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Case Report Open Access

A Rare Presentation of Pemphigus Erythematosus as Pustules

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Abstract

Pemphigus erythematosus (PE) is an autoimmune blistering disease that is characterized by erythema and blisters, but rarely pustules, on the upper torso, face, and scalp. A 37-year-old woman presented with a 20 day history of intense pruritus as well as multiple papules and pustules on the trunk and face. Initially, the disease was misdiagnosed as impetigo due to the presence of pustules, nontypical lesions, and pathology results. Histopathology of new bullas showed epidermal hyperkeratosis, blistering and typical acantholysis, and the presence of diffuse inflammatory cells. Direct immunofluorescence showed intraepidermal, intercellular deposition of IgG autoantibodies. A diagnosis of pemphigus erythematosus was made. Systemic steroid therapy was effective in controlling the disease. To the best of our knowledge, this is the first case report on pemphigus erythematosus presenting with pustules.

Keywords: Pemphigus Erythematosus; Pustules; Misdiagnosis

Introduction

Pemphigus is an autoimmune bullous skin disease characterized histologically by acantholysis in the epidermis. There are four clinical variants of pemphigus: pemphigus vulgaris, pemphigus vegetans, pemphigus foliaceus, and pemphigus erythematosus (PE) [**1**]. Pemphigus erythematosus is a variant of superficial pemphigus with features of both lupus erythematosus and pemphigus. Its clinical manifestation is variable, often accompanied by small, flaccid bullae with scaling and crusting. Generally, the scalp, face, upper chest and back are affected, with an erythematosus scaly-to-crusted rash of the face presenting in the typical butterfly distribution [**2**]. It is also known as Senear–Usher syndrome and is often initially misdiagnosed due to its resemblance to lupus erythematosus or seborrhoeic dermatitis by the typical pemphigus distribution of erythematous scaly lesions in the seborrhoeic areas [**3**].

Case Report

In August 2013, a 37-year-old woman presented to our dermatologic department with a 20 day history of intense pruritus as well as multiple papules and pustules on the trunk and face. Twenty days earlier, this patient was admitted to the dermatological department of the local area hospital after presenting with scattered millet grain size papules, blisters with clear contents, and conscious pruritus on the back, which were diagnosed as contact dermatitis. Following diagnosis, the patient had been treated ineffectively with boric acid solution, furacilin solution, and mupirocin ointment. With time, new lesions including pustules erupted widely on the chest and face. The patient was then hospitalized with a primary diagnosis of pemphigus in the local area hospital. On admission, the patient was biopsied and then treated with oral dexamethasone tablets (3 mg per dose, 3 doses per day) for one week, showing an excellent response. However, the diagnosis based on pathology was reported as impetigo. For further diagnosis and treatment, the patient was referred to the dermatologic department of our hospital.

At the time of admission, physical examination revealed multiple vesicles, bullae, and pustular lesions, with some having clear contents and some being purulent, erosive plaques and crusted lesions with slight erosions and exudation on the back and chest (Figure 1A-C). In addition, there were erythematous scaly lesions on both sides of the nose wings (Figure 1D). Neither the oral or genital mucosa nor the nails were affected. Nikolsky's sign appeared to be positive, albeit somewhat indistinct. Based on the history of the disease and clinical features, impetigo, pemphigus, and dermatitis herpetiformis were considered for differential diagnosis.

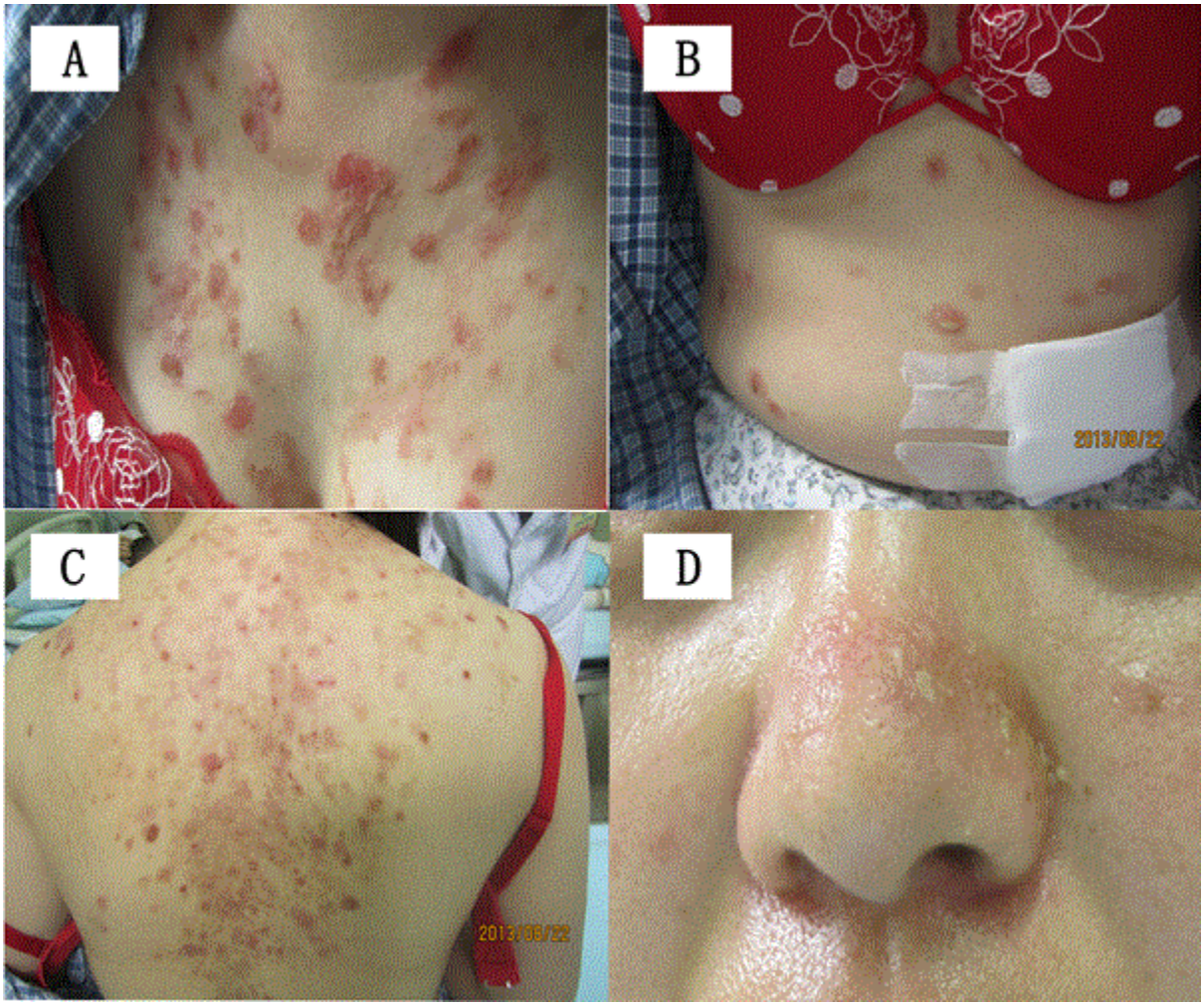

Figure 1: (A-C) Superficial crusted lesions widely distributed on the chest and back; (D) Erythematous scaly lesions on both sides of the nose wings

While consultations with the local area hospital regarding the pathological sections were undertaken, a battery of tests was nevertheless performed in order to enable diagnostication. Again, the diagnosis based on pathology was given as impetigo in our pathology department. The blood tests revealed eosinophilia while elevated total bilirubin while complement C3 (70.1 mg/dL) and C4 (15.0 mg/dL) were below the normal range. Other general examinations were normal.

Given the pathological manifestations of impetigo, the patient was treated orally with glycyrrhizin (80 mg/day), desloratadine dispersible tablets (5 mg/day), penicillin sodium (480 wu per dose, once every 12 hours), and topically with sodium fusidic ointment and other hospital preparations (e.g., chlorhexidine). Unfortunately, after 1 week, the patient did not show signs of improvement and therefore the treatment was deemed invalid and was promptly discontinued. Systemic cefazolin therapy was then given but the patient showed a weak response while new lesions continued to appear.

Because the therapy was unsuccessful, and clinical manifestations of the disease continued to be observed, we carried out repeated histopathologic and immunofluorescent examinations on skin biopsies taken from the patient. Tissue biopsy was performed on new bullas showing epidermal hyperkeratosis, blistering and typical acantholysis in superficial layers of the epidermis, and the presence of diffuse inflammatory cells. Direct immunofluorescence showed intraepidermal, intercellular deposition of IgG autoantibodies (Figure 2). Based on the clinical and laboratory results, and the close relationship between the butterfly erythema on the face of the patient and her response to treatment, a diagnosis of PE was made.

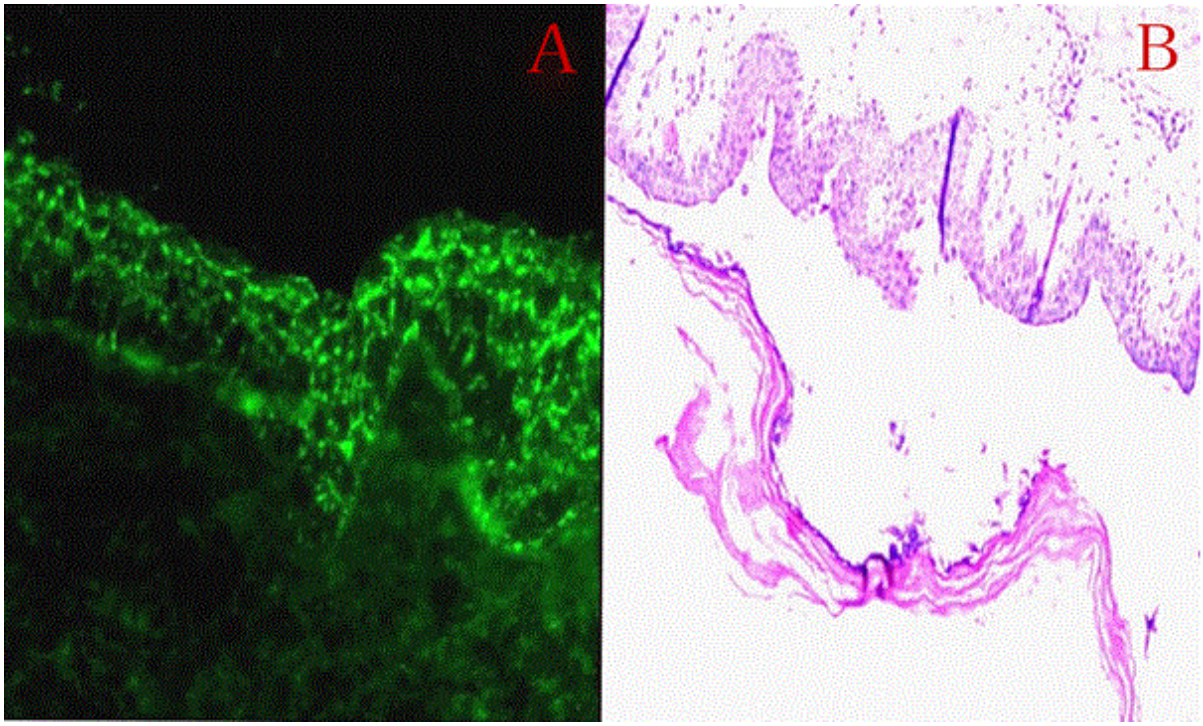

Figure 2: (A) Direct immunofluorescence performed on new bullas revealing intraepidermal intercellular deposition of IgG autoantibodies (FITC×400); (B) Typical acantholysis in superficial layers of the epidermis (H&E ×400).

After further pathological tests, systemic treatment with methylprednisolone (40 mg/day) for three days resulted in total clinical remission, after which she was discharged from our hospital.

Discussion

Clinically, lesions of PE usually begin with multiple pruritic, crusted, coin-sized patches on the upper torso, face, and scalp [**1**]. However, our clinical case study suggests that patients with PE may also show prominent pustular skin lesions.

PE in its typical presentation is infrequent, while PE as pustules is very rare. However, this rarity may be ascribed to the difficulty in its diagnosis. There are a few other clinical case studies that have found pemphigus presenting with pustules [**4**,**5**]. Most of the patients described showed extensive pustules that were quite different from the vesiculopustular lesions seen in our patient. In our patient, we observed papules and blisters accompanied by pruritus. Owing to its location on the body, pus deposits formed on the bottom of the pustules, taking on the characteristic lunate appearance of empyema with some undergoing breaks leading to red moist erosions. As a result, its clinical differentiation from seborrhoeic dermatitis and impetigo with bullae was made difficult.

Seborrhoeic dermatitis is a type of chronic relapsing inflammatory skin condition characterized by scaling and poorly defined erythematous patches in sebaceous glands-rich areas. PE differs in that it is an antibody-mediated autoimmune disease directed at desmoglein (a desmosomal protein important in keratinocyte adhesion), resulting in intraepidermal bullae [**6**].

Impetigo with bullae is a contagious infection of the skin in children, with purulence, typically caused by staphylococcus aureus. Bullae are of various sizes and are thin-roofed, flaccid or soon become flaccid, and initially contain clear yellow fluid before becoming turbid and pyoid after one day [**5**]. Histological examination reveals blisters in the subcorneal or granular region, while bacterial culture of pus yields staphylococci which are successfully treated with antibiotic therapy. However, in our patient, direct immunofluorescence demonstrated intercellular IgG deposition as in other forms of pemphigus, while histological examination revealed typical acantholysis in superficial layers of the epidermis. In this case, antibiotic therapy was ineffective. At the later stages, the cutaneous features of butterfly erythema appeared. These results strongly suggested a diagnosis of PE in our patient which was effectively controlled by systemic steroid therapy.

Our case demonstrates that PE may present with prominent pustular skin lesions. To our knowledge, very few cases with vesiculopustular lesions-as seen in our patient-have been reported. Diagnosis of PE with pustules will aid in determining the prognosis and course of the disease. Clinicians should to be vigilant for the rare occurrence of pustules in PE.

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