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RECURRENT ZOSTERIFORM ERUPTIONS IN A PATIENT WITH PEMPHIGUS VULGARIS

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A 31 year old female presented with painful oral ulceration, crusted erosive plaques of scalp and flaccid bullae of torso in 2017. She was diagnosed with pemphigus vulgaris based on characteristic clinical, histological and immunofluorescence findings and treated with monthly dexamethasone pulses and mycophenolate mofetil but unable to achieve remission for 3 years. Repeated evaluation for associated autoimmune disorders and malignancies were negative. While on treatment she developed recurrent episodes of blistering eruptions with dermatomal distribution.

Figure 1: Blistering eruptions involving various body regions with dermatomal distribution as depicted in the following figures.



Repeated tzanck smears, skin biopsies and immunofluorescence studies were compatible with pemphigus vulgaris and failed to provide any features in favour of herpes zoster (VZ)/ herpes simplex (HS) infection. Moreover, VZ, HSV, IgM and PCR were negative. She was later started on combination therapy with IV Ig and rituximab with an excellent response (Figure 1).

Figure 2: Following images depict resolution of skin lesions after the Rituximab treatment Skin biopsy of patient with pemphigus vulgaris direct immunofluorescence of pemphigus vulgaris



Pemphigus vulgaris-the most common and representative clinical form among the pemphigus group of blistering disorders, is recognized to have atypical clinical presentations such as isolated scalp erosions, foot ulcers, paronychia, pompholyx like presentation and macroglossia(1). Zosteriform/grouped vesicular blistering eruptions in a patient with diagnosed pemphigus patient on treatment can be due to several possibilities which include recurrent herpes infection (HSV/VZV) on the background of immune suppression, herpetiform pemphigus, isotopic phenomenon(2).

Figure 3: Skin biopsy of patient with pemphigus vulgaris

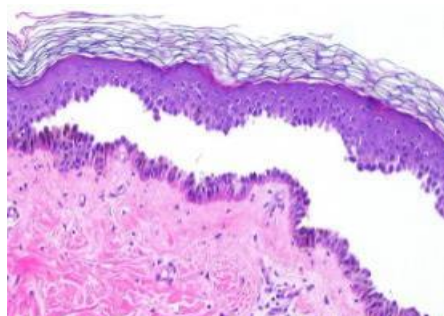
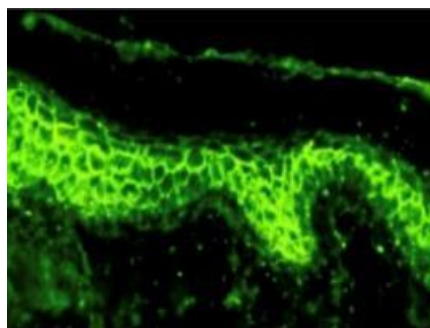


Figure 3: Direct immunofluorescence of pemphigus vulgaris pemphigus vulgaris



However, the zosteriform morphology is not considered as a morphological variant of PV in the medical literature up to date(3-5). Based on this patient's clinical presentation and investigation findings we thought the possibility of pemphigus vulgaris presenting with zosteriform morphology-could be an atypical and treatment modified variant but we had limited access to viral serological and PCR analysis and we were unable to arrange desmoglein ELISA which would be very helpful to differentiate our differentials(6). HSV and other herpes group of viruses are known to play a complex role in the pathogenesis of pemphigus, though the exact Patho mechanisms are still under evaluation(7,8). (may trigger/exacerbate PV and may cause treatment resistant disease) we consider the possibility of above mechanisms in our patient considering the zosteriform morphology, however this thought need lot of further studies.

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