

Pemphigus vegetans arising in umbilicus: Successful clearance with intralesional steroid

Sir,

Pemphigus vegetans is a rare clinical variant of pemphigus vulgaris accounting for 1%–2% of all pemphigus.^{1,2} All subtypes of pemphigus are known to affect all clinical sites, but there is a definite predilection for the oral mucosa and intertriginous areas.¹ Though the etiopathogenesis of pemphigus vegetans is not entirely clear, vegetating lesions often localize to intertriginous areas attributed to the occlusion and maceration in these areas with secondary bacterial infection.¹⁻³ On literature search, we found only one case report of a patient treated with systemic immunosuppressants having pemphigus vegetans lesions located on umbilicus.⁴ We report here a case of pemphigus vegetans originating on the umbilicus treated successfully with intralesional steroids.

A 74-year-old man presented to our outpatient clinic with a 2 months history of asymptomatic nodules in the right inguinal region and around the umbilicus. The lesions had first begun on the umbilicus followed by similar lesions on the right groin. These had not responded to oral and topical antibiotics. On dermatological examination, nodular vegetating lesions of size 1 to 4 cm were

seen over umbilicus and right inguinal region [Figure 1]. There were no mucosal lesions. A skin biopsy was performed with differential diagnoses of dermatofibrosarcoma protuberans, pemphigus vegetans and cutaneous metastasis from gastrointestinal adenocarcinoma. On histopathological examination, there was marked acanthosis with pseudoepitheliomatous hyperplasia of the epidermis with underlying dermis showing mixed inflammatory cell infiltrate with predominance of eosinophils, edema and proliferation of capillaries. Acantholytic keratinocytes were detected in the basal epidermis. [Figure 2] Intercellular immunoglobulin G and C3 deposits were observed on direct immunofluorescent examination [Figure 3]. Based on clinical, histopathological and immunofluorescence findings, a diagnosis of pemphigus vegetans was established.

On laboratory investigations, he had mild anemia and high erythrocyte sedimentation rate. Tumor markers were negative. Mycological direct examination and cultures were negative. On bacteriological culture performed from umbilicus, *Streptococcus agalactiae* growth was observed.



Figure 1a: Multiple nodular lesions showing vegetative growth on umbilicus



Figure 1b: Multiple nodular lesions showing vegetative growth on the right inguinal region

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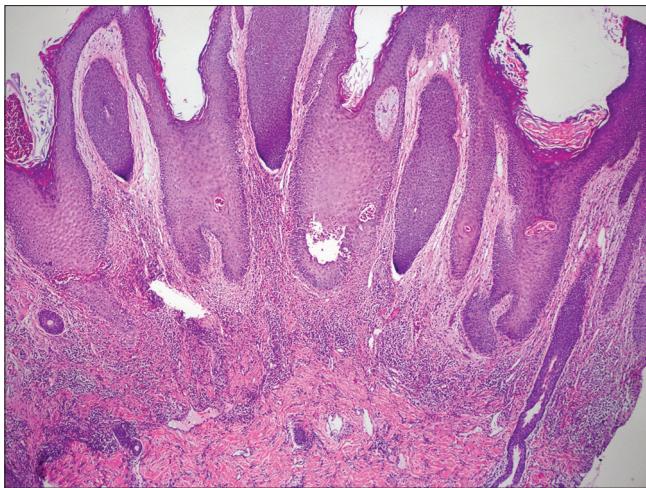


Figure 2a: Acanthosis with pseudoepitheliomatous hyperplasia in epidermis with intraepidermal split with acantholytic cells (H and E, $\times 400$)

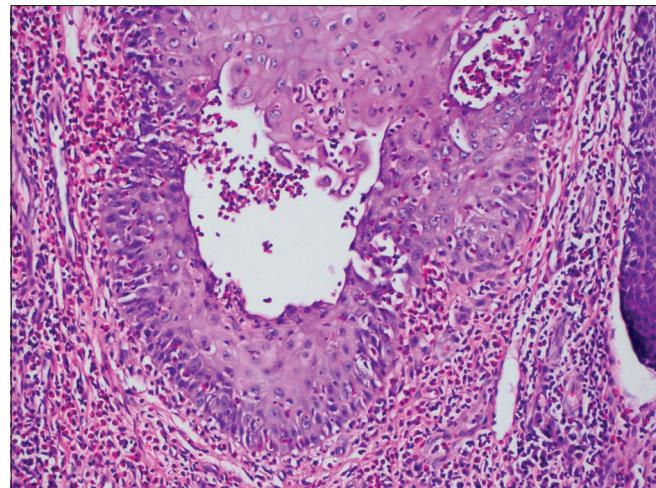


Figure 2b: Intraepidermal split with acantholytic cells, eosinophilic infiltrate in epidermis and dermis (H and E, $\times 200$)

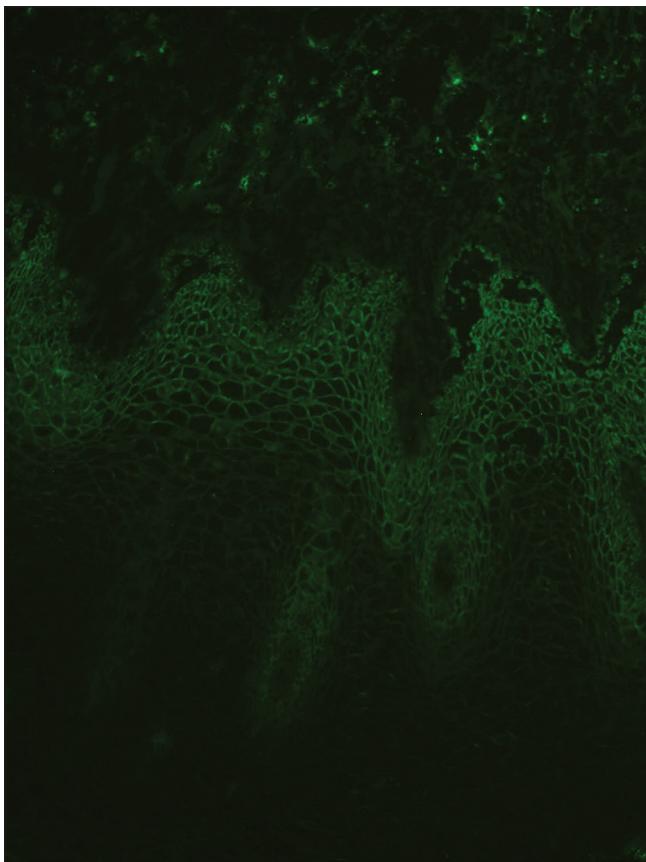


Figure 3: Intercellular immunoglobulin G deposition in epidermis, direct immunofluorescence examination ($\times 100$)

Daily dressings with rivanol 0.1% solution and systemic amoxicillin-clavulanic acid (1000 mg orally twice daily) were started for the patient. After completing a 10-day antibiotic course, the patient was started on intralesional steroid injections. We applied intralesional 40 mg triamcinolone acetonide, diluted with 3 ml of saline (10 mg/ml), once in 2 weeks for 4 sessions. Lesions



Figure 4a: Clinical view of the umbilicus after intralesional steroid treatment

healed completely with postinflammatory hyperpigmentation in 2 months and there was no recurrence on 9 month follow up [Figure 4].

Treatment of pemphigus vegetans is quite similar to that for pemphigus vulgaris and often requires systemic steroids and/or other immunosuppressants like azathioprine, mycophenolate mofetil, or cyclosporine. However, for the treatment of localized disease, intralesional steroids may be preferred.^{5,6} In our case, we achieved successful response using just intralesional steroid therapy.

In patients with limited lesions of pemphigus vegetans without mucosal involvement, intralesional steroid may be used as the first-line therapy to avoid the adverse effects of systemic steroids. These patients, however, need to be followed closely for recurrence as intralesional steroids are unlikely to cure the disease. In case recurrence is observed, systemic treatment may be administered at a later date.



Figure 4b: Clinical view of the right inguinal region after intralesional steroid treatment

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Conflicts of interest

There are no conflicts of interest.

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