

Severe COVID-19 along with Cytokine Storm in Pemphigus Vulgaris Managed Successfully with Dexamethasone Pulse Therapy

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During coronavirus disease-2019 (COVID-19) pandemic, the management of autoimmune disorders like pemphigus vulgaris poses a great challenge for dermatologists. Pemphigus is an autoimmune blistering disease, which presents with painful erosions over the skin and mucous membranes.¹

A 38-year-old female was admitted to our emergency department with complaints of multiple painful erosions in the oral cavity, over the trunk, and extremities for the last 2 months with intermittent high-grade fever for the last 4 days. The disease started 2 months back as vesicles and bullae over the trunk spontaneously ruptured to form nonhealing, expanding painful erosions. Dermatological examination revealed multiple erosions over the trunk, buttocks, and extremities and bilateral buccal mucosa (Figs 1A to C). The patient was clinically diagnosed with pemphigus vulgaris with suspected secondary infection and was managed in a high-dependency unit. All routine investigations were sent. We initiated our management with intravenous antibiotics and steroids (prednisolone 1 mg/kg body weight). Apart from raised total leukocyte count (TLC 11,600/mm³), other laboratory parameters were within normal limits. On the 4th day, the patient developed sudden shortness of breath with an increase in respiratory rate (RR 39 rate/minute), and her oxygen saturation was dropped below 92% at room air. The oxygen supplementation was started to maintain oxygen saturation above 94%. We send the nasopharyngeal swab for reverse transcription-polymerase chain reaction (RT-PCR) which was tested COVID-19 positive. On the 5th day, the patient has features of cytokine storm as inflammatory markers, like CRP, IL-6, and D-dimers, were raised. The severity of pemphigus vulgaris gradually increased in the form of expanding skin erosions covering around 60% of the total body surface area. On the background of the increased severity of pemphigus vulgaris and possible cytokine storm syndrome, we started a pulse dose of dexamethasone therapy (100 mg dexamethasone dissolved in 500 mL of 5% dextrose solution over and it was given over 2 hours for three consecutive days). The dexamethasone was added in dextrose to avoid the risks of steroid-induced hypernatremia and hypokalemia which may be more common with normal saline. During this pulse dose therapy, we closely monitored the vitals, serum electrolytes, and blood sugar. After completion of 3 days of dexamethasone pulse therapy, we observed a gradual improvement in her clinical condition and biochemical parameters. Her oxygen requirement reduced to 4 L/minute. While receiving pulse dexamethasone, she did not develop any electrolyte abnormalities and her blood sugar was maintained between 140 and 180 mg/dL. On the 9th day, the patient

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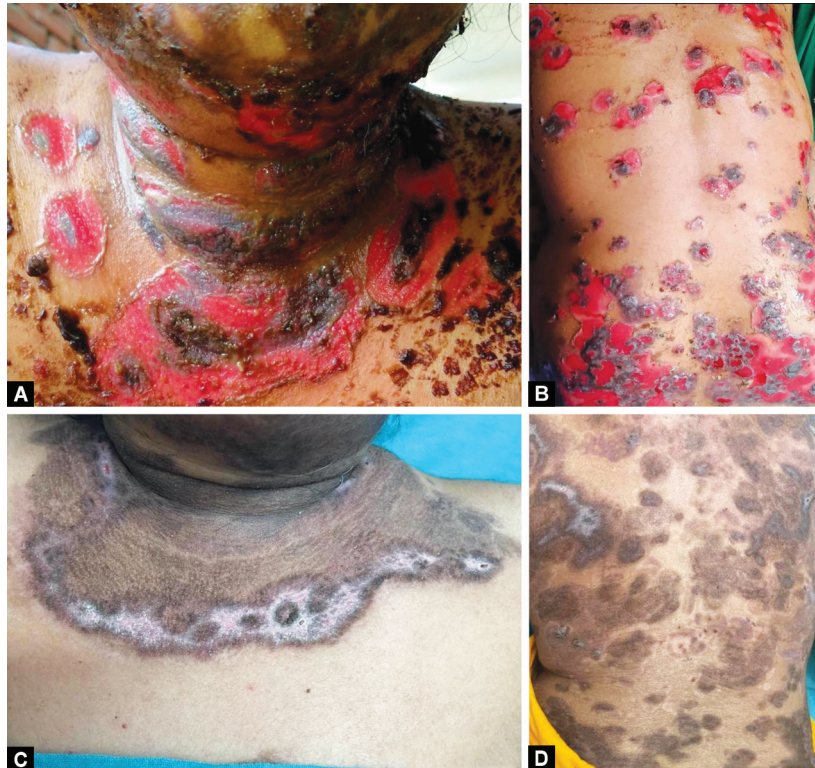
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was discharged. After 6 weeks of postpulse dexamethasone follow-up therapy, the erosions were healed completely with postinflammatory hyperpigmentation (Figs 1C and D). Her serum electrolytes were within the normal limit, and there were no signs and symptoms suggestive of any secondary opportunistic infection. Systemic corticosteroids are first-line drugs in treating pemphigus vulgaris.² Recently, high-dose corticosteroid pulse therapy is being practiced to control the cytokine storm of COVID 19.³ Glucocorticoids (GCs) exhibit anti-inflammatory and immunosuppressive effects through genomic action, activating the mitogen-activated protein kinase (MAPK) signaling pathway, and at high concentration, these GCs result in rapid immunosuppression using a non-genomic mechanism.⁴ Our patient was having features of severe pemphigus vulgaris with erosions all over the body and oral mucosa, and use of pulse dose of dexamethasone helped in a rapid clinical improvement of both pemphigus vulgaris and cytokine storm syndrome. However, to avoid further immunosuppression we have not used cyclophosphamide in our case. Hence, all patients on systemic corticosteroids may require a follow-up for at least 6 weeks to look for the development of any opportunistic infections. However, our patient did not have any signs and symptoms suggestive of opportunistic infection after 6 weeks of follow-up.

Informed Consent

Informed consent was taken from the patient.



Figs 1A to D: (A and B) Multiple erosions over trunk and chest; (C and D) Complete healing of erosions posttreatment

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