

Anterior scleritis in pemphigus vulgaris: A rare ocular manifestation

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Key words: Anterior scleritis, ocular manifestation, pemphigus vulgaris, rituximab

Pemphigus vulgaris (PV) is a rare severe autoimmune disease that is characterized by painful blisters and erosions on the skin and mucous membranes.^[1,2] It is characterized by the loss of keratinocytes adhesion caused by circulating autoantibodies, mainly circulating IgG antibodies against desmoglein 3 or both desmoglein 1 and 3, the intercellular adhesion molecules. Generally, patients of PV present with oral lesions, which may precede cutaneous lesions by several months.^[1] Ocular manifestations in PV are rare, the commonest reported being conjunctivitis.^[3,4]

We report a rare occurrence of anterior scleritis in a patient with PV on immunosuppressive therapy.

A 30-year-old female patient was referred to a tertiary care center with 7 days history of redness and severe pain in the right eye (RE). She was on a combination of moxifloxacin (0.5%) and ketorolac (0.5%) eye drops four times a day, started elsewhere, with no improvement in her symptoms. She had undergone an uneventful LASIK surgery in both eyes 2 years ago. She also had a history of oral ulcerations 8 months ago, with histological and immunofluorescence studies done at that time, diagnostic of PV. She was started on systemic prednisolone 30 mg/day, T. azathioprine 50 mg/day with topical application of triamcinolone acetonide (0.1%) ointment for local application over buccal mucosal erosions, by the attending dermatologist. As the oral lesions healed, she was weaned off topical and systemic steroids and continued on oral azathioprine for 6 months. She gave a history of recent aggravation of her oral lesions, 3 days after the onset of her ocular symptoms.

On ophthalmological examination, the best-corrected visual acuity in both eyes was 6/6. Slit-lamp examination of RE revealed mucoid discharge, diffuse conjunctival congestion with dilation of deep episcleral vessels with tenderness to

palpation [Fig. 1a and 1b]. The cornea was clear with a quiet anterior chamber. Examination of LE was normal. The posterior segment in both eyes was normal. Dermatological examination revealed multiple sublingual erosions [Fig. 1c] along with erosions on the buccal mucosa and hard palate [Fig. 1d]. The patient was diagnosed with RE diffuse non-necrotizing anterior scleritis secondary to PV. A conjunctival swab was sent for Gram stain and culture and drug sensitivity and was negative. A thorough systemic evaluation including immunological workup was carried out. She was started on topical prednisolone acetate six times a day along with carboxymethyl cellulose eye drops six times a day. Dermatologist and immunologist opinion was sought and intravenous rituximab (first dose of 1g), T. prednisolone (1 mg/kg/day), T. indomethacin 50 mg/day, and topical triamcinolone acetonide (0.1%) ointment for local application on the buccal mucosa was added to her regimen along with her ongoing therapy of oral azathioprine for PV.

It was found that the signs and symptoms of scleritis improved within 1 week [Fig. 1e] along with improvement in her oral mucosal erosions with complete resolution of scleritis observed after 2 weeks [Fig. 1f]. She was administered a second dose of intravenous (IV) rituximab (1g) on the 14th day as per the protocol and was later weaned off the topical and systemic steroids with the continuation of systemic azathioprine therapy.^[5] No recurrence was noted until a follow-up period of 3 months.

Discussion

Ocular involvement in PV can be explained by the presence of desmoglein 3 in the ocular epithelium. The commonest feature is non-cicatricial conjunctivitis and rarely erosions on the eyelids and conjunctiva may be observed. Scleritis is a rarely reported association of PV with only a few case reports describing it during the course of the disease.^[2,6-8] To the best of our knowledge, this is the first case of anterior scleritis in PV reported from western India.

Ocular involvement has been reported as a signal of severe disease and tends to occur several months after the onset of cutaneous and other mucosal lesions as seen in the present case where the anterior scleritis occurred after 8 months after the diagnosis of PV.

Infective anterior scleritis in PV responding to topical and oral anti-microbials has been reported previously.^[6,9] The occurrence of anterior scleritis a few days after the administration of IV rituximab for PV has also been reported.^[2,6,7] Alkeraye *et al.*^[2] reported a good response of ocular and oral lesions to the second dose of rituximab therapy in spite of its occurrence post the first dose. However, recurrence of scleral inflammation has been reported with rituximab with a majority of patients

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Access this article online	
Quick Response Code:	Website: www.ijjo.in
	DOI: 10.4103/ijjo.IJO_3169_21

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Received: 25-Dec-2021

Revision: 01-Mar-2022

Accepted: 22-Mar-2022

Published: 30-Jun-2022

Cite this article as: Bhole PK, Gandhi K, Parmar D. Anterior scleritis in pemphigus vulgaris: A rare ocular manifestation. Indian J Ophthalmol 2022;70:2669-71.

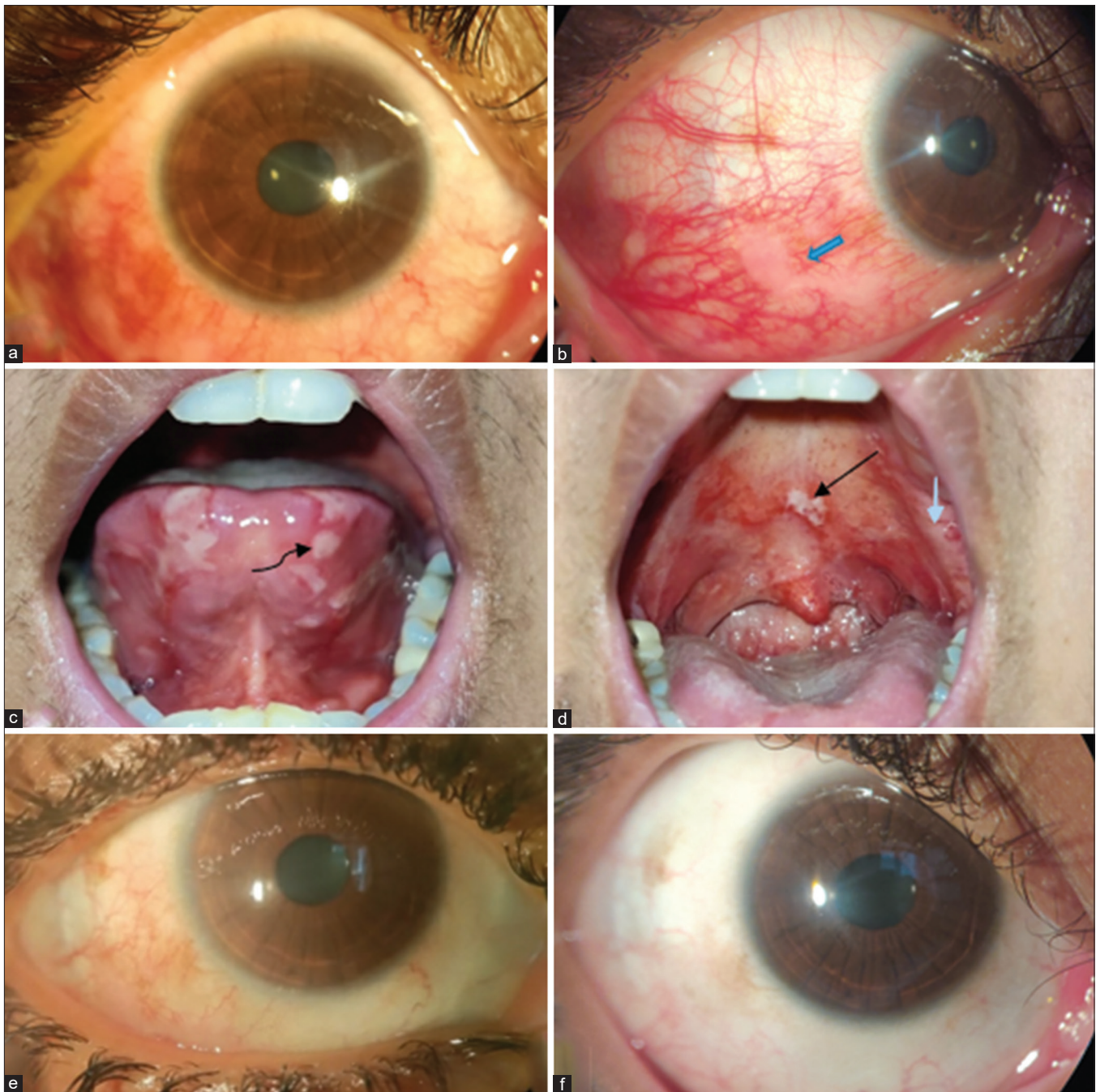


Figure 1: Anterior scleritis in pemphigus vulgaris. (a) Slit-lamp evaluation showing diffuse conjunctival congestion and dilated episcleral vessels. (b) Mucoïd discharge (blue arrow). (c) Oral sublingual erosions (curved black arrow). (d) Hard palate ulceration (black arrow). (e) Resolving scleritis (after 1 week). (f) Resolved scleritis (after 2 weeks)

responding to re-treatment with the same drug.^[10] The present case, however, primarily responded well to IV rituximab therapy along with oral steroids and azathioprine, with the healing of scleritis as well as oral lesions. Hence, rituximab may be considered a second-line agent for non-infectious anterior scleritis refractory to conventional immunosuppression although further studies with long-term follow-up may be required to understand its true potential.

To conclude, anterior scleritis is a rare ocular manifestation of PV and can be a signal of severe disease. Timely identification of its association with PV can help in guiding

the ongoing immunosuppressive therapy, as in the present case a good response was obtained to systemic steroids and rituximab.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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