

Pemphigus Erythematosus in a Middle Aged Nepali Male: Case Report and Literature Review

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Abstract

Introduction: Pemphigus erythematosus, also known as the Senear-Usher syndrome, is an autoimmune condition where there is overlap between the clinical and immunological features of pemphigus foliaceus and lupus erythematosus. Patients present with erythematous, scaly, and crusted lesions in seborrhoeic distribution along with malar rash in butterfly distribution. Diagnosis is based on clinical findings, characteristic histopathologic changes, presence of antinuclear antibodies and immunolabelling in direct immunofluorescence test.

Case Presentation: We report a case of a middle-aged Nepali male who presented with the complaints of photosensitivity, itchy rash on the cheeks and flaccid blisters and crusted erosions on the chest for a few weeks. His diagnosis of Senear-Usher syndrome was based on clinical, histological and immunological findings. He was subsequently started on oral prednisolone and hydroxychloroquine sulfate, along with topical corticosteroids, which induced remission within the first week. He is currently on maintenance therapy without any relapse. This is to our knowledge, the first case of Senear-Usher syndrome reported from Nepal.

Conclusion: Topical and oral corticosteroids along with hydroxychloroquine sulfate should be considered first line in the treatment of Senear-Usher syndrome.

Keywords: Pemphigus erythematosus; Senear-Usher syndrome; Immunological features

Abbreviations: ANA: Antinuclear Antibody; dsDNA: Double Stranded Deoxyribonucleic Acid; ESR: Erythrocyte Sedimentation Rate; IgG: Immunoglobulin G; C3: Complement 3, mL: Millilitre; mm: Millimetre; mm/Hg: Millimetre per 1 Column of Mercury

Introduction

Pemphigus erythematosus, also known as the Senear-Usher syndrome, is one of the variants of pemphigus, which is an autoimmune bullous disorder [1]. It is characterized by overlapping features of lupus erythematosus and pemphigus foliaceus. While the erythematous lesions on the face in a butterfly distribution resemble lupus erythematosus, the vesicles and crusted plaques on the trunk simulate pemphigus foliaceus [2]. Immunological features of both the diseases are also present. It is a rare condition, has been reported infrequently, with one review quoting a total of only 116 cases [1], and to the best of our knowledge, has never been reported from Nepal. We report a case of a middle aged male presenting with features of pemphigus erythematosus with the relevant literature review.

Case Presentation

Medical history

A 42-year-old male presented with small fluid filled lesions on his scalp on and off for the last 7 months. The patient was apparently well 7 months back, when he developed red itchy lesions on the malar regions of the cheeks and the bridge of the nose in a butterfly distribution. This was accompanied by photosensitivity and increased loss of hair on the scalp but there were no oral or nasal ulcers, joint pain, chest pain and neurological symptoms. At the same time, he developed fluid filled lesions on the scalp, chest and upper back, which followed the previous course. The blisters were flaccid containing clear fluid, which ruptured within 1-2 days or immediately after manipulation. Rupture of the vesicles resulted in the formation of crusts, which healed with residual hyperpigmentation. Crops of new lesions occurred, which followed

a similar course to leave hyperpigmentation. He visited a local clinic where he was given oral cloxacillin 500 mg capsules to be taken four times a day for 10 days and topical combination of betamethasone valerate cream and neomycin cream to be applied on the affected sites for 2 weeks. After administering these medications as prescribed, his lesions healed in a month.

He was symptom free for the next 6 months following which there was severe relapse of the previous symptoms of fluid filled lesions on the scalp, chest and upper back, red rash on the face in a butterfly distribution, photosensitivity, malaise and diffuse loss of hair on the scalp. There was no history of prior admission to any hospital for medical or surgical reasons and no history of any chronic disease. There was no sudden gain or loss in weight and he was not on any medication prior to the eruption of the lesions.

Clinical presentation

When he presented to our department, we initially suspected a bullous disorder. His vitals were stable; there was no pallor, icterus, lymphadenopathy, cyanosis, clubbing, oedema or dehydration. No abnormalities were detected on systemic examination.

On dermatologic examination, he had Fitzpatrick skin type IV. Flaccid vesicles and crusted erosions and plaques were present on the scalp, upper chest and back (Figure 1), malar rash in a butterfly

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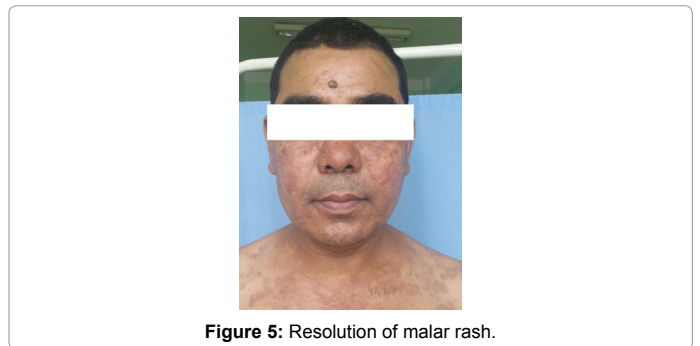
distribution on the face and discoid rashes on the forehead (Figure 2). Nikolsky sign was positive while the Asboe-Hansen sign was negative. Oral and genital ulcers were absent.

Laboratory tests

His laboratory investigations revealed ANA positivity with dsDNA value of only 5 IU/ml. There were no abnormalities in his haematological and biochemistry profiles and his routine urine microscopic examination did not reveal any anomalies except for a raised ESR of 45 mm in 1 hour.

Histopathological and immunofluorescence studies

Skin biopsies were taken from a fresh blister for histopathologic examination and from perilesional skin for direct immunofluorescence study. The histopathological examination revealed an acanthotic epidermis with large subcorneal blisters containing numerous neutrophils (Figure 3). Acantholytic cells were not detected. The



dermis showed increase in number of capillaries and inflammatory cell infiltrates. The direct immunofluorescence revealed IgG 3+ and C3 1+ in intercellular space of epidermis and at the dermo-epidermal junction.

Treatment

He was subsequently diagnosed as suffering from lupus erythematosus and was put on topical fluticasone propionate 0.05% w/w, oral prednisolone at a dose of 1 mg/kg once daily, oral hydroxychloroquine sulfate 200 mg twice daily, a broad-spectrum sunscreen and prophylactic doses of rabeprazole, calcium and alendronate.

Outcome

He made a full recovery (Figures 4 and 5) within a month and has been maintained on hydroxychloroquine sulfate 100 mg twice daily and tapering doses of oral prednisolone.

Discussion

Pemphigus erythematosus, is a rare condition that combines the clinical and immunological features of pemphigus foliaceus and lupus erythematosus [3]. Clinical features of the Senear Usher syndrome include the pemphigus foliaceus-like scattered scaly flaccid blisters with erosions and crusts on 'seborrhoeic' areas of the scalp, face, chest and upper back along with malar rash simulating lupus erythematosus [2] with absence of mucous membrane involvement [4]. The immunological features include IgG and C3 in the suprabasal layers as in pemphigus foliaceus and along the basement membrane zone similar to lupus erythematosus [5].

Many authors have disputed the existence of this syndrome as a distinct entity and consider it to be an abortive form of pemphigus foliaceus [6]. This could be supported by the study done by Otkarina et al. [7], where they demonstrated deposition of antibodies against desmoglein 1 on the basement membrane zone mimicking the lupus band test. This could explain the direct immunofluorescence findings when ANA reactivity is negative or weakly positive.

Other authors have however stated that while pemphigus erythematosus has concomitant basement membrane zone deposition of immunoglobulin and complement in addition to intercellular epidermal staining, pemphigus foliaceus lacks basement membrane zone staining [8]. Studies have confirmed multiple-autoimmune process in this syndrome [7,9]. In fact, Chorzelski et al. [9] described the presence of immunoglobulins and complement at the basement membrane zone along with anti-epithelial antibodies in this syndrome in 1968. Then, 44 years later, Perez-Perez et. al. [5] not only confirmed the co-existence of anti-epithelial and antinuclear antibodies but also ruled out cross-reactivity by immunochemical studies. These findings do support the hypothesis claiming the Senear-Usher syndrome to be a distinct entity and not just a variant of pemphigus foliaceus. Our patient satisfied both clinical and immunological features of the Senear-Usher syndrome.

The differentials other than pemphigus erythematosus that can be considered are seborrheic dermatitis, bullous pemphigoid, and dermatitis herpetiformis. The characteristic appearance of seborrheic dermatitis is that of red, flaking, greasy scales on the scalp, nasolabial folds, ears, eyebrows and chest and not that of flaccid vesicles and bullae. The histopathology usually reveals 'squirting' of granulocytes from the dermal papilla. Oedema, hyperkeratosis and focal parakeratosis are also commonly seen. Bullous pemphigoid lesions are tense blisters occurring on normal or erythematous skin associated with severe pruritus. The blisters are typically located on flexor surfaces of the arms and legs, axillae, groin, and abdomen. Dermatitis herpetiformis has a chronic course, is characterized by pruritic papules and vesicles distributed on extensor surfaces, is associated with gluten sensitive enteropathy, demonstrates presence of neutrophils in dermis in histopathological sections, and IgA deposition on the papillary tips in direct immunofluorescence.

The course is often chronic lasting months to years. The treatment options for patients with pemphigus erythematosus include oral and topical corticosteroids, β -blockers, immunosuppressive agents, Jamarson therapy, antimalarials, sulfapyridine and anticholinergic agents [1]. The use of these medications can lead to partial or complete remission. However, they are associated with significant side effects and have to be monitored appropriately.

To the best of our knowledge, this is the first case report on this syndrome in Nepal. An extensive search of Pubmed, Ovid Medline, Ovid Embase and Google Scholar did not reveal any previously published case reports or case series from Nepal, although there have been case reports and case series published from the USA, India [6], Italy, Nigeria and Sudan.

Conclusion

Our patient was diagnosed with pemphigus erythematosus based on clinical and immunological findings. He responded promptly to topical and oral corticosteroids and hydroxychloroquine sulfate not necessitating the use of adjuvant immunosuppressants.

Topical and oral corticosteroids along with hydroxychloroquine sulfate should be considered first line in the treatment of such conditions.

Declaration

Ethics approval and consent to participate

Written informed consent was taken from the patient for collection

of photographs, digital recording of clinical information and for dissemination of the information in a printed or digital form. All efforts have been made to ensure anonymity of the subject. The patient has been informed of their right to withdraw consent at any future point in time before online publication.

Consent for publication

Written informed consent was taken from the patient for publication of this case report. The patient has been informed of his right to withdraw consent at any future point in time before online publication.

Availability of data and materials

A copy of all the records of the patient including the photographs can be retrieved from the corresponding author.

Competing interests

The authors declare that they have no competing interests.

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Authors' Contributions

BA was involved in writing the first drafts of case report and provided the photographs. SMMA was involved in critical revision of the manuscript and re-writing of the discussion and conclusion. LM searched all the relevant literature, re-wrote the report, collected the informed consent from the participants and made revisions to the draft. All the authors gave their final approval for this version of the manuscript to be published.

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