Clinical Image

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Woman with a 3-month history of left ulcerative bulbar conjunctiva accompanied by severe scleral irritation and soreness

**Clinical Presentation**

In practice, symptoms of severe scleral irritation and soreness is often followed by scleral hyperemia, edema, episcleral angiogenesis obliterans, scleral lesions with nearby pale avascular zones, and scleral necrosis, loss or perforation, involving tissues such as the cornea, ciliary body, and trabecular meshwork. Here, we present a case of mixed bacterial (*Enterococcus faecalis* [*E. faecalis*]) and fungal infection of the sclera of the eye as a clinical quiz. Our goal is to highlight the clinical features, etiology, and management of a rare case of left ulcerative bulbar conjunctiva accompanied by severe scleral irritation and soreness.

A 49-year-old female presented to our clinic with a 3-month history of a red and swollen left eye. Her past medical history was notable for treatment at another hospital with tobramycin-dexamethasone and pranoprofen eye drops for almost 2 months since symptom onset. Slit-lamp examination showed mixed conjunctival congestion, with 2 conjunctival ulcers at the 3- and 6-o’clock positions, situated on the bulbar surface, 5 and 2 mm from the corneal limbus (Figure 1A & 1B). A small volume of purulent discharge was adherent to the ulcers. The corneal limbus was thin (1 mm wide) from the 3:30- to 5- o’clock position. The cornea was transparent at other places.

An exploratory operation of the left conjunctiva was performed with the bulbous conjunctiva being resected along the corneal limbus. After the subconjunctival tissue was separated, a large field of necrotic, dissolved, and caducous sclera was identified over the superior, medial, and inferior rectus. The sclera was melted, and the choroid could be seen. A pulmonary CT scan showed localized pulmonary emphysema and tubercle inflammation. Subsequent rheumatologic examinations were consistent with rheumatoid arthritis (RA).

**Figure 1** - An image showing: A) conjunctival ulcer at the 3-o’clock position measuring 2.5 × 1.5 mm; and B) conjunctival ulcer at the 6-o’clock position measuring 0.5 mm

**Figure 2** - A large number of fungal hyphae visible under microscopic examination (arrows).
**Clinical Image**

**Questions**

1. What is the diagnosis?
2. What is the etiology of this condition?
3. What is the management?

**Answers**

1. The woman has necrotizing scleritis and a conjunctival ulcer, combined with bacterial (E. faecalis) and fungal infection.
2. Necrotizing scleritis may be a presenting symptom of systemic acquired connective tissue disorders and vasculitic diseases. Approximately 50-80% of patients with necrotizing scleritis report these conditions, particularly systemic vascular autoimmune disorders, and collagen metabolic diseases. Identifying such conditions can help in the early diagnosis of necrotizing scleritis. The most common type of rheumatoid disease is RA (18-33%), followed by Wegener's granulomatosis systemic vasculitis (7-19%). In this case, the patient was diagnosed with necrotic nodular vasculitis by skin biopsies of both lower limbs. She had suspicious lung changes on CT and rheumatologic examinations that were consistent with RA. Therefore, necrotizing scleritis would be a likely diagnosis. When the necrotic tissue and pus were examined with potassium hydroxide (KOH) under microscopy, copious amounts of fungal hyphae were seen. Bacterial culture and sensitivity tests of the secretions grew large amounts of E. faecalis. The cause of the mixed infection is currently thought to be the patient’s past history of autoimmune disease, hepatitis B infection, and hormonal therapy.
3. Glucocorticoid and immunosuppressant treatments can often have positive effects. However, in this case, drug treatment was not possible because of the mixed infection, and the severity of the scleral lesions. We carried out surgical resection and allogeneic corneal transplantation successively after the failed sclera transplantation. There were no further symptoms or episodes of recurrence after treatment.

**Discussion**

Scleritis is relatively common among 30-50-year-old females. Approximately 14% of scleritis cases become necrotizing, which is hugely destructive for the eye structure and results in vision loss in approximately 40% of affected patients. Few cases of necrotizing scleritis combined with conjunctival ulcer have been reported in the literature. Reddy described the atypical presentation of fungal infection and necrotizing scleritis, but mixed bacterial and fungal infections are highly unusual. This is a case of scleritis due to an E. faecalis infection.

Recent treatment options for necrotizing scleritis include the biological agent rituximab, a chimeric monoclonal antibody directed against the B-lymphocyte protein CD20. Combination therapy with rituximab and glucocorticosteroids is a safe and effective alternative treatment for vasculitis. In the present case, surgical resection and allogeneic scleral transplantation were considered. We selected a cryopreserved homogeneous allogeneic corneal tissue graft to repair the scleral coloboma. This vascularized graft had regularly arranged collagen fibers in the substantia propria layer. It was in a dehydrated state, which can reduce the chance of local immune rejection.
Clinical Image

Allogeneic corneal transplantation therapy for the repair of large scleritic lesions has been shown to be an effective method of saving the eye.

In conclusion, exploratory surgery of the conjunctiva is essential to perform in cases of conjunctival ulceration where necrotizing scleritis is suspected. Necrotic tissues should be thoroughly resected, and the etiology determined intra-operatively to guide intra- and post-operative treatment. Use of an allogeneic tissue graft could preserve both the eyeball and the vision.

References


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