphigus erythematosus is clinically and serologically overlaps with pemphigus foliaceus and lupus erythematosus, but a positive antinuclear antibody (ANA) is only detected in 30%-80% pemphigus

erythematosus patients.[9] Histopathology of pemphigus erythematosus was found acantholysis in superficial layers of the epidermis, and cleft in subcorneal.[1] From direct immunofluorescence staining demonstrated intercellular IgG deposition and complement C3 in cells surface as in pemphigus foliaceus or pemphigus erythematosus.[2] IgG and C3 are mostly found in the sun exposed area due to ultraviolet B (UVB) exposure of the skin promotes histologic acantholysis in pemphigus patients.[10] Based on clinical manifestation, histopathology, and direct immunofluorescence, these results strongly diagnosed as pemphigus erythematosus. Except direct immunofluorescence, enzyme-linked immunosorbent assay (ELISA) could be performed to diagnose pemphigus for its high sensitivity of anti desmoglein 1 (Dsg1).[11, 12] Anti-Dsg1 antibodies deposite along dermo-epidermal junction mimicking in ANA negative patients.[8, 13] As none of the direct immunofluorescence-positive patients was anti-Dsg-negative,[12] in this case we did not perform ELISA because we already found IgG and C3 from direct immunofluorescence staining.

Later, the patient was effectively controlled by systemic and topical steroid therapy. To our knowledge, this is the first case of prominent plaques and pustular lesions in pemphigus erythematosus, as seen in our patient, have been reported. Our case demonstrates that infrequent case of atypical pemphigus erythematosus may cause difficulties in diagnosis and needed histopathological examination.

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CONFLICTS OF INTEREST DISCLOSURE

All authors declared no competing interests.

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