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Case Report **OPEN ACCESS**

Pemphigus foliaceus: A rare blistering skin lesion

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ABSTRACT

Pemphigus foliaceus is a benign variety of pemphigus group. It is a rare autoimmune blistering disease that affects the skin without mucosal involvement. It can present in endemic and sporadic form; whereby endemic form is common in Brazil and Tunisia, known as fogo selvagem. There was no previous case reported in Malaysia. We report a case of a 43-year-old man, who presented with generalized extensive blisters and vesicles that form crust and scale. The full evaluation established the diagnosis of pemphigus foliaceous. Delay in diagnosis can lead to poor outcomes. Primary care practitioners should be aware of this since the condition can mimic eczema, specifically photodermatitis.

Keywords: pemphigus, acantholysis, blister, skin diseases, autoimmune diseases

INTRODUCTION

Pemphigus foliaceous is a rare autoimmune disease presenting in endemic and sporadic forms [1]. It belongs to pemphigus, which comprises a group of potential life-threatening blistering diseases of the skin and mucous membranes [2]. The term “pemphigus” was first described by Boissier de Sauvages in his classification of skin diseases and further described by Wichmann as a chronic bullous disease [3]. The term then was further described as pemphigus foliaceus by Cazenave [3].

Pemphigus foliaceous is clinically characterized by blisters and erosions, but scaly or crusted erythematous patches are common [4]. It is less common than pemphigus vulgaris worldwide [5]. It equally affects men and women, whereby the mean age at onset is usually 50 to 60 years old [5]. However, it has also been reported to occur in children [6]. Endemically it is being reported primarily in Brazil and Tunisia [5]. To the best of our knowledge, it is being reported in the neighboring country, which is in Singapore in a retrospective study [7]. However, in Malaysia, there was no case report found on this. There was reported fatality, and a delay in diagnosis and treatment can lead to fatal outcomes [8].

CASE REPORT

A 43- year-old Malaysian man, who works as a chef was initially referred to a dermatology clinic from a local government clinic for photo contact dermatitis. He presented with a generalized skin lesion for a one-year duration. Initially, the lesion started at the right cheek and was described as plaque that worsened with sun exposure. The lesion then spread to the left cheek, scalp and the whole body. Lesion initially started with a small blister and vesicles, then ruptured, forming crust. The lesions were itchy and also painful. He went multiple times to private practitioners and has been treated with multiple oral antibiotics, antihistamines and topical steroids. The lesion temporarily resolved, then recurred. He denied taking other over-the-counter drugs.

Physical examination revealed that the patient was afebrile and hemodynamically stable. Glucometer reading was normal. Scalp examination reveals multiple hyperpigmented crusted patches and plaques. On face examination, there was a greasy yellowish crust with sparing over the periorbital area. No oral lesions were seen. Trunk examination shows multiple crusted erosions, scabs and post-inflammatory hyperpigmentation. Arms and legs examination reveals multiple dry erosion with few intact vesicles (**Figure 1**). The body surface area involved was around 60%. The Nikolsky sign was positive.

A full blood count result revealed a hemoglobin level of 14.8 g/dL, white blood cell of 14.35×10^9/L and platelet count of 244×10^9/L. The renal function shows mild hyperkalemia with potassium 5.5 mmol/L. Other renal functions were normal. The liver function test was normal. There was also mild hypocalcemia with total calcium of 2.06 mmol/L. Corrected calcium was 2.10 mmol/L. Magnesium and phosphate were normal. Anti-nuclear antibody-IgG (ANA) was negative. There was no growth of bacteria on blood culture and sensitivity.

A skin biopsy taken from the intact vesicles over the left forearm shows intraepidermal bullae (sub-corneal). The bullae contain fibrin, acantholytic cells, eosinophils and neutrophil. There is mild dermal perivascular infiltration by neutrophils, eosinophils and lymphocytes (**Figure 2**). Direct Immunofluorescence study (DIF) showed epidermal intercellular deposition of IgG.

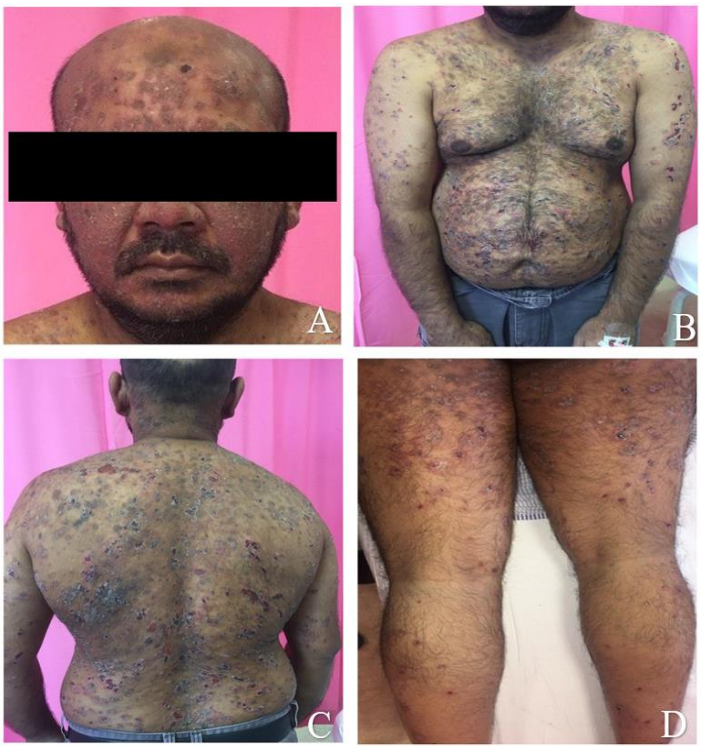

Figure 1. Greasy yellowish crust over the facial area (A): The anterior part of the body shows multiple crusted erosions, scabs, & post-inflammatory hyperpigmentation. Arms reveals multiple dry erosion with few intact vesicles (B): The posterior part of the body shows multiple crusted erosions, scabs, & post-inflammatory hyperpigmentation. Arms reveals multiple dry erosion with few intact vesicles (C). Lower limbs reveal multiple dry erosion with few intact vesicles (D) (reprinted with permission of the patient)

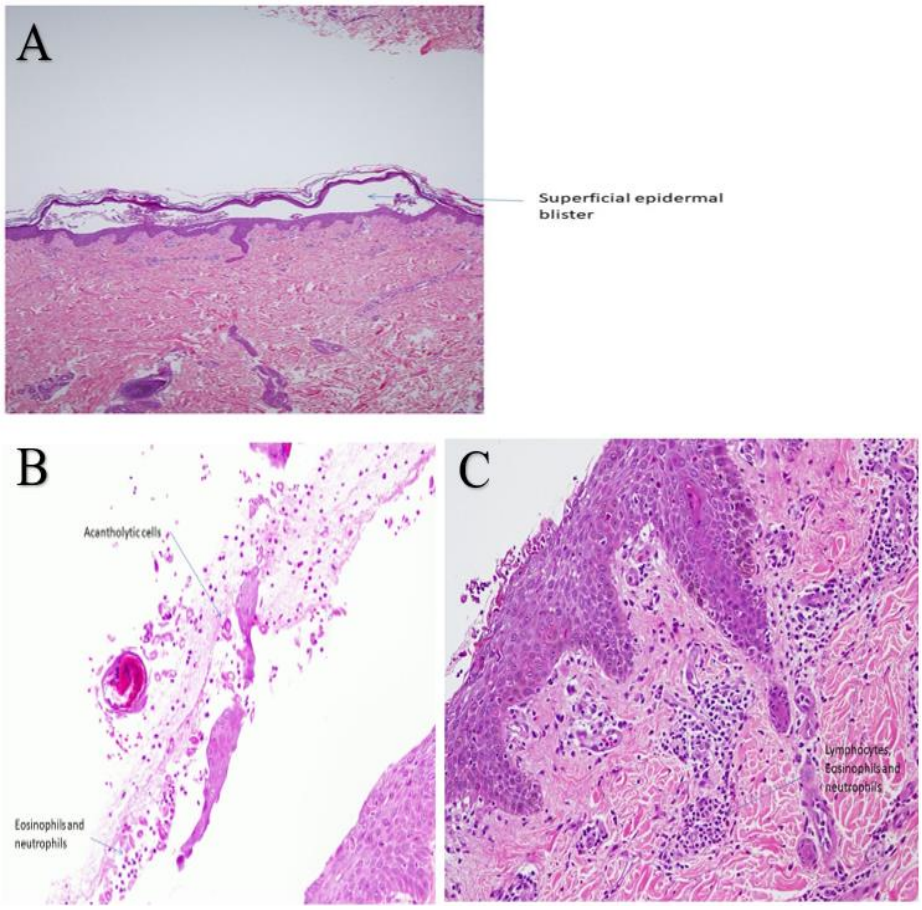

Figure 2. Intraepidermal bullae (sub-corneal) (A). The bullae contain fibrin, acantholytic cells, eosinophils, & neutrophil (B). Dermal perivascular infiltration by neutrophils, eosinophils, & lymphocytes (C) (reprinted with permission of the patient)

A diagnosis of pemphigus foliaceus was made based on clinical features, history and physical examinations supported by tissue biopsy. He was admitted to the ward and was started on IV cefuroxime 750 mg every eight hours to cover secondary bacterial infections. Intravenous hydrocortisone 100 mg every eight hours was started. Topical betamethasone cream 1:2 twice daily and emollient which was aqueous cream three times daily was prescribed. Antihistamines which include tab loratadine 10 mg in the morning and tab chlorpheniramine 4 mg at night, were also given to reduce itchiness. Eventually, after day 5 of admission, he showed improvement, and no new lesions were observed. He was discharged with tablet prednisolone (1mg/kg/day)–a total of 75 mg daily, tablet cefuroxime 500 mg twice daily for another four days (to complete antibiotics for one week duration), tablet azathioprine 50 mg daily (steroid sparing agent started on the day of discharge), tablet calcium carbonate 500 mg every eight hours, tablet calcitriol 0.25 mcg once daily, tablet loratadine 10 mg once at morning and tablet levocetirizine 5 mg once at night. The patient provided written informed consent for the publication of clinical details and images.

DISCUSSION

Pemphigus foliaceus should be suspected in a patient presented with a recurrent blistering skin lesion. The lesion usually begins at the trunk but can also occur at the face or scalp, which can later become widespread. Furthermore, the blisters can easily be ruptured due to brittle and subsequent transient nature [1, 5]. Compared to pemphigus vulgaris, typically, there are no oral or other mucosal lesions [1]. In pemphigus erythematosus, the lesion is usually in the sun-exposed areas which combines the features of lupus erythematosus that is associated with a positive ANA in the serum [1]. In this patient, the clinical features suggest pemphigus foliaceous; moreover, the history of worsened skin lesion in sun-exposed areas can suggest pemphigus erythematosus or systemic lupus erythematous. However, the antinuclear antibody is negative. The Nikolsky sign had a moderately sensitive but highly specific tool for diagnosing pemphigus [1]. It is elicited by applying lateral pressure using a finger or a thumb at affected skin, which causes the upper layers of the epidermis to dislodges from the lower layers. This sign was present in this patient.

There were a lot of differentials for blistering skin lesions apart from other types of pemphigus. Other differentials include bullous impetigo, cutaneous lupus erythematosus, drug eruption and sub-corneal pustular dermatosis, seborrheic dermatitis. Thus, the diagnosis requires clinical and histopathology/DIF that can suggest pemphigus foliaceous [1, 9]. Superficial sub-corneal acantholysis in the upper stratum spinosum or granulosum and a slight inflammatory infiltration of the upper dermis are the features of pemphigus foliaceous [9, 10]. The hallmark of pemphigus is the presence of IgG autoantibodies towards the cell surface of keratinocytes with DIF whereby in pemphigus foliaceous Immuno-fluorescence study shows intercellular deposition of IgG [10]. Diagnosis of pemphigus foliaceous was confirmed in our patient from clinical presentations and supported by skin biopsy results.

The goal of treatment is to promote healing as well as limit the new development of blisters and erosions [9]. In addition, the treatment will lead to improve functional status, quality of life and limit the side effects associated with long-term usage of corticosteroid or other immunosuppressive agents [9]. It was noted that mortality was reduced with corticosteroid treatment [5].

Therefore, the first line of treatment includes systemic corticosteroid (prednisolone at 0.5 mg to 1.5 mg/kg/day), either oral or intravenous [9]. Combining azathioprine 1 to 3 mg/kg per day or mycophenolate mofetil 2 g per day with oral steroids is efficacious, but the optimal time for its commencement is still unclear [5]. It is suggested that systemic corticosteroid can be combined with immunosuppressive agents at the onset of therapy, in case of expected complications from prolonged use of corticosteroid therapy (>4 months) or dose-dependency above minimal therapy (10 mg/day) [9].

CONCLUSION

Our case highlights the pemphigus can masquerade many skin disorders especially dermatitis. Due to rarity of the disease, primary care doctors should have a high index of suspicions in the patient presented with a recurrent blistering skin lesion. Early referral to a dermatologist is needed to prevent morbidities and mortalities. The confirmation of diagnosis via skin biopsy and DIF. Earlier treatment will improve outcomes, improved quality of life and recovery.

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Declaration of interest: No conflict of interest is declared by authors.

Data sharing statement: Data supporting the findings and conclusions are available upon request from the corresponding author.

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