

**PEMPHIGUS VULGARIS MASQUERADING AS  
SUBCORNEAL PUSTULAR DERMATOSES – A CASE  
REPORT**

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**Abstract**

Both pemphigus vulgaris and subcorneal pustular dermatoses are intraepidermal blistering disorders though the treatment for the two varies. We present a 26 years old male patient with multiple vesicles and bullae filled with clear fluid as well as pus, hypopyon sign present, predominantly over the trunk and crusted lesions over the scalp. The patient did not have mucosal involvement at presentation and Nikolsky's and bulla spread sign were negative. A clinical diagnosis of subcorneal pustular dermatosis and IgA pemphigus was made; however, direct immunofluorescence was suggestive of pemphigus vulgaris as was histopathology examination. The patient responded to treatment with oral corticosteroids. A previous case report of pemphigus foliaceus presenting as IgA pemphigus and responding to dapsone has been reported and so has a report of pemphigus vulgaris presenting with multiple pustules.

**Key words:** Pemphigus vulgaris; IgA pemphigus; Subcorneal pustular dermatosis

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**Introduction**

Pemphigus is a group of chronic autoimmune bullous diseases of the skin and/or mucosae characterized by the presence of desmoglein 3 and/or 1 antibody. There are mainly two types of pemphigus, pemphigus vulgaris (and its variant pemphigus vegetans), and pemphigus foliaceus (and its variant pemphigus erythematosus) [1]. In pemphigus vulgaris flaccid blisters filled with clear fluid arise either on normal skin or an erythematous base. Mucosal erosions may precede cutaneous lesions by many days or months. Direct immunofluorescence shows deposition of IgG and C3 in the intercellular spaces in a 'fishnet' pattern.

Subcorneal pustular dermatosis presents with flaccid pustules in which the pus characteristically accumulates in the lower half. Both direct and indirect immunofluorescence is negative.

IgA pemphigus presents as flaccid vesicles or pustules usually associated with pruritus. The lesions have a predilection for the axillae and groins. Intercellular IgA deposition is seen in the epidermis either at different levels or throughout.

This case is unique as the patient presented with lesions which

were clinically suggestive of subcorneal pustular dermatosis but DIF proved otherwise.

**Case Report**

26 years old married male, fisherman, presented to us with painful lesions over the scalp since two months and fluid and pus filled vesicles and bullae over the trunk and upper limbs since 5-6 days associated with itching. Few lesions had ruptured after scratching to give rise to erosions. No history of spontaneous rupture of lesions, peripheral extension or history suggestive of healing with milia formation. There was no history of mucosal lesions at presentation. No history of prior drug intake, or any constitutional symptoms. No history suggestive of wheal formation or severe itching prior to development of lesions. There was no history suggestive of systemic involvement. On examination he had crusting, erosions and matting of hair over the scalp. Multiple tense vesicles and bullae filled with both clear fluid and pus predominantly over the trunk (Fig. 1) and few over the arms, neck and medial aspect of thigh. Hypopyon sign was positive (Fig. 2).

Few erosions over trunk were seen. Oral cavity examination revealed single erosion over the left buccal mucosa. Genitals were normal. Both marginal and direct Nikolsky's and bulla spread sign were negative. A clinical diagnosis of subcorneal pustular dermatoses and IgA pemphigus was made and he was investigated.

Tzanck smear showed only pus cells and gram staining from pustule revealed gram positive cocci and pus cells. A perilesional skin biopsy for direct immunofluorescence showed intercellular staining with Ig G and C3 and skin biopsy from a vesicle showed suprabasal acantholysis with cleft formation and detached roof of the bullae. (Fig. 3) Basal cells with increased melanin

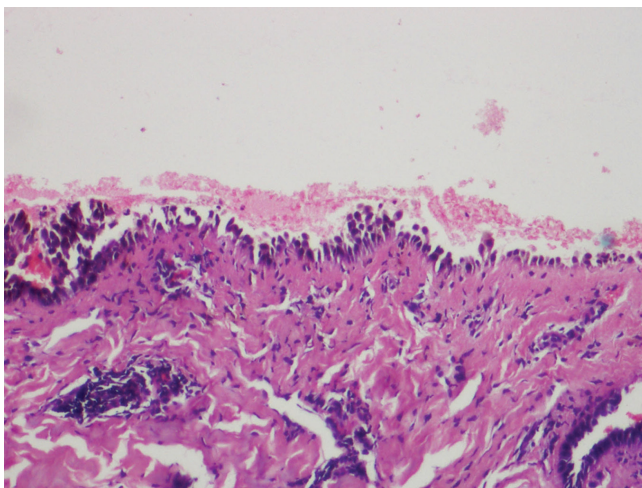
pigmentation arranged in a row of tomb stone fashion in the base of the bulla were also seen. Indirect immunofluorescence showed intercellular staining with Ig G at 1:100 dilutions. ELISA for desmoglein 1 and 3 was positive with >200 RU/ml. Complete blood count, fasting, post prandial blood sugars, renal function, liver function tests and urine examination were within normal limits. Chest x-ray was normal. Serum protein electrophoresis showed normal pattern. On the basis of DIF, IIF, ELISA and histopathology findings a diagnosis of pemphigus vulgaris was made and patient as treated with oral prednisolone 1mg per kg body weight, to which he had a dramatic response.



**Figure 1. Multiple tense vesicles and bullae filled with both clear fluid and pus predominantly over the trunk.**



**Figure 2. Hypopyon sign positive.**



**Figure 2. suprabasal cleft with few acantholytic cells and tombstoning of basal cells (the roof of bulla has denuded). (H&E 200X)**

## Discussion

The typical presentation of pemphigus vulgaris is as flaccid blisters which may occur anywhere on the skin surface. The

blisters burst to give rise to erosions which have a tendency to spread at their periphery. 50-70 % of the patients may present with oral erosions which are irregularly shaped over the palate or buccal mucosa [2].

Many atypical presentations of pemphigus vulgaris have been reported. A 60 years old patient presented with ulceration over bilateral dorsa of feet which persisted for four months before the characteristic lesions of pemphigus vulgaris appeared. A 30 year old female patient presented with a single erythematous crusted plaque in the right nasal wing. On histologic examination and immunofluorescence it was found to be pemphigus vulgaris [3]. A 50 years old male patient presented with erythematous scaly plaques and was diagnosed as psoriasis which however did not respond to treatment for the same.

Direct immunofluorescence revealed pemphigus foliaceus [4]. Pemphigus foliaceus masquerading as IgA pemphigus and responding to dapsone has been reported [5]. There has also been a case report of pemphigus foliaceus presenting with prominent neutrophilic pustules where the lesions mimicked subcorneal pustular dermatosis clinically [6]. In both cases a correct diagnosis was made based on the findings of direct immunofluorescence. No case reports have yet been reported where pemphigus vulgaris presented with subcorneal pustular dermatoses like lesions.

In our case a diagnosis of subcorneal pustular dermatosis and IgA pemphigus (subcorneal pustular dermatosis type) was made clinically but direct and indirect immunofluorescence showed features of pemphigus vulgaris. The treatment for pemphigus vulgaris and IgA pemphigus/subcorneal pustular dermatosis varies and thus a correct diagnosis is a must for proper treatment.

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