ACUTE UNILATERAL ADAMANTIADES BEHÇET UVEITIS – A CASE REPORT

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01. Introduction

Adamantiades-Behçet's disease (ABD) is a multisystem inflammatory disorder marked by recurrent oral aphthous ulcers, genital ulcers, and uveitis.(1) Its cause remains unclear, but several epidemiologic findings suggest autoimmune triggering from external (probably infectious) factors in genetically predisposed individuals.(2)

ABD is sometimes referred to as the "Silk Road disease" due to its higher prevalence along the historical route that joins the Mediterranean with the Middle and Far East and the regional variability that is observed can be partly attributed to spatial differences in the expression of HLA-B51 antigen.(3) The main age of onset is typically in the third or fourth decade, with an equal distribution between genders, though regional exceptions exist.(4)

Ocular manifestations occur in 50-90% of ABD patients, featuring bilateral nongranulomatous panuveitis and retinal vasculitis, however, some patients may only show unilateral involvement for several years. (1)

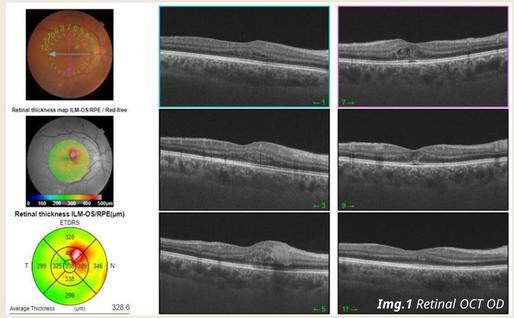
02. Case Presentation

A 40-year-old male patient presented to the emergency department with mild acute painless blurring of vision and floaters in the right eye, after first-time oral administration of naproxen.

Visual acuity was 20/20 and IOP was 10/11 mmHg. Cornea and anterior chamber were unremarkable. Dilated fundoscopy revealed mild localized vitritis over two white fluffy patches in the posterior pole of the affected eye. (Img.2)

Optical coherence tomography (OCT) of the right eye showed disruption of outer retinal architecture in the macular region, with intraretinal cystoid spaces along with a small amount of subretinal foveal fluid. (Img.1)

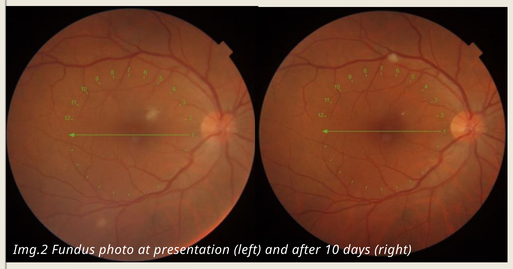
Peripapillary Retinal nerve fiber layer (RNFL) thickness map had no abnormal findings. Left eye imaging was unremarkable.



***Img.1** Retinal OCT OD*

The patient was undergoing laboratory investigation for autoimmune arthritis, experiencing wrist, ankle, and hip pain for over two weeks, hence the administration of oral naproxen. The patient also reported a maculopapular rash around the orbits and although denying genital ulcers, he admitted to a history of recurrent aphthous stomatitis for years without ever seeking medical attention.

The combination of recurrent oral ulceration, arthralgia, facial rash, and posterior uveitis led to a suspicion of autoimmune/systemic etiologies. A complete posterior uveitis panel was ordered for further investigation.

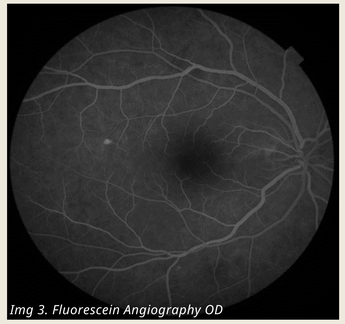


Img.2 Fundus photo at presentation (left) and after 10 days (right)

Topical dexamethasone drops were immediately started x4 daily, resulting in mild improvement.

Fluorescein angiography revealed no active vasculitis or ischemic areas, only an occult inflammatory lesion not initially detected in fundoscopy or OCT imaging. (Img.3)

Ocular symptoms recurred a few days later, with new inflammatory foci emerging in different posterior pole areas, stressing the need to promptly suppress inflammation.



Img 3. Fluorescein Angiography OD

Close collaboration with the rheumatologists included a positive pathergy test, contributing to the clinical diagnosis of Adamantiades-Behçet's disease with a score of 6 according to the International Criteria for Behçet disease.(4)

Immediate initiation of oral steroids and azathioprine was chosen over waiting for laboratory results to expedite therapy. This approach resulted in both functional and anatomical improvement, evidenced by resolution of floaters and restoration of retinal OCT architecture.

Subsequent positive HLAB51 results supported the diagnosis, while all other exams for the differential diagnosis of posterior uveitis, including HLAB27, were negative.

03. Discussion

Blurring of vision caused by non-steroidal anti-inflammatory drugs is rare but has been documented in literature. (5) In our case, this initially misleading symptom accelerated the diagnosis of ABD. Our differential diagnosis included other autoimmune causes of uveitis (especially ankylosing spondylitis and lupus retinopathy), HIV retinitis, CMV retinitis, syphilis, tuberculosis, sarcoidosis, toxoplasmosis, rickettsiosis and bartonellosis, all of which were excluded during laboratory investigation.

The interesting element in this case was that ocular signs of posterior uveitis were the hint that steered ophthalmologists toward ABD. Initially, seronegative arthritis was considered by the rheumatologist as the most probable diagnosis based on available radiologic and laboratory findings. However, the absence of anterior segment involvement, the presence of posterior uveitis inflammatory foci, and the related systemic symptoms prompted further investigation, leading to a confirmatory diagnosis of ABD with positive HLA genotyping.

At this point, we would like to underline that there are no specific diagnostic tests for ABD, stressing the importance of detailed history-taking to support a clinical diagnosis. (6)

Moreover, ABD typically follows a relapsing course, marked by the emergence of new retinal infiltrates (even without concurrent vitritis or angiitis), which often resolve spontaneously within days without scarring, serving as indicators of disease activation. In our case, the patient exhibited migratory infiltrates across various posterior pole areas, representing relapsing and remitting lesions, some detectable only angiographically.

Prompt initiation of systemic steroids and immunosuppressive agents effectively controlled the disease, preserving the patient's visual acuity. Ongoing assessments are conducted to monitor disease reactivation and potential drug-related side effects.

This case underscores the importance of interprofessional collaboration between ophthalmologists and rheumatologists for optimal patient care outcomes in autoimmune diseases with ocular manifestations.

04. Conclusion

-Adamantiades-Behçet's disease (ABD) is a clinical diagnosis, and no specific diagnostic tests are available, so detailed history-taking is essential for establishing sound diagnostic choices.

-Ocular manifestations may be the key that aids rheumatologists towards the correct diagnosis of ABD, especially in relation to other uveitides with systemic symptoms.

-ABD runs a relapsing course that calls for timely initiation of effective treatment, with minimal delay, in order to suppress inflammation, restore tissue damage and prevent recurrent attacks.

-This case serves as a chance to remind clinicians that partnership between ophthalmologists and rheumatologists plays a crucial role in enhancing diagnostic accuracy and therapeutic interventions in autoimmune uveitis patients.

05. References

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