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Ocular Manifestations Revealing of Behçet's Disease: About 42 Cases

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Abstract **Original Research Article**

Introduction: Behçet's disease is a chronic, autoimmune, multisystem of unknown etiology, characterized by oral ulcers, genital ulcers, and eye lesions. Ophthalmological manifestations of Behcet's disease are numerous and are often indicative of the disease. Our job is to study these events in a group of patients in whom the ophthalmologic involvement was indicative of Behcet's disease. ***Materials and Methods***: A retrospective study in the ophthalmology department at the military hospital of instruction Mohamed V in Rabat between January 2009 and December 2015. It involved 42 patients who initially consulted for eye symptoms and in that the diagnosis of Behçet's disease was selected based on the criteria of ISG. ***Results:*** The initial ocular demonstrations were non-granulomatous anterior uveitis in (15,06%) of which (10,9%) with hypopyon, intermediate uveitis (9.5%), ocular hyalite (12.32%), panuveitis in (41,1%), retinal vasculitis (16.43%), retinal vein occlusion (3.6%), papillary edema (1.8%). The criteria used for the diagnosis of Behcet's disease were essentially have oral ulceration, genital ulceration and pathergy test. ***Discussion*:** In 10% to 20% of cases, ocular involvement may be opening, remaining isolated for several years. The most frequently revealing clinical aspects of Behcet's disease are the anterior uveitis, and vasculitis panuveites. ***Conclusion*:** the ethnic and geographic origin of the patient, the young age and some clinical features of ocular lesions should remind ophthalmologists to Behcet's disease to inflammation of one of the ocular structures and encourages him to seek other diagnostic criteria in collaboration with other specialities.

Keywords: Behçet's disease, diagnostic criteria, ocular manifestation, uveite, aphtose.

INTRODUCTION

Behcet's disease is a chronic autoimmune and multisystemic disease of unknown etiology. It was first identified in 1937 by Hulusi Behcet, who noted that patients exhibited three primary symptoms: recurrent oral ulcers, genital ulcers, and iritis [1]. Since then, several diagnostic criteria have been employed until the International Study Group for Behcet's Disease established the current criteria in 1990 [2-4]. According to these criteria, recurrent oral ulcers are mandatory, as well as recurrent genital ulcers, skin lesions, uveitis, and a positive pathergy test.

Ophthalmological manifestations are common in Behcet's disease and may be its first revealing sign. Our study aims to examine these manifestations in a group of patients whose ocular findings led to a diagnosis of Behcet's disease.

METHODS AND METHODS

This study was conducted in the ophthalmology department of the Military Hospital of Instruction Mohammed V in Rabat from January 2020 to December 2023. It is a retrospective study that included 42 patients who initially consulted for ocular symptoms and were later diagnosed with Behcet's disease based on the criteria set by the International Study Group. The study parameters covered various aspects, including age, sex, clinical findings, bilaterality of the attack, Behcet's diagnostic criteria, ophthalmological complications, and the treatment administered.

RESULTS

This study included a total of 42 patients (73 eyes). Of these patients, 26 were men (61.9%) and 16 were women (38.1%). Patients' ages ranged from 17 to 66 years old, with an average age of 29.4 +/- 12.58.

The patients' initial ocular manifestations included nongranulomatous anterior uveitis (11 eyes; 15.06%) with hypopyon in eight of those eyes (10.9%), intermediate uveitis in seven eyes (9.5%), hyalitis in nine eyes (12.32%), retinal vasculitis in 12 eyes (16.43%) (Figure 1), retinal vein occlusion in three eyes (4.1%),

and papilledema in one eye (1.3%) (Tabe 1). The ocular manifestations were bilateral in 27 patients (64%). The most common bilateral manifestations were panuveitis, retinal vasculitis, and anterior uveitis.

The diagnosis of Behcet's disease was made based on the presence of oral and genital ulcers along with a positive Pathergy test (Table 2). After the diagnosis, the patients were treated with a three-day course of Solumedrol with an initial dose of 10mg/kg/day, followed by oral administration of Prednisone at a rate of 1mg/kg/day. Topical medication with corticosteroids and Tropicamide has also been prescribed.

A treatment consisting of immunosuppressive therapy (Azathioprine) in association with oral corticosteroids was initiated in 27 patients. Other patients did not receive this treatment as their ocular manifestation was purely anterior.

All patients responded well to the treatment, and ocular inflammation has been controlled. Some ophthalmological complications arose during the follow-up period. The most common complications were cataracts, posterior synechiae (Figure 2), cystoid macular edema (Figure 3), and ocular hypertonia. Three patients developed retinal ischemia and optic neuropathy.

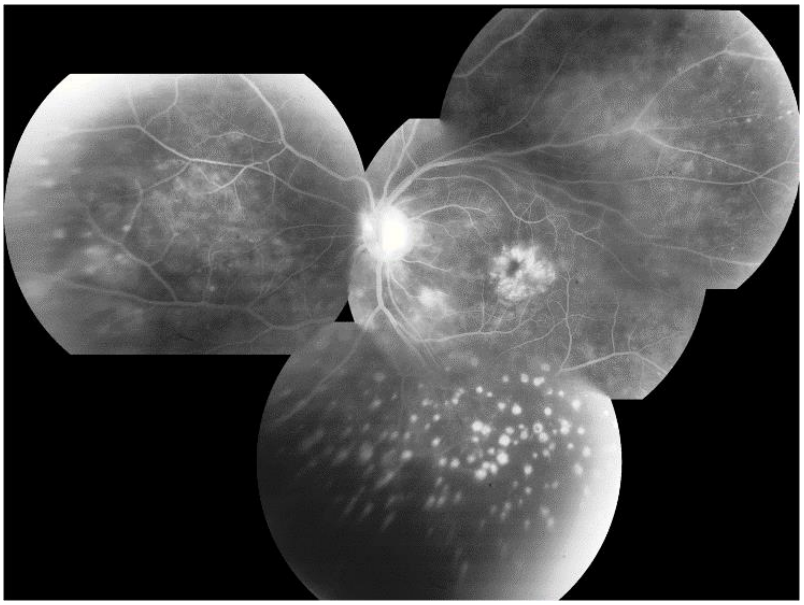

Figure 1: Retinal angiogram showing retinal vasculitis with macular edema and hyalitis

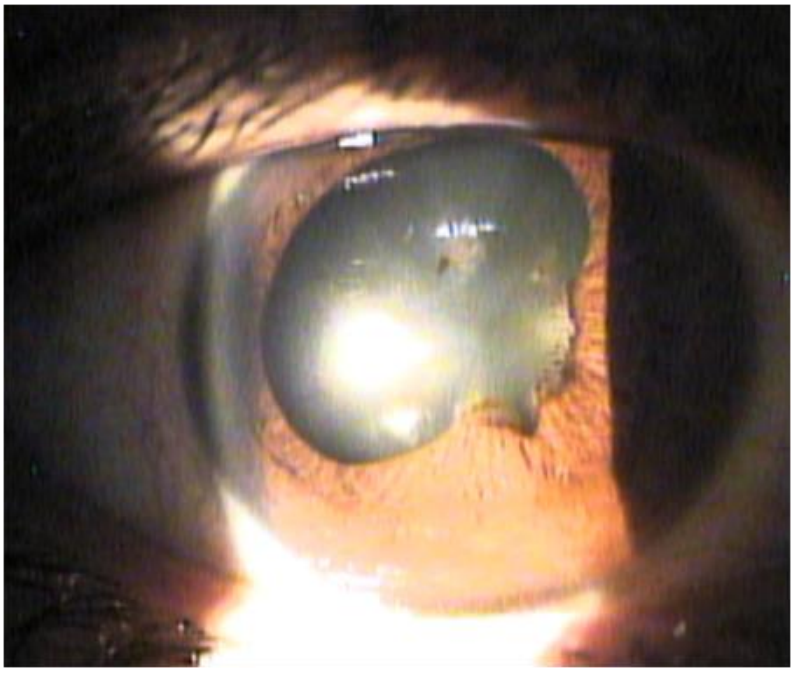

Figure 2: Photography of the anterior segment showing posterior synechiae

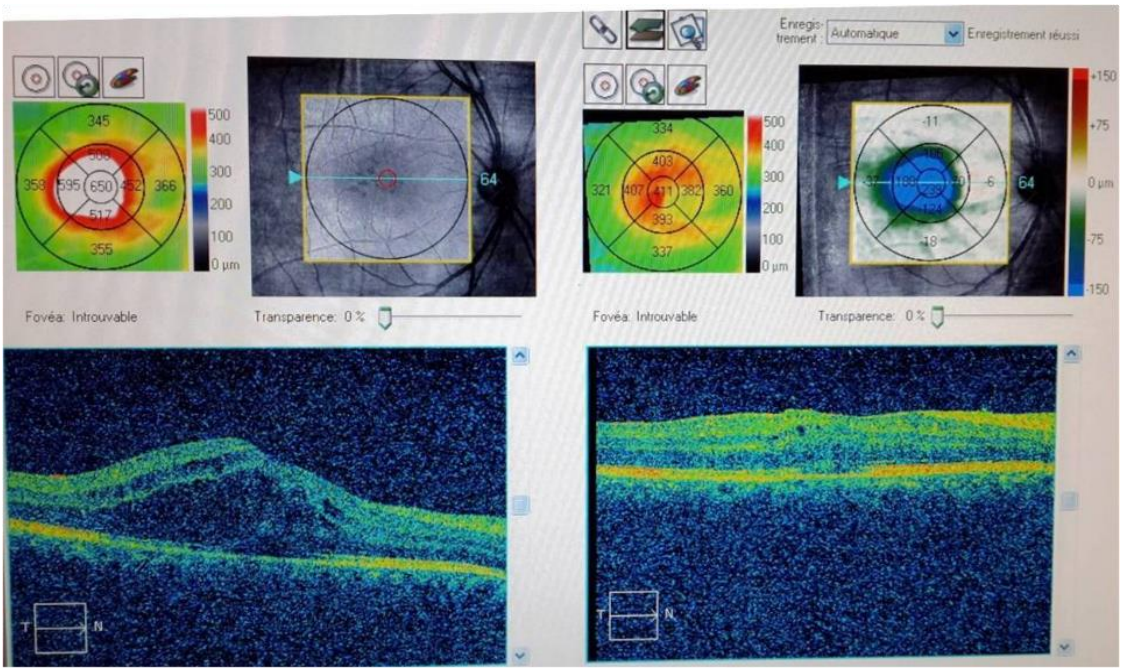

Figure 3: OCT-scan showing cystoid macular edema before and after treatment

Table 1: Initial ocular manifestations of Behcet's disease in our patients

Initial ocular manifestations	(% des yeux)	
	men	women
anterior uveitis	9,59	5,47
Intermediate uveitis	5,47	4,1
hyalitis	6,8	5,47
panuveitis	27,39	13,69
retinal vasculitis	9,58	6,84
retinal vein occlusion	2,73	1,36
Pappiloedema	0	1,36

Table 2: Diagnostic criteria of Behcet's disease in our patients

The criteria for making the diagnosis of Behçet's disease	% des patients
Eye damage	100
Mouth aphtosis	100
Genital aphthosis	80,95
Skin lesions	33,33
Pathergy test	90,47
Neurological damage	0
Vascular damage	0

Table 3: Ophthalmological complications in our patients

Ophthalmological complications	Nb des cas / 73 yeux	Pourcentage (%)
Cataract	11	15,06
Irido-lens synechiae	20	27,39
Ocular hypertonia	5	6,84
Intravitreal hemorrhage	1	1,36
Macular edema	11	15,06
Blindness	3	4,1

DISCUSSION

Behcet's disease is a worldwide condition, but it is more common in countries corresponding to the ancient Silk Road, especially in the Middle and Far East, East Asia, and the Mediterranean region [3-5]. It affects both men and women, but it tends to be more severe in men. The male-female ratio varies depending on the region, with a ratio of 0.98 in Japan, 0.63 in Korea, 1.19 in Iran, 1.03 in Turkey, and 1.8 in India [3, 4, 6, 7]. In our study, the male-female ratio was 1.62. The disease

usually starts between 25 and 30 years. But can also appear in childhood or after 60 [4]. The average age of our patients was 29, which is similar to the average age reported in other studies, such as 35.7 years in Japan, 29 years in Korea, 25 years in Iran, 25.6 in Turkey, 28 in Greece, and 24.5 in the United Kingdom [4, 6]. Behcet's disease is a type of systemic inflammatory condition that is characterized by oral and genital ulcerations, ocular inflammatory damage, skin lesions, vascular damage, as well as numerous visceral damages that can be potentially life-threatening [2, 3, 4, 6]. The eye is the most commonly affected organ in Behcet's disease. The initial ocular inflammatory process is typically anterior and unilateral, and it tends to involve the posterior segment of the eye. Later, it can become bilateral [3, 4, 7-9].

The incidence of ocular involvement in Behcet's disease varies depending on the studies, ranging between 28% and 70%. Ocular involvement typically occurs around four years after the onset of systemic symptoms, particularly oral aphthosis [2, 10-13]. However, in 10% to 20% of cases, ocular manifestations may appear first and remain isolated for a few years [2, 7, 14, 15]. Anterior uveitis is a rare condition that may occur in 10% of cases [7, 11, 14, 16]. It is always characterized as nongranulomatous and can be unilateral or bilateral. Typically, it manifests as an anterior uveitis, associated with hypopion, indicating a severe inflammation in the anterior chamber in 12 to 30% of cases [3, 16].

Posterior uveitis is a severe condition that may compromise visual function. It is observed in 32% to 53% of cases and can manifest in various forms, including variable severity hyalitis, white-yellow retinal lesions, or hemorrhagic lesions with varying numbers and locations [3, 14, 17]. Additionally, occlusive retinal vasculitis is a crucial sign of ocular involvement in Behçet's disease, and it's found in over 90% of patients. This occlusive vasculitis can lead to ischemia and may affect the visual functional prognosis. It impacts significantly the veins [3, 14, 15]. Papilledema is a rare condition that can be linked to intracranial hypertension, which may occur due to venous sinus thrombophlebitis or inflammatory and/or ischemic optic neuropathy [2, 4, 5].

The diagnosis of Behcet's disease is mainly based on a group of clinical elements found in the same patient [18]. There have been different diagnostic criteria proposed over the years. However, the most widely used criteria nowadays are the ones developed by the International Study Group for Behçet's disease in 1990 [19]. According to these criteria, oral aphthosis is a mandatory diagnostic symptom and must be recurrent, along with two of the other criteria, which include recurrent genital ulcers, eye damage, skin damage, and a positive pathogenic test. International Team for the Revision of the International Criteria proposed new international criteria to diagnose Behçet's disease. The criteria assign a score of 2 to ocular manifestations, oral and genital aphthosis, and a score of 1 to skin, neurological, and vascular damage, as well as a positive pathergic test. When the total score is greater than or equal to 4, the diagnosis of Behçet's disease is established [4]. No biological test is specific for the disease, and HLA typing is generally not helpful for diagnosis. However, in regions where Behçet's disease is not common, HLA typing could help with diagnosis. There is a higher prevalence of HLA B5, particularly HLA B51. Furthermore, a recent Japanese series highlighted a correlation with the phenotype of HLA-A26, found in patients who present the HLA-B51 allele. This phenotype would increase susceptibility to the disease and may be associated with a poor visual prognosis [2].

Several complications can mark the progression of uveitis associated with Behçet's disease and compromise the visual prognosis. They are attributable to inflammatory exacerbations, lesion progression, or anti-inflammatory treatment. The most common complications are macular edema (20 to 75% of cases), posterior synechiae in 26% of cases, cataracts in 39% of cases, and optic atrophy and glaucoma in 14% of cases [3, 4, 20]. Retinal neovascularization can occur due to ischemic occlusive vasculitis in 3.4-8% of cases. Vitreous hemorrhage, retinal dehiscence or retinal detachment, as well as phthisis bulbi, are rare complications [2, 4, 5].

Management of the ophthalmic manifestations linked with Behçet's disease aims to swiftly and effectively control acute inflammatory exacerbations and lessen the frequency and intensity of recurrences to minimize the risk of complications and sequelae that could eventually lead to visual impairment. This treatment usually combines corticosteroids, used for their immediate anti-inflammatory action on acute attacks, and immunosuppressants as long-term background therapy [12, 14, 20].

Cases of isolated anterior uveitis require local corticosteroid treatment without associated systemic therapy [14]. The mere presence of vitreous cells or purely angiographic vasculitis, even in the absence of another clinical abnormality in the posterior segment, is a clear indication for therapy with immunosuppressants [3, 5, 13]. Surgical intervention may be necessary to treat ocular complications related to Behçet's disease, such as cataracts or glaucoma. Cataract surgery, if needed, must be performed in a calm eye, in remission for at least three mounths. Vitreoretinal surgery is indicated for the treatment of complications such as macular holes, retinal detachment, or persistent vitreous hemorrhage. Finally, laser photocoagulation is recommended in the management of secondary neovascular complications resulting from occlusive vasculitis.

CONCLUSION

Ocular involvement is the initial manifestation of Behçet's disease in 10% to 20% of cases. The diagnosis must be suspected based on the ethnic and geographical origin of the patient, the young age, and clinical findings. Collaboration with other specialties to identify additional diagnostic criteria is crucial to confirm the diagnosis.

The authors declare that they have no conflicts of interest

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