

Bilateral nodular sclerokeratitis secondary to syphilis - A case report

*Siddhi Goel, Arjun Desai, Pranita Sahay,
Prafulla K Maharana, Namrata Sharma,
Jeevan S Titiyal*

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Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Correspondence to: Dr. Prafulla K Maharana, Department of Ophthalmology, Cornea, Cataract and Refractive Surgery Services, Room Number S-5, First Floor, Dr. Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi, India. E-mail: drpraful13@gmail.com

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A 30-year-old female patient presented with bilateral anterior nodular sclerokeratitis and multiple erythematous skin lesions involving the face, trunk, arms, and legs. The patient had a history of temporary relief with steroids, however the lesions recurred. A dermatology consultation was sought and the patient was diagnosed to have syphilis, consequent to which she was started on benzathine penicillin and showed a dramatic improvement in both skin and ocular lesions. A high index of suspicion for syphilis should be kept in mind for patients presenting with nodular scleritis to initiate timely and appropriate management with penicillin.

Key words: Nodular scleritis, sclerokeratitis, syphilis

Ocular manifestations of syphilis have been rarely reported in literature.^[1-6] Anterior nodular sclerokeratitis can occur in syphilis, and often this can be a presenting symptom.^[3,7,8] The treatment of choice for these cases is systemic benzathine

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penicillin; however, the use of topical steroids shows an initial good response in symptoms without altering the underlying pathology resulting in a prolonged course of the disease as well as consequences of steroid therapy.^[8] Herein, we present a similar case wherein anterior nodular sclerokeratitis was initially managed with steroids for a prolonged period of time without due consideration of the underlying systemic association. On systemic evaluation, the patient was diagnosed with syphilis for which systemic benzathine penicillin was started to which the patient responded well with resolution of ocular nodules.

Case Report

A 30-year-old lady presented with complains of bilateral acute onset pain, redness, and nodular lesions in the eye for the past three weeks. She also had painful erythematous rash and elevated skin lesions over her forehead, forearms, and trunk. She did not give a history of any other medication/drug intake or addictions. Sexual and reproductive history was unremarkable. The patient had been started on topical steroids and lubricant eye drops by her local ophthalmologist, which had caused a mild improvement in her ocular symptoms.

On ocular examination, she had a best-corrected distance visual acuity (BCVA) of 6/6 in both the eyes. There was diffuse conjunctival injection in both eyes with a nebulomacular corneal opacity (NMCO) in the inferior quadrant of right eye measuring 1 × 1 mm and two elevated scleral nodules on the ocular surface (inferior and temporal to the cornea). In the left

eye, a small temporal NMCO with corneal infiltrate and an elevated temporal scleral nodule were seen [Fig. 1]. The nodule was positive on sodium fluorescein staining. Posterior segment evaluation was normal.

General examination revealed multiple erythematous, tender indurated papules to plaques distributed over forehead, extensor surface of both the forearms, feet, and the lower back [Fig. 2]. Scaling was seen overlying some lesions while there was central atrophy in the larger lesions. None of the skin lesions were hypopigmented and there was no hypoesthesia over the lesion. Sensory examination revealed reduced sensation to fine touch and pain over both upper limbs below elbow and over the palmar

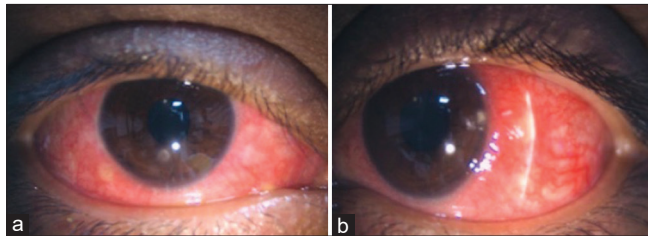


Figure 1: Slit-lamp image of both eyes at baseline (a) right eye showing diffuse congestion with nebulomacular corneal opacity on inferior cornea, and inferotemporal scleral nodule (b) left eye showing temporal corneal infiltrates with temporal scleral nodule

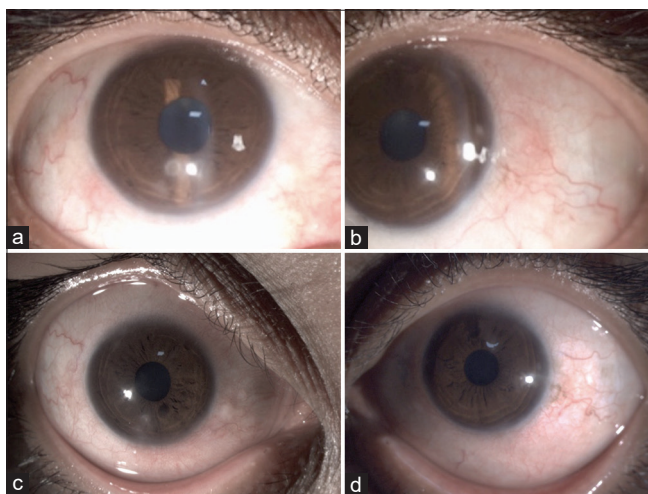


Figure 3: Slit-lamp image of both eyes (a and b) after 1st dose of penicillin and after 1 month of treatment (c and d) showing marked decrease in inflammation and size of the nodules

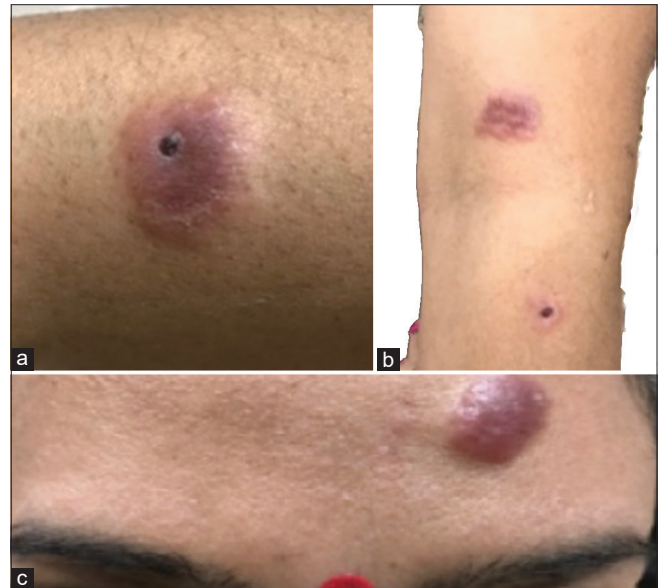


Figure 2: Clinical photograph of skin lesions at baseline showing (a) erythematous indurated papule with central atrophy on the right forearm (b) left forearm, and (c) forehead



Figure 4: Clinical photograph of skin lesions after 1st dose of penicillin showing decreased erythema and flattening of lesions on (a) right forearm, (b) left forearm, and (c) forehead

Table 1: Review of literature for cases of scleritis in syphilis with negative HIV serology

Author (Year)	Type of Study	n	Stage of syphilis	Treatment	Outcome
Casey <i>et al.</i> (1996)	Case report	1	Secondary syphilis	Parenteral Benzathine Penicillin (intramuscular)	Complete resolution in 2 weeks
Wilhelmus <i>et al.</i> ^[5] (1987)	Case series	2	-	Parenteral Benzathine Penicillin-1; Parenteral Aqueous Penicillin-1	Complete resolution in 1-3 weeks
Shaikh <i>et al.</i> (2015)	Case report	1	-	Parenteral Benzathine Penicillin (intramuscular)	Good response at 1 month
Ahmadreza Moradi <i>et al.</i> (2015)	Case series	4	-	-	-
Furtado <i>et al.</i> (2018)	Case series	3	-	-	-

and dorsal surface of the hand. The left ulnar and radial cutaneous nerve were found to be firm and thickened on palpation.

Baseline hematological investigations — complete blood count, renal function test, liver function tests, and erythrocyte sedimentation rate (ESR) were found to be normal. Investigations for cytoplasmic antineutrophil cytoplasmic antibody (c-ANCA), perinuclear antineutrophil cytoplasmic antibodies (p-ANCA), single-stranded DNA (ss-DNA), and rheumatoid factor were negative. Test for human immunodeficiency virus 1 (HIV-1) antibody, HIV-2 antibody, hepatitis B surface antigen (HbSAg), and antihepatitis C virus antibody were nonreactive. The Venereal Disease Research Laboratory (VDRL) was reactive with a titer of 1:64. The *Treponema pallidum* hemagglutination assay (TPHA) was positive. Serological tests for these infectious markers were negative in the patient's spouse.

Mantoux test resulted in a 70 mm induration. Chest X-ray was normal. Quantiferon tuberculosis (TB) gold test and high-resolution computed tomography of chest were normal.

Skin biopsy was taken from one lesion each from both the forearms that revealed focal parakeratosis and mild atrophy of epidermis. Microscopic examination revealed presence of a diffuse, dense, perivascular, and focal perieccrine lymphohistiocytic infiltrate extending beyond the mid dermis. There was no evidence of granulation tissue or acid-fast bacilli in the examined specimen.

Based on these findings a diagnosis of anterior nodular sclerokeratitis with syphilis was made.

The patient was started on 0.5% preservative-free moxifloxacin eye drops thrice a day, 0.5% carboxy methyl cellulose eye drops every 2 h, and 1% prednisolone phosphate eye drops 4 times/day with 75 mg oral indomethacin once a day for ocular symptoms. Intramuscular injection of benzathine penicillin 2.4 million International Units (4 mL) was given in the buttock (2 mL in each buttock). A total of three injections were given at weekly intervals. There was a marked decrease in ocular symptoms after initiating therapy with near total resolution of ocular pain and congestion at 2 weeks [Fig. 3]. Flattening of skin lesions with a 30%–40% decrease in size of the lesions was noted one-month post penicillin injection [Fig. 4]. The VDRL titer was repeated at 3 months, which showed a decrease in titer to 1:8.

Discussion

Anterior nodular sclerokeratitis can occur as an idiopathic entity or may be associated with systemic disease in approximately 39%–50% of patients.^[5] While autoimmune and connective tissue

disorders like rheumatoid arthritis, Wegener's granulomatosis, systemic lupus erythematosus, relapsing polychondritis, and polyarteritis nodosa are the more common associations, infectious scleritis has been reported in 4.2% to 7.5% of the cases.^[9]

Ocular syphilis is known as the "great imitator" and a diverse range of ocular manifestations have been reported: keratitis, iritis, posterior uveitis, pan uveitis, chorioretinitis, optic neuritis, and scleritis.^[1,2,4,6,10,11] Ocular involvement is relatively less common in primary and secondary syphilis and is an uncommon clinical entity in an immuno-competent host.^[11] However, few cases of scleritis have been reported in medical literature in patients with secondary syphilis and negative serology for HIV^[2,4,7,8] [Table 1]. Wilhelmus *et al.* reported complete clinical resolution in all cases of syphilitic episcleritis (n=2) and scleritis (n=2) following parenteral therapy with penicillin.^[8] Similarly, Shaikh *et al.* reported a case of nodular syphilitic scleritis masquerading as an ocular tumor, which responded well to penicillin.^[9]

Thus, based on the clinical presentation of the patient, a positive VDRL titre of 1:64, positive *Treponema pallidum* hemagglutination (TPHA), and dramatic improvement of both ocular and skin lesions after intramuscular administration of benzathine penicillin, we concluded that the patient was affected with anterior nodular sclerokeratitis secondary to syphilis, the source of which could not be traced. It is important to note that the patient had highly positive Mantoux test, 70 mm in this case. However, such a result should not distract a clinician from simultaneously ordering for other laboratory investigations such as TPHA for syphilis, which although is uncommon but is a great mimicker.

Conclusion

All cases of scleritis, especially those with nonophthalmic features and not responding to standard treatment protocol should be carefully evaluated for rare causes and systemic associations preferably by an integrated team of healthcare professionals. Laboratory tests for syphilis should be routinely carried out in all patients presenting with scleritis without an underlying obvious cause for optimal outcomes.

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.

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Conflicts of interest

There are no conflicts of interest.

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