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Differential Diagnosis of Pan-Uveitis: Behçet's Disease

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Abstract

This report describes the case of a 56-year-old man who presented with blurry vision, increased intraocular pressure, and conjunctival injection after posterior chamber intraocular lens implantation. Initially, post-operative endophthalmitis and foreign body inflammation were considered as differential diagnoses, but after further examination uveitis was diagnosed. Uveitis may present as an isolated finding or associated with a wide range of systemic diseases. During an interview following multiple visits and referrals, the patient mentioned a history of aphthous ulcers and genital ulcers, which then lead to the clinical diagnosis of Behçet's Disease. This report emphasizes that Behçet's Disease is rare in Caucasians and has variable presentations. Therefore, it is frequently misdiagnosed in North America due to failure to include it in the differential diagnosis. Inclusion of Behçet's Disease in the differential is crucial as potential adverse outcomes include blindness and death.

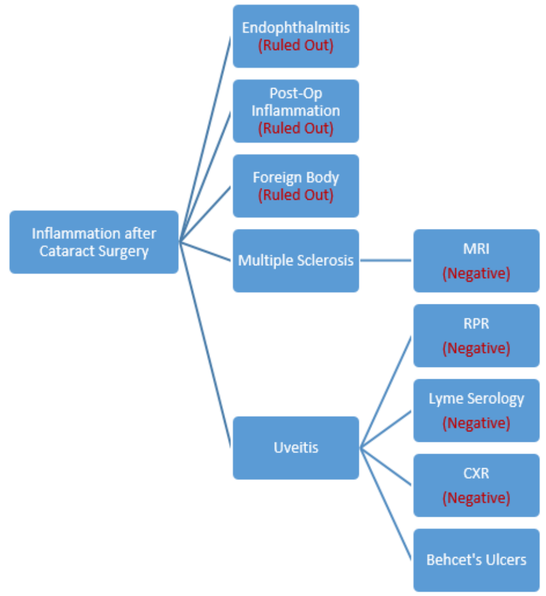
Introduction

- Differential diagnoses of uveitis include a post-inflammatory condition, foreign body inflammation, HLA-B27 Associated Uveitis, Reactive Arthritis, Sarcoidosis, Tuberculosis, Syphilis, Lyme Disease, Systemic Lupus Erythematosus, Acute Retinal Necrosis, and Behçet's Disease.
- The prevalence of Behçet's Disease in North America ranges from 1 in 15,000 to 1 in 500,000 depending on the region (1).
- The clinical triad includes uveitis, oral ulcers, and genital ulcers, but the clinical array of findings for the disease is diverse and includes erythema nodosum, ileocecal inflammation, epididymitis, pericarditis, aneurysms, as well as other ocular findings such as retinal vasculitis (2).
- Early recognition and treatment of this potentially blinding and fatal disorder is crucial (3), but the disease is frequently misdiagnosed (4).

Presentation

- Our patient was a 56-year-old Caucasian male with a past medical history including HTN, MI, arthritis, skin rashes, and skin cancer. Initial complaints were fluctuating blurry vision, dry eyes, and eye pain bilaterally. Examination revealed elevated intraocular pressure (IOP) with trace debris, corneal edema, and injection on anterior segment exam. This exam was performed one-week following cataract surgery with posterior chamber lens implantation, raising the concern for post-operative inflammation, foreign body reaction, or post-operative endophthalmitis. Prednisone treatment was initiated.
- Referral was made to a retinal specialist to rule out endophthalmitis. Exam revealed mild anterior chamber flare with a normal retinal exam and no sign of infection.
- The patient returned to the referring physician with worsening vision and a sensation of pressure upon awakening. IOP remained elevated, and ocular hypertension was diagnosed. Anterior segment inflammation remained and vision measured 20/50 OD and 20/40 OS. Timolol, Combigan, and artificial tears were prescribed along with steroids.
- Referral to a glaucoma specialist led to a diagnosis of uveitis. He also complained of pain with extraocular movement, and multiple sclerosis (MS) was added to the differential.
- At follow-up, IOP remained elevated and mild retinal vasculitis was noted. Differential diagnosis at this point included recurrent uveitis, MS, and Behçet's Disease.
- Rheumatology was consulted, and the patient was treated with cyclophosphamide. MRI was negative for inflammatory changes, ruling out MS. RPR testing, Lyme serology, and chest x-ray were also negative.Optic nerve edema, macular edema, and fluorescein angiography with characteristic retinal findings and vessel leakage were noted. The patient at this point stated that he had a history of aphthous ulcers and genital ulcers, which pointed to the diagnosis of Behçet's Disease although the patient had no ulcers on presentation.

Decision Tree



Discussion

- Our patient was a North American Caucasian, but he did not present with other clinical signs more commonly seen in Western Behçet's Disease, such as intestinal inflammation or pericarditis (5).
- Ocular involvement is a less common finding in Western Behçet's Disease, but our patient presented with uveitis, retinal vasculitis, and ocular hypertension. Uveitis is a classic ocular finding in Behçet's Disease, and retinal vasculitis can also classically be observed (6).
- Ocular hypertension is not a commonly reported finding in Behçet's Disease but was present in our patient, potentially representing inflammatory ocular hypertension syndrome related to uveitis (7).
- Our patient initially presented with blurry vision and had mild inflammation on exam. Given that he was presenting one week after posterior chamber intraocular lens implantation, further observation was recommended to determine if the inflammation was secondary to the stress of surgery.

Discussion (cont.)

- After further workup and examination, he was diagnosed with uveitis. When uveitis is diagnosed, ruling out an infectious cause is urgent, and workup should include an RPR to rule out possible syphilis infection, chest x-ray to check for sarcoidosis or tuberculosis, and Lyme serology.
- RPR has a sensitivity of 100% and specificity of 78%-86% in the secondary stage of infection (8). For disseminated Lyme disease, serology sensitivity ranges from 70%-100%, and specificity is 95% (9). Chest X-Ray assessing for TB has a sensitivity of 78% and specificity of 51% (10).
- If these tests are negative, inquiring about a past history of aphthous ulcers, genital ulcers, or other International Criteria for Behçet's Disease should be considered (3).

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