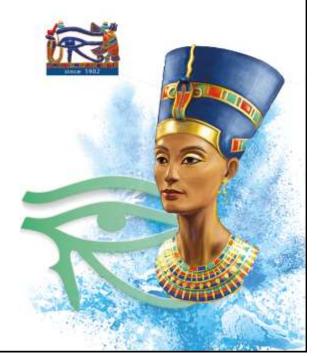


One disease with different faces

Ashgan M. Asaad., MSc.



Case (1)

- 32 years old male.
- Complaint: outward deviation of both eyes.
- Seeking for strabismus surgery.
- Examination





Case (1)

	Right Eye	Left Eye
BCVA	0.2	0.2
EOMs	Free EOM movements Intermittent XT /Fair control	
IOP	11	12
Cornea	Clear	Clear
AC	No cells/ No flare	No cells/ No flare
Pupil	RRR	RRR
Lens	Clear	Cear
Vitreous	Vitreous cells/haze +3	Vitreous cells /haze +3
Fundus	Hazy view of attenuated and sheathed vessels	





Case (1)

- On meticulous history taking:
- He mentioned that he was smoker and had previous attacks of oral and genital ulcerations.
- Behçet disease was considered and immunology refer was done.
- The exotropia was explained that it was intermittent with good control and he lost the control after developing posterior uveitis and diminution of vision.



Case (2)

- 23 years old male patient.
- Complaint: Bilateral diminution of vision > 1 year ago.
- History of weight loss, poor appetite, skin brown patches.
- TB diagnosis was made by +ve quantiferon gold test 1 year ago and antituberculous therapy was completed with no improvement of VA.
- Examination:



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Case (2)

	Right Eye	Left Eye	
BCVA	CF 50 cm	1/60	
IOP	10	12	
cornea	Prominent corneal nerves Iris pigments on the back of the cornea		
AC	cells +3/ flare +1	cells +2 / flare +1	
Pupil	RRR Persistent pupillary membrane	RRR	
Lens	Iris pigments on anterior capsule		
Vitreous	Vitreous cells / haze +2	Vitreous cells / haze +1	
Fundus	Hazy view of attenuated and sheathed vessels		

3

Case (2)

Laboratory investigations:

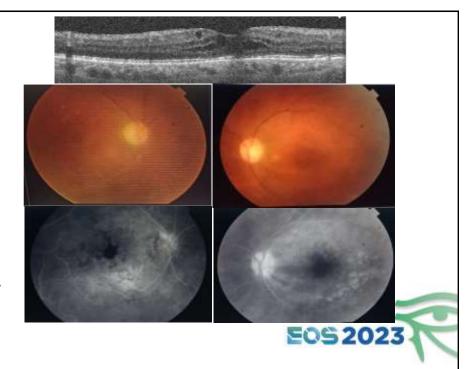
- ESR .. 1st hour (10)- 2nd (22).
- CRP....(3.2).
- Liver function tests Normal.
- Kidney function tests Normal.
- Quantiferon gold test ... positive.



Case (2)

FFA and OCT macula were done:

- Perivascular leakage, vasculitis and papillitis.
- Macular edema.



Case (2)

- Refer to both immunology and chest consultation.
- After immunology consultation Behçet disease was diagnosed by skin lesions and appearance of both oral and penile ulcers.
- After chest consultation: Bilateral subtenon injection of steroids was done and immunosuppression therapy was started.
- VA was improved to 1/60 (OD) and 3/60 (OS) after one month of immunosuppression.



Case (3)

- 28 years old male patient, smoker.
- Complaint: Bilateral diminution of vision since childhood with worsening few months ago.
- Vague history of corneal trauma in infancy.
- Seeking for corneal transplantation.
- On examination: skin lesions were observed and history of recurrence was taken.
- Herpetic keratitis was suspected and oral acyclovir 400 (1×5) was taken with no improvement.
- Examination:





	Case (3)		
	Right Eye	Left Eye	A 1000
BCVA	1/60	4/60	
IOP	5	6	100000000000000000000000000000000000000
Cornea	Vascularized central opacity	Paracentral opacity	
AC	No cells / flare.	No cells / flare.	
Pupil	RRR	RRR	
Lens	clear	clear	
Vitreous	Posterior vitreous cells / haze +2		THE TOTAL
Fundus	Hazy view of retinitis, sheathed and ghost vessels		
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Case (3)

Laboratory investigations:

RBCs: Normocytic Normochromic. WBCs: Mild absolute Neutrophilia,

Platelets: Adequate

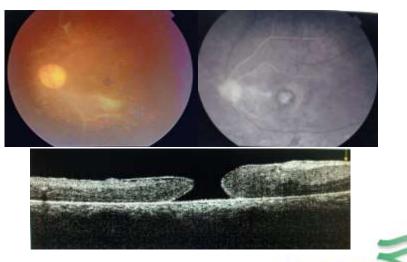




FFA of left eye showed:

- vasculitis and papillitis.
- Macular hole confirmed by OCT.

Case (3)



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Case (3)

On meticulous history taking:

- He was suffering from attacks of oral and genital ulcers.
- Behçet disease was considered and immunology refer was done.
- Steroid and immunosuppressive therapy was started.



Behçet disease (BD)

- Behçet disease is a chronic, recurrent, multisystem disease.
- Characterized by occlusive and necrotizing vasculitis.
- Ocular involvement, which is seen in up to 70% of patients, is characterized with anterior uveitis, retinitis, retinal vasculitis, vitritis, retinal vein occlusion, macular edema, optic disc hyperemia, and edema.



Behçet disease

Diagnostic Criteria.... The International Study Group for Behçet Disease (ISGBD),

- Recurrent oral ulceration characterized by oral ulcers at least three times in a 12 month period, plus at least two of:
- · Genital ulceration.
- · Ocular inflammation.
- Characteristic skin lesions: (erythema nodosum, pseudofolliculitis, acneiform nodules, papulopustular lesions) and
- Positive pathergy reaction: pustule 24–48 hours after a sterile needle prick (>95 %specific, but often negative in European and North American patients).
- But, presentation does not always conform to the criteria above.



Behçet disease

- Additional features include:
- Vascular lesions. Aneurysms, including pulmonary and coronary and venous thrombosis or thrombophlebitis.
- Arthritis occurs in 30%, though arthralgia is more common.
- Dermatographia, similar to the pathergy reaction, indicates skin hypersensitivity and consists of the formation of erythematous lines following stroking or scratching.
- Neurological manifestations (5%) such as meningoencephalitis of the brainstem, dural sinus thrombosis and cerebral aneurysms.
- Gastrointestinal inflammation, especially ileocecal.
- Hepatic and renal lesions are relatively uncommon.



Association of BD with Strabismus

Journal of the Korean Ophthobosiograd Society 1991;32(8):675-682.
Published online August 1, 1991.

Isolated Unilateral Lateral Rectus Muscle Palsy in Neuro-Behret Syndrom.

Chong Kun Lee, Jong Bok Lee, Hong Bok Kim

Department of Ophthalmology, Yonsei University College of Medicine, Seoul, Korea.

Abstract

Behcet syndrom is a chronic systemic disease developing recurrent oral and genital ulceration, ocular inflammation, arthritis and neurological manifestations. Anterior uveitis and occlusive retinal vasculitis are the most commom ocular manifestation of Behcet syndrome. A 29 years old Korean male with abducens palsy (6th cranial nerve palsy) was found to have oral aphthae, genital ulcer, skin lesion and other neurological manifestations (weakness and sensory loss of lower extremeties, ataxic gait, speech disturbance and urinary difficulty), but he had neither ocular manifestation such as uveitis nor retinal vasculitis.



Association of BD with TB

Yan Shen", Haifen Ma", Dan Luo, Jianfel Cal, Jun Zou, Zhijun Bao", Jianlong Guan'

Behçet's disease with latent Mycobacterium

tuberculosis infection

tions. Environmental influences and genetic factors may play a role in the etiopathogenetic mechanisms that lead to the development of the disease, indicating the autoimmune and autoinflammatory nature of BD. In

Behçet's Disease and Tuberculosis: A Complex Relationship

Sara Mendonca Footas, Asara Silva Marques, Ana Grito, Rudolfo Gomes, Fernando Martos Gonçalves Serviço de Medicina Interna, Hospital Beatriz Ángelo, Loures, Portugal

Behçet's disease (BD) is a systemic vasculitis characterized by recurrent orogenital ulceration and several systemic manifestations (such as gastrointestinal involvement, vancular disease or arthritis). The pathogenesis is still unknown but the trigger rule of certain pathogens such as Mycobacterium tuberculosis is well-documented. Furthermore, patients with BD are more susceptible to tuberculosis due to immunity. defects. Here, we describe the case of a 70-year-old woman with a history of recurrent oral aghithae and inflammatory arthritis presenting

JAMA Ophthalmology | Original Investigation

Zhenyu Zhong, MD: Guannan Su, PhD; Qingyun Zhou, MO, Akira Meguro, PhD; Masaki Takeuchi, MD, PhD; Nobuhisa Mizuki, MD, PhD; Shigwaki Ohno, MD, PhD; Welting Liao, MD; Xiaojie Feng, MD; Jiadong Ding, MD; Yingnan Gao, MD; Wei Chi, MD, PhD; Meifer Zhang, MD, PhD; Xianto Zuo, PhD; Peizeng Yang, MD, PhD

Tuberculosis Exposure With Risk of Behçet Disease
Among Patients With Uveitis

In conclusion, human genetic and bromarker data demonstrated that an increased risk of subsecutiosis infection was associated with a history of uveitis. Our study provides novel evidence linking an infectious agent to the risk for noninfec-



Association of BD with HSV and keratitis

March 1991

Bilateral Corneal Immune Ring Opacity in Behcet's Syndrome

Smadar Cohen, MD; Israel Kremer, MD

3 Author Affiliations

Arch Ophthalmol. 1991;109(3):324-325. doi:10.1001/archopht.1991.01080030026 023

Immunopathogenic Role of Herpes Simplex Virus in Behçet's Disease

Do Young Kim, Suhyun Cho, Min Ju Choi, Sconghyang Sohn, 2 Eun-So Lee,3 and Dongsik Bang

9. Conclusion

The exact pathogenesis of BD remains elusive, although research continues to increase our understanding of the complex pathogenetic mechanisms of the disease. Infectious agents, such as HSV and Streptococcus sanguinis, have long been postulated as possible environmental triggers of BD, and the role of HSV continues to be a main focus of BD research. Clinical observations and the detection of HSV DNA in saliva and genital ulcers of BD patients prompted us to investigate the effects of HSV in BD in vitro and in vivo.

Take home message

- Don't rush for the first presentation before systematic examination and meticulous history taking.
- Fundus examination is mandatory in strabismus cases to exclude sensory causes.
- Fundus examination is mandatory in all cases of keratitis and uveitis to exclude posterior uveitis and for classification of the case (anterior, intermediate, posterior or pan-uveitis)according to SUN classification.
- Combined pathologies exist in the field of uveitis.
- Infectious agents such as TB and HSV act as environmental triggers of BD in addition to genetic factors.

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Thank you

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