CASE REPORT

Sclerokeratouveitis and lens dislocation in a patient with genital ulcer: was the great imitator imitated?

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Accepted 15 September 2017

SUMMARY

Uveitis is the most common ocular manifestation of syphilis, while scleritis is rare. A case of nodular scleritis, peripheral keratitis and uveitis in a patient with genital ulcer is presented in this report. This patient had a clinical profile suggestive of syphilis, though the serology was negative for treponemal antibodies. Other possible differential diagnosis imitating syphilis are discussed.

BACKGROUND

Syphilis is an infectious disease caused by *Treponema pallidum* and has been labelled as the great imitator due to its varied presentations.¹ Ocular syphilis usually manifests as uveitis and is usually seen in the latent phase of the infection, while scleritis is a rare presentation.^{1–5} The diagnosis is based on positive serology or direct demonstration of the spirochetes in human tissue.^{1,6} It is important to rule out the disease as early as possible as about a third of patients with latent infection eventually develop the potentially fatal form of tertiary syphilis. In fact serological testing has been suggested for every undiagnosed case of ocular inflammation.¹

We present a case of successfully managed sclerokeratouveitis that had clinically definitive features of systemic and ocular syphilis though laboratory diagnosis proved otherwise. We discuss the differential diagnosis and the possible imitator of the great imitator itself!

CASE PRESENTATION

A 45-year-old chronic alcoholic male was brought to our emergency services by a social worker with complaints of pain, redness and photophobia in both eyes (BE) since 5 days. There was history of multiple episodes of acute red eye treated with topical drugs. The patient denied history of systemic disease. Visual acuity was 5/60 in right eye (RE) and improved with correction to 6/12, while it was best corrected at hand motions close to face in left eye (LE). BE had anterior chamber cells (1+ in RE and 3+ in LE, standard uveitis nomenclature) fine keratic precipitates, a pupillary membrane, posterior synechiae and aphakia on slit lamp examination (figure 1). LE additionally had a perforated peripheral corneal ulcer measuring 5.5 mm in longest dimension with prolapsed iris tissue beneath pseudocornea along with multiple mildly tender scleral nodules (roughly 2×2 mm) in the temporal and inferior-temporal area. A sector of necrotising scleritis was also present adjacent to

the perforated cornea in LE, while RE had a small isolated scleral nodule (figure 1). Ultrasound B scans of BE showed posterior dislocation of crystalline lens (figure 2). On leading questions, patient gave history of multiple sexual partners and history of high risk exposure in the last 15–20 years, the last contact being 20 days back. A solitary indurated painless genital ulcer measuring 1.3 cm in longest dimension was detected on the scrotum during systemic examination (figure 3).

A presumptive diagnosis of BE infective nodular scleritis, anterior uveitis and aphakia, with LE perforated peripheral ulcerative keratitis and necrotising scleritis was made.

INVESTIGATIONS

Gram stain and potassium hydroxide preparation preparation of corneal scraping of LE were negative as were the bacterial and fungal culture reports obtained in due course. He was diagnosed to have syphilitic chancre by the dermatologist but serology reports obtained after 3 days were negative for viral markers (HIV, herpes simplex virus (HSV), hepatitis C virus and hepatitis B virus), while venereal disease research laboratory (VDRL) and fluorescent treponemal antibody absorption (FTA-ABS) were also negative. Chest X-ray, Mantoux test and immune markers (antinuclear antibody, antineutrophil cytoplasmic antibody and rheumatoid factor) were also negative.

TREATMENT

The patient was advised BE topical moxifloxacin (0.3%) two hourly, cycloplegia, oral vitamin C and oral doxycycline.

OUTCOME AND FOLLOW-UP

While the patient was being investigated, there was a dramatic improvement in BE within 48 hours. By 10th day of follow-up, corrected visual acuity had improved to 6/9 in RE and 6/60 in LE. There were no congestion and no anterior chamber cells in BE, BE scleral nodules resolved while LE perforated corneal ulcer had healed further with formation of a vascularised corneoid scar (figure 4). Even the genital ulcer showed marked improvement with no induration and significant reduction in size (figure 3). The condition remained stable at 1 month of follow-up.

DISCUSSION

Sclerouveitis can be due to infective causes like tuberculosis, syphilis, Lyme's disease, HSV, leprosy or due to inflammatory causes like systemic lupus

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To cite: Rathj A, Takkar B, Sharma N. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-221668

Figure 1 Clinical anterior segment photographs of (A) right eye and (B) left eye. Both eyes had aphakia with pupillary membrane, posterior synechiae and nodular scleritis (stars). Additionally, left eye had a perforated peripheral corneal ulcer with prolapsed iris tissue and pseudocornea (arrow).

erythematosus, sarcoidosis, granulomatosis with polyangitis and Behcet disease.³⁻⁹ Differentiating infectious causes from inflammatory is paramount for prevention of untoward complications. We diagnosed the patient to have an infectious disease due to his profile and possible exposure to a venereal disease and due to rapidly progressive scleral and corneal necrosis in his LE. The rationale for starting doxycycline was prevention of further corneal melting, promotion of corneal healing and provision of a broad spectrum anti-infective cover. The quick response of the ocular condition to antibiotics without steroids within 48 hours indicates that disease was indeed infectious in aetiology.¹

Ocular signs of bilateral scleritis, lens dislocation, iris nodules and uveitis are known to be among the classical signs of syphilitic anterior uveitis.¹ The solitary painless genital ulcer was diagnosed to be syphilitic chancre by the dermatologist, which is the diagnostic sign of primary stage of syphilis and may rarely also be seen in secondary syphilis.^{1,10} However, ocular signs like lens dislocation and history of episodes of acute red eye indicate

latent or tertiary stage of syphilis, which are the most common stages of ocular involvement by the organism.¹ Further, we could not prove the disease serologically and dark field microscopy was not available to us.⁶ Therefore, we decided against penicillin therapy and continued doxycycline, which is a known alternative therapy for syphilis.¹¹ Every case of ocular syphilis should be evaluated for possibility of neurosyphilis by cerebrospinal fluid analysis,¹ but our patient did not give consent for the invasive procedure due to the doubtful diagnosis. One may explain the presence of chancre in latent/tertiary phase by re-exposure of a patient with syphilis to the organism, but laboratory evidence points otherwise.⁶ Thus, we concluded in retrospect that another disease may have imitated syphilis in this patient. A biopsy, scleral-corneal or from the scrotal ulcer, should be considered in such suspicious cases. However, they were not performed in our case as both the ocular and genital lesions showed a rapid response to therapy, even before the blood could be analysed for the presence of antibodies.

Figure 2 Ultrasound B scan of (A) right eye and (B) left eye showing posteriorly dislocated lens. Note the high spikes on vector scan corresponding to the lenses.

Figure 3 (A) Clinical photograph of the scrotum of the patient showing solitary indurated genital ulcer that measured to 1.3 cm in longest dimension. (B) Clinical photograph of the lesion following treatment for 2 weeks showing the healed ulcer.

Lyme disease or Borreliosis is the other probable imitator as scleritis is reported in it and it may also cause genital lesions when disseminated.^{9 12} Of note, Borreliosis is also caused by a spirochete which may explain the response to therapy. However, the absence of erythema migrans, tick bite and 'late manifestations' of the disease virtually rule out a confirmed/probable diagnosis.¹³ The genital ulcer caused by HSV is known to have multiple vesicles and is painful and was ruled out on serology. Inflammatory causes like Behcet disease and MAGIC syndrome

may also be considered in a case of genital ulcer with sclerouveitis, but the resolution without steroids is unlikely.¹⁴ In latent syphilis, VDRL test can be falsely negative in 30% of cases while FTA-ABS test is false negative very rarely, in 1%–2% of cases.¹ Therefore serology negative syphilis is a distant possibility in view of clinical presentation. Although immunodeficiency may be responsible for the negative FTA-ABS test, the systemic history and blood investigations of our patient were not indicative of such a state. Bilateral peripheral keratitis in the setting

Figure 4 Clinical photographs of the anterior segment of (A) right eye and (B) left eye showing resolution of scleritis and the scleral nodules. The corneal ulcer of the left eye also healed with the formation of a scar (arrow). Minimal dilation and congestion of the vessels can be seen at the site of the scleral nodule (star).

of chronic alcohol abuse raises a distant possibility of distinct corneal infections related to general debilitation and immune dysfunction.^{15 16} But we could not attain any evidence of local infection on microscopic examination or microbial culture of corneal scraping. Trauma and neurotrophic ulcers can also be considered as the cause of bilateral peripheral corneal ulcers in patients with such a profile, but the absence of history of trauma and the presence of scleral nodules and the scrotal ulcer indicated otherwise.

Learning points

- ▶ Ruling out infectious causes of sclerouveitis is pivotal to its successful management.
- ▶ Despite best efforts, confirmatory laboratory diagnosis may remain elusive and relying on clinical signs in such cases is recommended.

Contributors All authors equally contributed to the workup diagnosis and management. AR did the imaging. BT wrote the manuscript and NS did critical revision of the script. AR holds the overall responsibility.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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