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Case report

Diagnostic dilemma: Unilateral panuveitis mimicking endophthalmitis in very severe HLA B27-associated uveitis

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ABSTRACT

Purpose: To report a case of a 28-years-old male who presented with a worsening unilateral panuveitis after intensive topical steroid therapy which resulted in a diagnostic and treatment dilemma as to whether the patient should be treated as for infective endophthalmitis despite no immediate known infective risk factors.

Observations: A patient presented initially with unilateral non-granulomatous acute anterior uveitis which worsened after being started on intensive steroid therapy, developing fibrinous panuveitis. The rapid worsening of inflammation and vision deterioration despite being on intensive steroid therapy resulted in the patient subsequently being treated as for infective endophthalmitis. Anterior chamber and vitreous tap were done and intravitreal antibiotics were administered, along with topical antibiotics therapy. Vitrectomy was withheld due to the lack of conclusive evidence of infective etiology and risk factors. Full uveitis and infective workup were done. Investigations were largely unremarkable, and fluid and vitreous cultures were negative. HLA B27 was positive. The patient was subsequently started on systemic oral corticosteroids and improved in terms of his visual acuity, signs and symptoms.

Conclusions: HLA B27-associated uveitis can present in a range of clinical spectrum with the extreme being of unusual severity in the form of fibrinous panuveitis that can mimic infective endophthalmitis. This can lead to a dilemma in management and subject patients to unnecessary risks from diagnostic and therapeutic interventions. Patients under this subgroup may require long term systemic immunosuppression therapy for disease remission and will require long term follow up.

1. Introduction

HLA B27-associated uveitis is typically characterised by recurrent acute alternating bilateral non-granulomatous anterior uveitis (with full remission between attacks) in young adults between the ages of 20 and 40, with a preponderance towards males.[1] The inflammation is frequently associated with significant cellular and protein extravasation into the aqueous humor (including fibrin and hypopyon in the anterior chamber) and is characterised by the absence of mutton-fat keratic precipitates.[2] This condition has a frequent association with seronegative arthritic syndromes, of which the most prevalent association is ankylosing spondylitis.[3] The prevalence of HLA B27 is known to differ amongst racial groups, higher in the Caucasian population (8–10%) as compared to the Asian population (1–6%).[2,4]

Although acute anterior uveitis is the typical manifestation of HLA B27-associated uveitis, posterior segment involvement such as vitritis, cystoid macula edema, papillitis and retinal vasculitis can occur in up to 25% of the patients.[5–7]

In this report, we describe the unusual presentation of an unilateral HLA B27-associated uveitis. This patient developed worsening inflammation and subsequently panuveitis despite being on intensive topical steroids from the very start. The rapid deterioration of his condition on steroid therapy posed a diagnostic and treatment dilemma due to the differential diagnosis of infective endophthalmitis, with the patient's demographic and nature of work in mind.

2. Case report

A 28-years-old Indian construction worker, with no significant past medical or ocular history, presented with 4 days history of right eye redness, pain and photophobia. There was no foreign body history nor history of trauma. Systemic, travel and contact history were unremarkable. His visual acuity (VA) was 20/20 in both eyes and slit-lamp examination revealed right eye circumcilliary injection and anterior chamber activity of 2 + cells with no fibrin nor hypopyon. Fine keratic precipitates were seen. There were no vitreous cells seen and posterior

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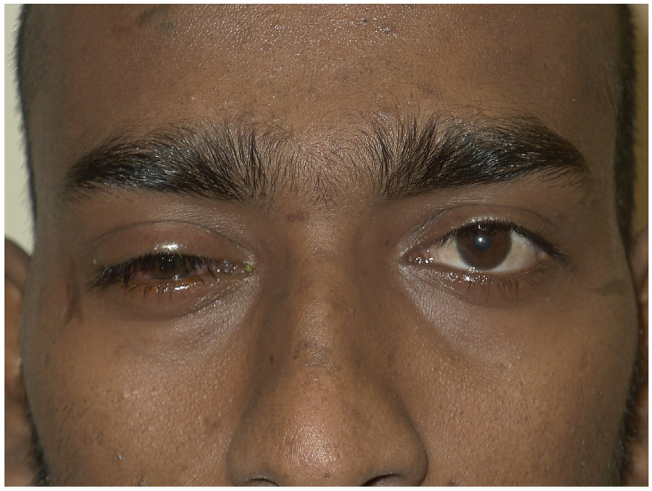

Fig. 1a. Worsening of right eye inflammation with ptosis from contiguous lacrimal gland inflammation after use of intensive topical steroid therapy.

segment examination was normal. There was no relative afferent pupillary defect seen. The diagnosis was that of a right eye non-granulomatous acute anterior uveitis (AAU). No further investigations were done as this was a first episode of AAU for the patient. He was commenced on hourly Guttae Pred Forte and 1% Atropine twice a day.

The patient returned 5 days later with worsening right eye pain and redness. His VA had worsened to 20/80 and examination revealed a right upper eyelid ptosis (Fig. 1a), along with development of right fibrinous anterior uveitis (Fig. 1b), vitritis (Fig. 1c) and cystoid macular edema. With the worsening of signs and symptoms whilst the patient was on intensive steroid therapy, along with the patient's occupation in construction work, the possibility of a foreign body entry resulting in infective endophthalmitis could not be ruled out, despite the negative history.

Anterior chamber paracentesis was performed with the fluid sent for Gram stain testing and Tetraplex Polymerase Chain Reaction (PCR) analysis. Vitreous paracentesis was also performed with the vitreous sample sent for bacterial and fungal cultures. Intravitreal antibiotics were administered and intensive topical antibiotics were commenced immediately. Oral Ciprofloxacin was also started. Computed Tomography of the Orbits done did not show any intraocular foreign body, but noted right lacrimal gland inflammation, explaining the ptosis.

Both the fluid and vitreous samples did not yield any positive

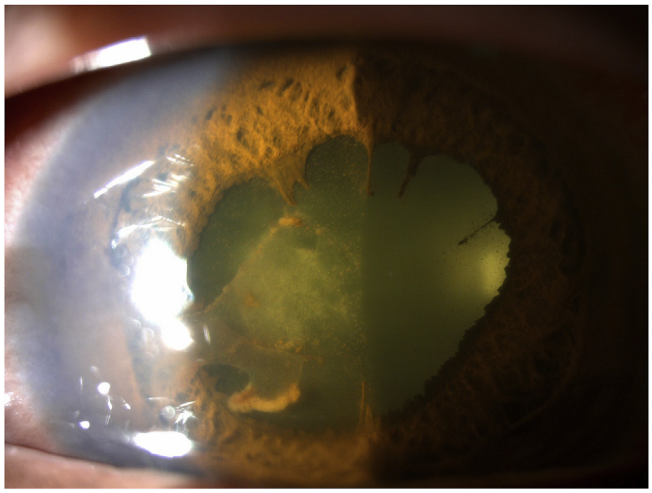

Fig. 1b. Fibrinous anterior uveitis.

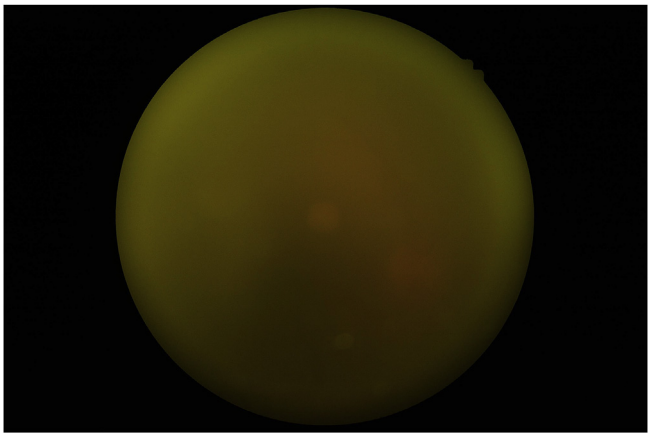

Fig. 1c. Presence of vitritis.

cultures and PCR analysis was negative. Uveitis screen including full blood count, urine analysis and cultures, chest radiograph, syphilis serologies, retroviral screen, toxoplasma screen and tuberculosis T-SPOT were normal. Autoimmune markers such as ANCA, Anti Ro/La, dsDNA and ANA were negative. Despite the intensive antibiotics treatment, the patient's VA deteriorated further to hand movement vision.

The patient eventually proved to be HLA B27 positive and was then started on high dose oral corticosteroids. Lumbo-sacral radiograph did not show signs of ankylosing spondylitis. On review one week later, the patient's right eye VA had improved to 20/30 with improvement in his uveitis (Fig. 2a and b). He was monitored at close intervals and was given a course of gentle tapering dose of topical and oral steroids.

3. Discussion

HLA B27-associated acute anterior uveitis typically has sufficiently characteristic symptoms and signs to make the diagnosis without laboratory confirmation.[8] There can be a wide range of severity but most attacks require only topical therapy and will subside within a few weeks with generally good prognosis.[9] While vitritis and posterior segment involvement in HLA B27-associated uveitis are not uncommon,[1,7,10] several factors combined in making the diagnosis and treatment decisions challenging for this patient.

Firstly, this was an unusually severe first presentation of uveitis for a patient who had no previous attacks and whom displayed no

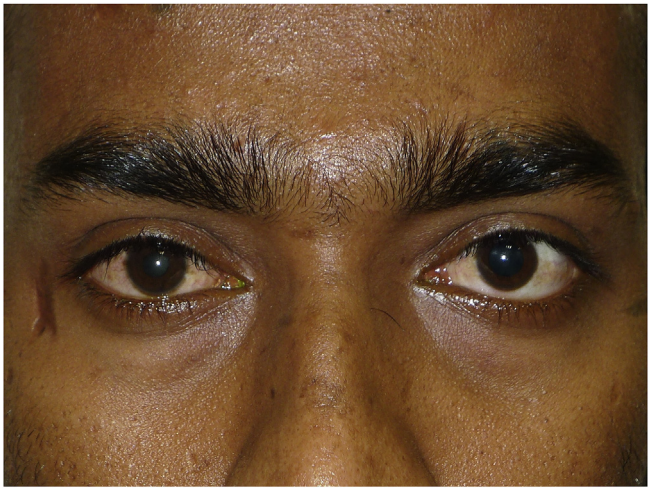

Fig. 2a. Improvement of signs and symptoms after use of topical and systemic corticosteroids with resolution of lacrimal gland inflammation and ptosis.

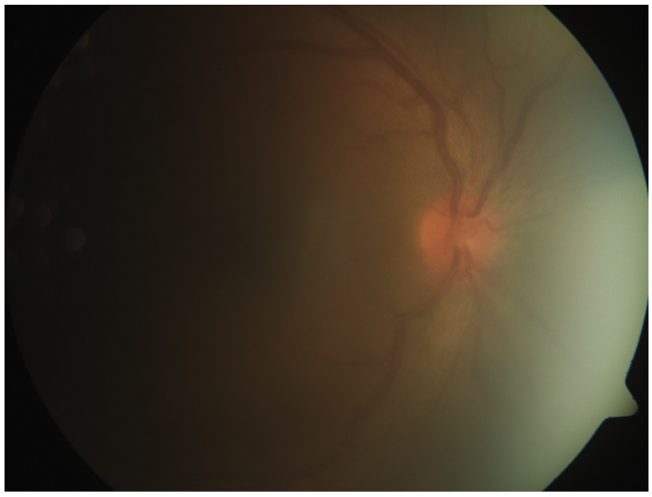

Fig. 2b. Resolving uveitis.

symptoms of any systemic infections or autoimmune diseases. Secondly, this patient's occupation as a construction worker predisposed him to higher risks of ocular trauma, which could have inadvertently resulted in an intraocular foreign body. Thirdly, the rapid development of panuveitis happened after the patient was started on intensive topical steroid therapy, increasing the suspicion of a possible infective cause. In retrospect, while we do know that posterior segment involvement in HLA B27-associated uveitis require systemic steroids for treatment,[7,10] the above factors made it difficult for a straightforward diagnosis.

For this patient, 'tap and inject' was done and thereafter repeated due to the uncertain diagnosis and suspicion of infective endophthalmitis. While the inflammation was rapidly progressive, decision was made to delay surgical intervention due to the fact that there was also no conclusive evidence for any possible causes of exogenous or endogenous endophthalmitis. This uncertainty in diagnosis meant that the patient had to undergo invasive diagnostic and therapeutic procedures with their inherent risks and systemic steroid treatment had to be delayed until an infection could be conclusively ruled out. A review by Kim et al.[7] found that posterior segment involvement in HLA B27-associated uveitis had poorer visual outcomes compared to anterior uveitis despite systemic immunosuppression, and this delay in treatment in our patient could lead to poorer visual outcomes in the long term.

A case series by Sanghvi et al.[10] described 4 cases of similar presentations of HLA B27-associated panuveitis, of which 2 patients underwent diagnostic and therapeutic vitrectomy due to suspicions of infective endophthalmitis. These patients were not known HLA B27-positive on presentation and had no risk factors for infection but presented with severe fibrinous uveitis with rapidly deteriorating vision, prompting the clinicians to treat as for infective endophthalmitis, which was similar to our patient. These patients eventually required systemic corticosteroids, with some requiring life-long immunosuppression for long-term control of their ocular inflammation.

4. Conclusions

HLA B27-associated uveitis can present in a range of clinical spectrum and posterior segment involvement can be seen uncommonly. In this patient, his symptoms were hyperacute; very severe fibrinous panuveitis and rapidly deteriorating VA occurring after intensive topical steroid therapy, giving rise to suspicions of infective endophthalmitis. This case highlights the diagnostic challenge and difficulty in management in cases of acute HLA B27-associated panuveitis which may result in unnecessary invasive diagnostic and therapeutic procedures.

Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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