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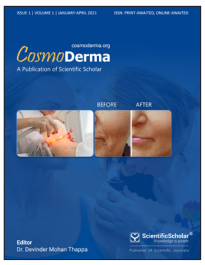

Letter to the Editor

A difficult case of pemphigus

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Dear Sir,

The term "pemphigus" refers to a group of autoimmune blistering diseases of skin and mucous membranes characterized by erosions, ulcerations, and crusting. Infections caused by members of the herpesviridae family are commonly seen in immunocompromised hosts which further complicate the course of disease process, thereby posing as a diagnostic and therapeutic challenge. We report a case of pemphigus erythematosus (PE) complicated by cutaneous herpes simplex infection.

A 40-years-old female presented to the dermatology OPD with blackish-brown colored lesions on both the cheeks for 1.5 months, a solitary wound over the scalp for 1.5 months, and fluid filled lesions over the body for the past 5 days. History of application of topical ayurvedic medication was present after which patient complained of increased pruritus and worsening of symptoms.

History of photosensitivity was also present. There was no history of fever, itching before onset of lesions, mucosal involvement (eyes/oral cavity/genitalia), drug intake, joint pain, weight loss, loss of appetite, insect bite, etc. Past history and family history were insignificant. On cutaneous examination, a solitary ulcer of size 3 × 3 cm was present over the scalp with yellowish slough present over the floor of the ulcer. Blackish-brown crusted plaques were present on malar area of the face while extensive crusting was present over seborrheic area of the body. There were multiple erosions of variable shape and sizes present all over the body. [Figure 1] Nikolsky's sign was positive while the bulla spread sign negative. The clinical differential diagnoses were pemphigus foliaceous, PE, systemic lupus erythematosus, pemphigus vulgaris, and bullous pemphigoid. Systemic examination was within normal limits. Blood tests showed leukocytosis (total leukocyte count – 17610 cells/mm^3) and raised erythrocyte sedimentation rate (erythrocyte sedimentation rate in 1st h – 62 mm/h). An incisional skin biopsy was subsequently taken and sent for examination. Histopathology showed subcorneal blistering and acantholysis. Special investigations such as ANA were found to be positive showing homogenous pattern. Direct immunofluorescence (DIF) was done with the epidermis exhibiting intercellular deposits of IgG and C3c in a characteristic fish net pattern [Figure 2]. The diagnosis of PE was made based on classical facial rash, ANA positivity, and fish net pattern on DIF (clinicopathological correlation). The case was managed with oral prednisolone 60 mg daily along with systemic antibiotics – intravenous cefotaxime 500 mg and metronidazole 100 mL infusions once every 8 h. Sodium fusidate cream was applied over erosions and raw areas. Povidone-iodine shampoo was advised for scalp lesions along with antihistamines for pruritis. Surprisingly, after 10 days of initiation of therapy, patient developed oral and genital ulcers. Tzanck smear from the lesions showed multi-nucleated giant cells. Patient was started on valacyclovir at a dose of 1 g every 8 h for 5 days. In addition, azathioprine at a dose of 50 mg twice daily was started as a steroid sparing agent. The

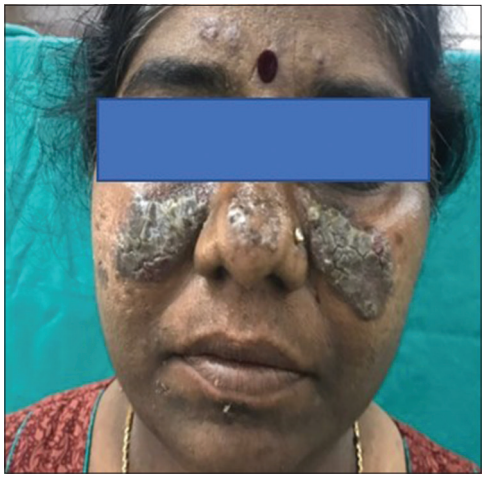

Figure 1: Blackish-brown crusted plaques present over the malar area of face.

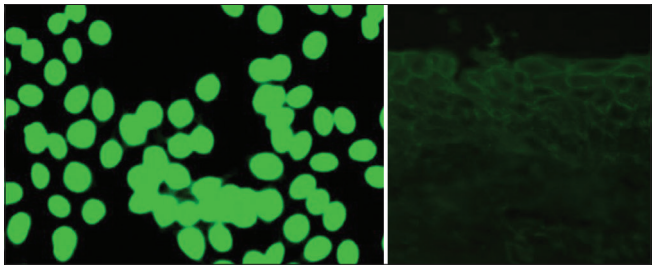

Figure 2: ANA-positive showing homogenous pattern. Direct immunofluorescence showing oriented stratified squamous epithelium and epidermis exhibiting intercellular deposits of IgG and C3c.

oral and genital lesions subsided within 5 days of antiviral therapy. Subsequently steroids were tapered to dose of 20 mg daily at the time of discharge. The patient was periodically followed up and showed complete remission in 6 months.

PE is a rare autoimmune skin condition with clinical, histopathological, and serological features that show overlap between lupus erythematosus and pemphigus foliaceus.[1,2] Cutaneous Human *herpesvirus* Type 1, Type 2 *herpes simplex* virus 2 – (HSV_2), and Type 3 varicella zoster virus are known to occur in various bullous disease of the skin, particularly when patients are receiving immunosuppressive therapy. The severity of this infection ranges from mild to fatal, with dissemination of lesions. The possible mechanisms include interruption of T-cell tolerance, up-regulation of pro-inflammatory factors in those genetically predisposed, structural damage to keratinocytes resulting in exposure of endogenous antigens, and epitope spreading.[3-5] The morphology of PE and HSV infection closely resemble each other and often pose a diagnostic and therapeutic challenge as seen in our case. The prudent use of steroids is therefore to be kept in mind during management of such cases. While localized HSV infection per se could mimic immunobullous disease, it may also simulate an active flare of the immunobullous disease in case of extensive disease. The refractoriness to conventional therapy in PE should raise the possibility of a herpes simplex skin infection, so that necessary and timely laboratory investigations may be performed for the detection of viruses with tropism to skin. Timely management not only improves patient well-being but also helps in saving lives.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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