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Case Report

Management of Recalcitrant Case of Oral Pemphigus with Intralesional Corticosteroids: A case report

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Abstract:
Oral pemphigus is one of the prevalent autoimmune oral mucosal diseases. However, the recalcitrant variant is infrequent and challenging to manage. These shallow ulcers may persist for several months and interfere with the normal state of health. We present a case of a female patient with long standing ulcers in oral cavity involving multiple sites. She was referred from dermatologist. She was on conventional corticosteroids and immunosuppressants, but had no relief symptomatically for the past 6 months. We managed the patient with intralesional corticosteroids and within 3 months, resulted in disease-free state. The treatment methods followed in our case could be successful to implement by oral physicians when the situation demands.

Keywords: *Oral Lesions, Intralesional corticosteroids, Pemphigus, Recalcitrant Type*
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Introduction

Pemphigus is an autoimmune blistering disease affecting middle-aged and elderly individuals, targeting skin and mucosal surfaces through IgG autoantibodies against desmoglein 3 and 1 proteins on keratinocytes. When both are involved, skin and mucosal surfaces are affected; if only dsg3 is involved, it primarily affects the mouth. Oral pemphigus presents as vesicles or bullae that rupture, forming chronic ulcers in areas like the buccal mucosa, gingiva, palate, and lips [1]. First-line treatment involves systemic corticosteroids, sometimes with agents like azathioprine or cyclophosphamide. This report details a rare case of oral pemphigus with an annual incidence of 0.1 to 0.5 per 100,000 [3]. The patient, unresponsive to six months of oral corticosteroids, was successfully treated with intralesional corticosteroid injections [4].

Case History

A 46-year-old woman presented with mouth ulcers, difficulty eating, bleeding while brushing, and increased salivation. She developed blisters that ruptured into ulcers and was diagnosed with oral pemphigus. Initially treated with topical Tenovate 0.05%, her condition worsened after 5-6 weeks, prompting systemic treatment with Omnacortil 20 mg and Endoxan 500 mg, twice daily. After six months with no improvement, she stopped taking Dexamethasone-Cyclophosphamide Pulse therapy due to cancer-phobia and was referred to our hospital.

Clinical Findings

On examination, the patient (161 cm, 49 kg) had multiple ulcers on the palate, buccal mucosa, and lower lip, with generalized gingival erythema [Figure 1]. Profuse bleeding and severe pain were noted. The pain score was 9 on the VAS. Recalcitrant pemphigus was diagnosed, with bullous pemphigoid and bullous lichen planus as differentials.

Investigations

The Tzanck test showed acantholytic cells, indicating pemphigus. (Figure 2A) Histopathology revealed acanthosis with suprabasilar bullae and acantholytic cells, distinguishing pemphigus from sub-epithelial blistering diseases. (Figure 2B) Direct Immunofluorescence (DIF) showed IgG positivity on desmosomes, confirming pemphigus vulgaris. (Figure 2C)

Therapeutic intervention

Systemic immunosuppressants were stopped, and intralesional corticosteroid injections (ICSI) of dexamethasone and lignocaine hydrochloride were administered biweekly for six weeks. The patient had no adverse reactions. (Figure 2D) The treatment response is detailed in Table 1.

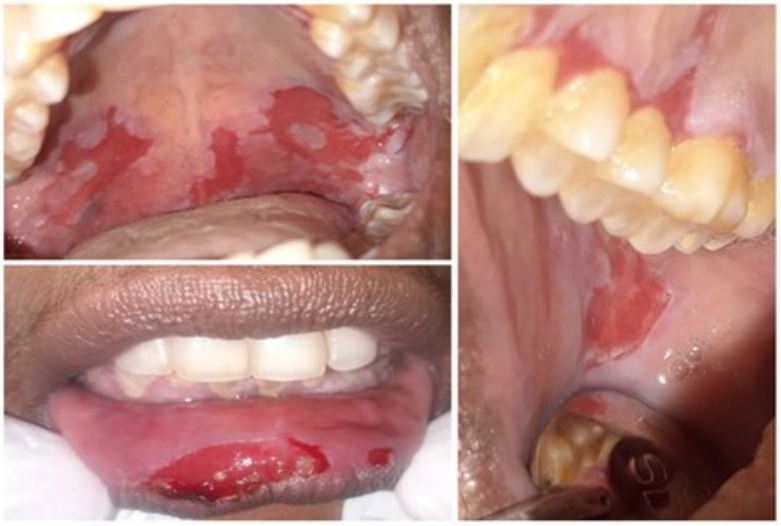

Figure 1: Multiple shallow irregular ulcers at the junction of hard & soft palate, lower lip and right buccal mucosa.

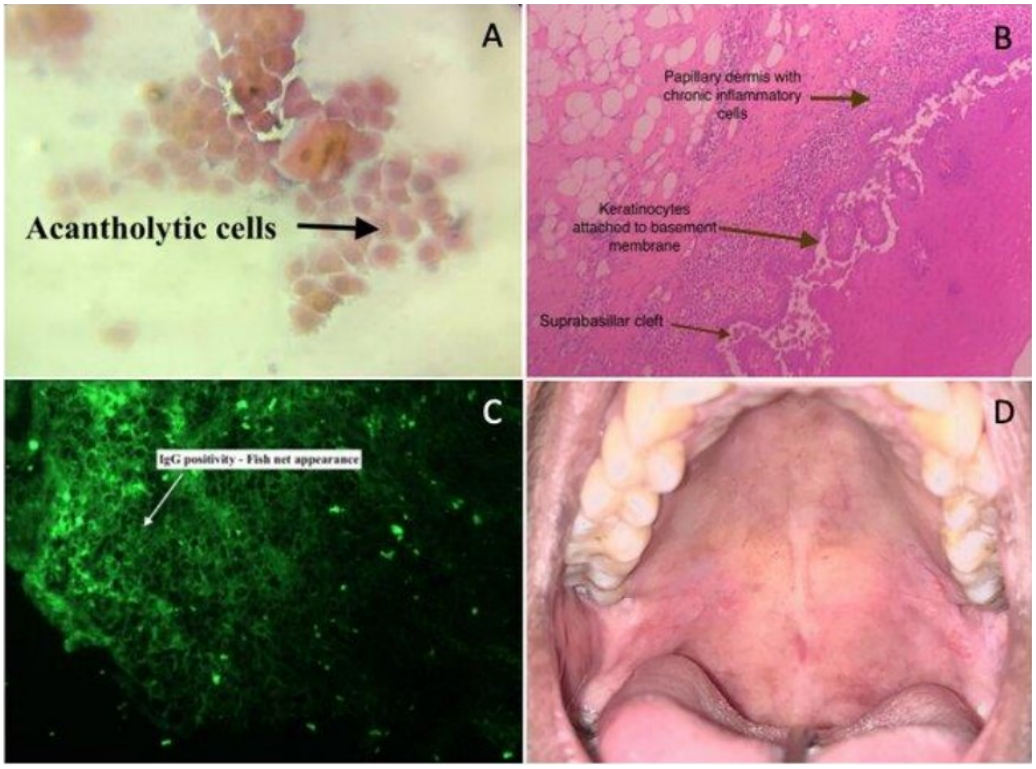

Figure 2A: Tzanck smear shows presence of acantholytic cells.Tzanck smear shows the presence of acantholytic cells; (B) Histopathological image (haematoxylin-eosin staining) reveals supra-basilar bullae along with acantholytic cells; (C) Direct Immunofluorescence evaluation shows IgG positivity with desmosomes giving a fish-net appearance; (D) The patient shows no adverse reaction for ICSI.

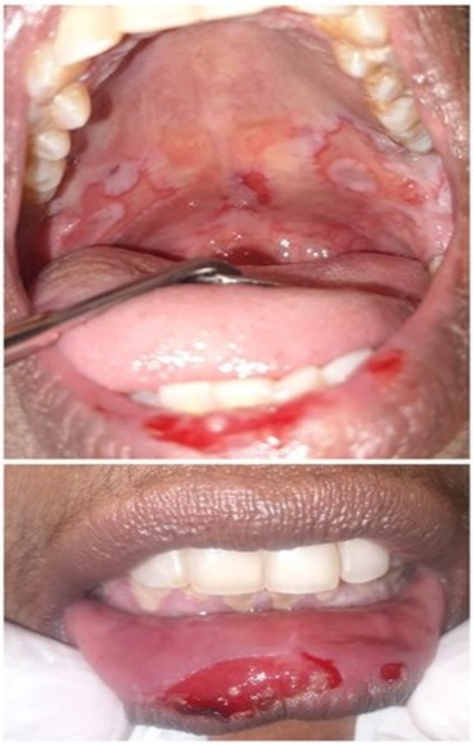

Figure 3: Healing of lesion by 2nd week of ICSI

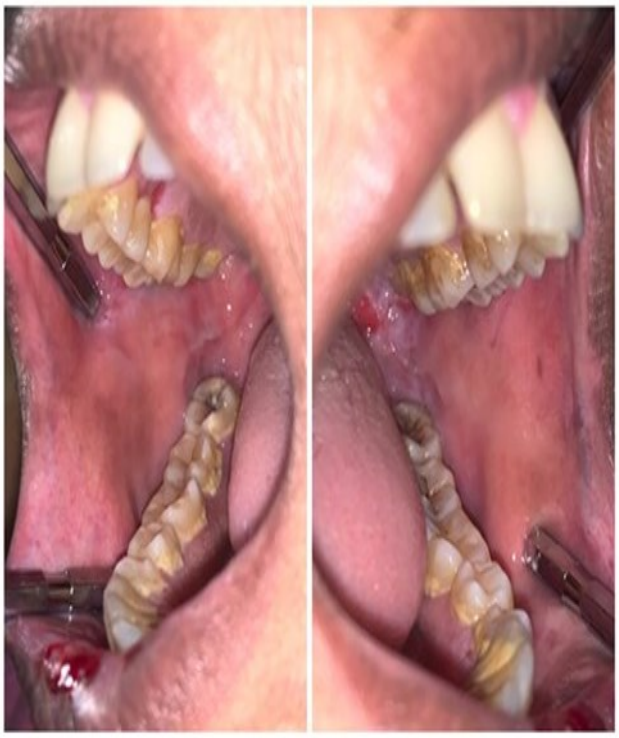

Figure 4: Healing of lesion by 4th week of ICSI

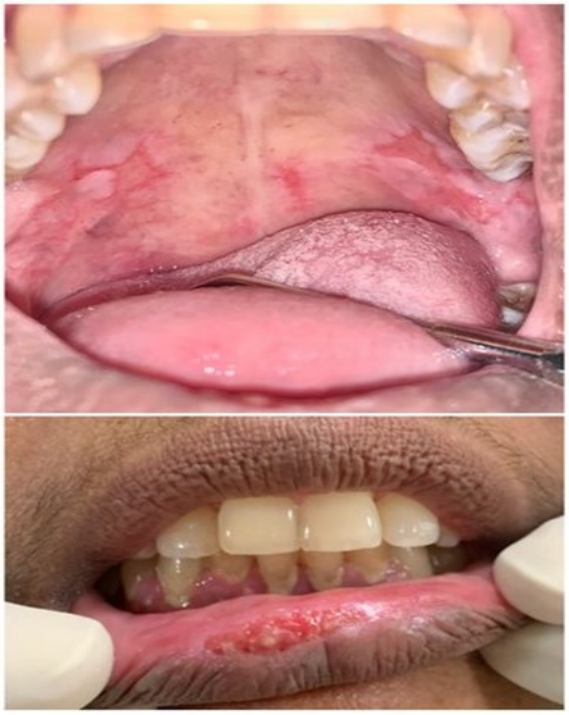

Figure 5: Healing of lesion by 6th week of ICSI

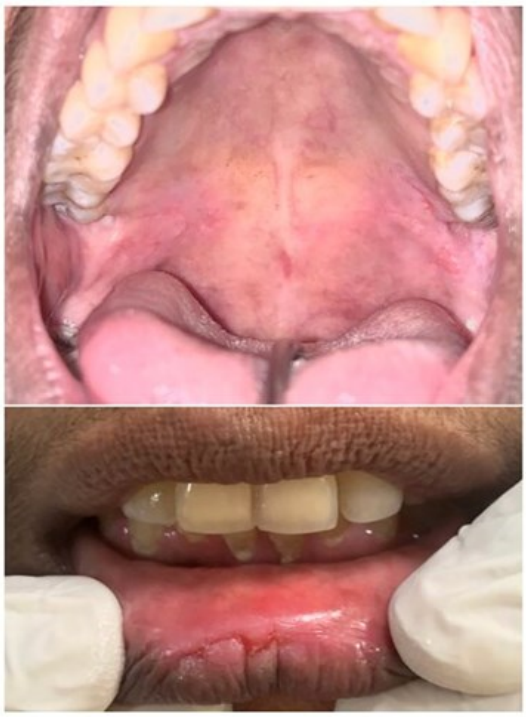

Figure 6: Healing of lesion by 8th & 9th week of ICSI

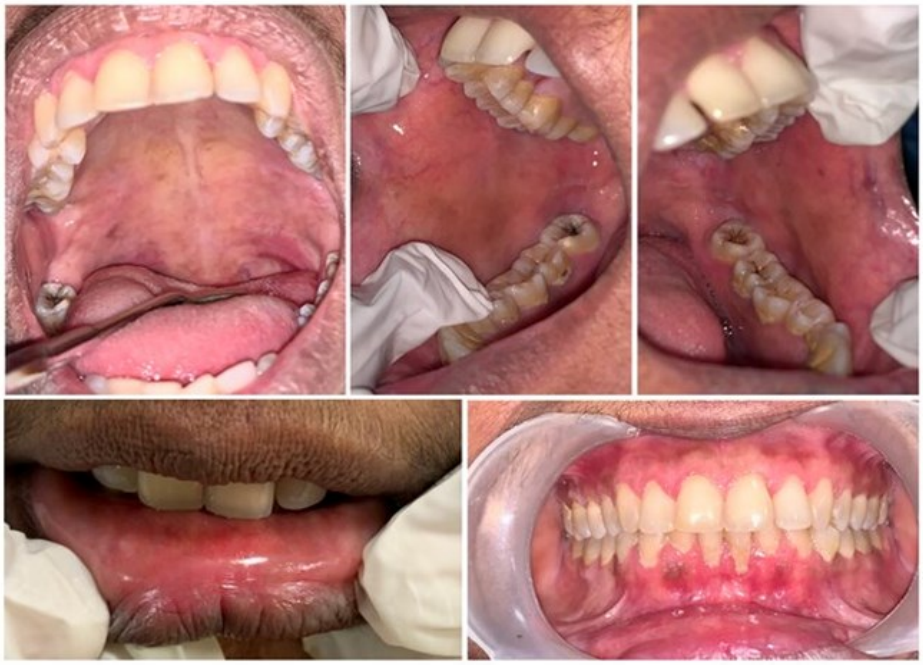

Figure 7: Complete resolution of lesions in the palate and lips by 12th week of ICSI & Patient remained free of oral ulceration without any relapse after 4 reviews

Table 1: Patients response to the ICSI, at Chennai, 2021(March – May).

Treatment	Review report			
	2nd week review	4th week review	6th week review	8th week review
ICSI – Twice weekly Syrup. Dexorange 0 - 1tsp-0 Tab. Zu- C 500mg 0-1 -0 Povidine iodine (Betadine) gargle, twice daily	Appreciable healing of the lesion in right buccal mucosa and gingiva, with symptomatic relief. No response in Lip and palatal lesions. [Figure 3] VAS score - 6.	Lesions on the buccal mucosa healed completely. significant healing of the gingival lesions. [Figure 4] Lip and palatal lesions remind the same. VAS score – 4	Gingival erythema subsided. signs of healing was noted in the lip and palate. [Figure 5] VAS score – 3	Lesions on the palate healed completely with the track-down of erythema. Lip lesion showed remarkable response. [Figure 6] VAS score - 0

Follow-up and Outcome

By the second week of therapy, new lesion formation stopped, and existing lesions began to heal. By the ninth week, 80% of lesions had healed, and no new lesions appeared. Intralesional corticosteroid injections (ICSI) were discontinued, and the patient was prescribed topical Tenovate 0.05% thrice daily. By week 12, significant healing was noted, with complete resolution of palatal lesions and lip lesions healing the following week. [Figure 7] As all symptoms resolved and no new lesions formed, topical therapy was withdrawn. The patient completed four follow-up visits and remained free of oral ulcerations. At a one-year follow-up, there was no relapse, and annual review visits were planned.

Discussion

Oral pemphigus causes chronic ulcers due to bullae formation and rupture [5]. The primary treatment goal is symptom relief using corticosteroids. Initially, the patient received low-dose prednisolone and cyclophosphamide.

Dexamethasone-Cyclophosphamide Pulse (DCP) therapy, known for higher efficacy and rapid healing,

was considered but avoided due to its complications, such as diabetes and hypertension [6]. Instead, ICSI was chosen to deliver high local drug concentrations, accelerating healing and minimizing systemic absorption [7]. This approach reduces mucosal atrophy risk and ensures longer drug retention at the lesion site. Despite the potential side effects like bleeding and mucosal atrophy, the patient experienced none. The total dose was limited to 2 ml per session, and significant improvement was observed by the fourth week, with complete healing by the eighth week. There was no recurrence during the one-year follow-up, but sustained remission for at least five years is necessary to reduce recurrence risk.

Conclusion

Though systemic corticosteroids, with or without immunosuppressive drugs, are the standard treatment for pemphigus, a favorable response to intralesional steroid injections achievied a symptom-free, recurrence-free state one-year post-treatment in the present case.

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