

## Localized pemphigus vulgaris on cheeks responding to topical steroids

Sir,

Pemphigus vulgaris (PV) is an autoimmune blistering disease characterized by extensive mucocutaneous vesicobullous lesions. Generally, the mucosal lesions precede skin lesions by weeks or months. Severity of the disease may vary, from localized crusted erosions involving the skin or mucosa or both, to very extensive life-threatening mucocutaneous blistering and erosions.

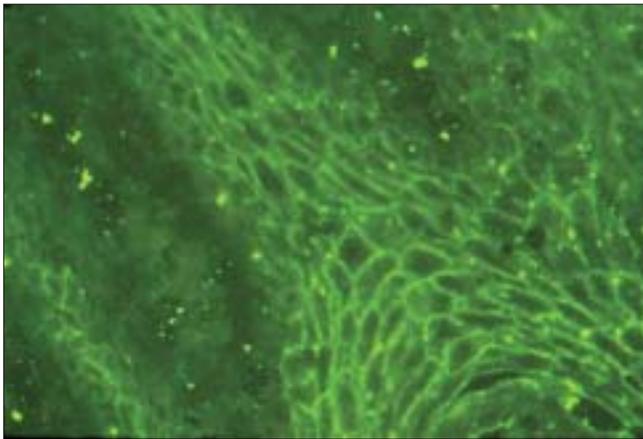
A 58-year-old woman presented with persistent crusted erosions on both cheeks just below her eyes

since four months. She complained of mild discomfort in the lesions. There was no history of drug intake or topical application, trauma, surgery, or irradiation in the involved area prior to eruption of lesions. She was prescribed oral antibiotics and topical framycetin cream (1% w/w) off and on with partial relief. Patient had been a field worker working in agricultural farms since childhood. Examination revealed an ill defined, superficial, crusted lesion with irregular margins and surrounding rim of erythema of about 2 x 3 cm in size below the left eye and about 1 x 1 cm sized lesion below the right eye [Figure 1]. There was no regional lymphadenopathy. She had lost her left eye due to blunt trauma in early childhood. A differential diagnosis of basal cell carcinoma and bullous impetigo were considered. Gram's smear from the lesion showed only polymorphs. Pus for culture from undersurface of the crust was sterile. A skin biopsy from left cheek revealed a suprabasal cleft with acantholytic cells and ulceration of the overlying epidermis. Subsequently, direct immunofluorescence (DIF) from the perilesional skin revealed IgG and C3 deposits in the intercellular spaces of the epidermis [Figure 2]. Based on histopathology and DIF, a diagnosis of localized PV was made and patient was treated with topical clobetasol propionate 0.05% cream. There was complete clearance of the lesions within four weeks without any recurrence in a six month follow-up.

Localized PV represents a different and relatively benign subset of PV and reflects one end of the spectrum of pemphigus with lesions usually limited to the sun exposed areas. Ultraviolet (UV) radiation has been used experimentally to induce acantholysis in patients with pemphigus foliaceus.<sup>[1]</sup> PV lesions developing after surgery, irradiation, and at the site of burn scar have been reported previously and these unusual presentations were attributed to Koebner-like phenomenon. Long standing localized PV lesions have also been described on oral mucous membrane, vaginal wall, and penile foreskin.<sup>[2,3]</sup> Egan *et al* and Lapiere *et al*, documented PV lesions limited to scalp that responded to topical steroids.<sup>[4,5]</sup> Baykal *et al*, reported four cases of localized PV over the nose and cheeks.<sup>[6]</sup> As in our patient, an initial clinical diagnosis of PV was not considered in any of them and diagnosis was based on histopathology, DIF, or serology results. The disease responded favorably to topical or systemic therapy with glucocorticoids alone or in combination with azathioprine. Localization of the PV lesions on nose and cheek, the light exposed



**Figure 1:** Crusted erosions with irregular margins on bilateral cheeks (stitch represents the biopsy site)



**Figure 2:** Direct immunofluorescence showing IgG deposits in the intercellular spaces with greater density in the lower part of the epidermis

skin, suggest that PV may be triggered or maintained by prolonged sun exposure. This case is reported for its atypical presentation and adequate response to potent topical steroid. It is suggested that localized PV should be considered in the differential diagnosis of persistent erosive lesions, more so when present over sun exposed areas.

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