

Macular Complications of Behcet Disease: What to Expect?

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ABSTRACT

Purpose: To determine the rate of incidence and the most common type of macular complications in patients with Behcet disease.

Methods: In this monocentric retrospective clinical study we examined all patients diagnosed with Behcet disease between April 2018 and April 2022 (150 patients). Each individual would undergo a complete ophthalmological examination, a fundus retinography, OCT scan and fluorescein angiography.

Results: 54 patients of 150 presented a macular complication (36%). The macular involvement was bilateral in 49 patients. 60% of patients presented with an initial best corrected visual acuity (BCVA) of equal or less than 1/10. The most common types of maculopathy found in our study was cystoid macular edema (60.4% of patients), macular degeneration (32%), macular hole (5.3%) and serous retinal detachment/epimacular membrane (2.3%).

Conclusion: Macular involvement is very common in Behcet syndrome evolution and is responsible for severe and, most often, irreversible sight loss. Therefore a proper and early management of the disease is mandatory in order to minimize visual impairment.

Keywords: Behcet disease, macular complications, macular edema, ophthalmology.

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I. INTRODUCTION

Behcet's disease is a dysimmune multisystemic vasculitis that affects different organs in a variable way. Ocular manifestations of Behcet's disease are one of the disorder's major criteria. Macular complications are variable and, most of the time, are responsible for a severe decrease of visual acuity and a poor prognosis of the disease [1].

The purpose of our study is to evaluate the incidence rate of macular complications in Behcet disease, as well as to determine the most frequent types of maculopathies in the pathological entity.

II. MATERIAL AND METHODS

Our study of macular complications in Behcet's disease is a mono-centric retrospective clinical study conducted at both Military teaching hospital of Rabat and Provincial hospital of Tetouan between April 2018 and April 2022. The patient's data was gathered in both ophthalmology and medicine departments.

We included all admitted patients that were diagnosed with Behcet disease according to the International Study Group of Behcet Disease.

Each patient underwent a complete ophthalmological

examination including visual acuity testing, slit lamp and fundus examination. Then all patients had their macular oct scans using Zeiss OCT Cirrus 4000, a retinal photography and fluorescein angiography.

III. RESULTS

150 patients were included in our retrospective study, all diagnosed with Behcet disease (BD). Among these patients 54 had macular complications of BD.

A. Demographic Features

The mean age of our study group was 52 ± 4 years old. As for gender, 75% were males while 25% were females. The mean evolution time of the disease was 3 years.

36% of the examined patients presented a macular complication of BD (54 patients for a total of 150 included subjects). 27% had the maculopathy at the time of the examination while 9% presented them during the follow up period.

The macular complications were bilateral in 49 patients, be it 98 eyes among 300 examined (32.6%).

B. Clinical features (Fig. 1 to 5)

For 54 patients who presented a macular complication of BD during the disorder's evolution we have noticed that:

- 60% of patients presented with an initial best corrected visual acuity (BCVA) of equal or less than 1/10. While 25.50% had a BCVA in the range of 2/10 and 5/10. Only 15.50% had a BCVA \geq 5/10 (Fig. 1).

Upon the ophthalmological examination, the OCT scans and Fluorescein angiography findings the distribution of macular complication types was as follow:

- The main maculopathy type in BD for our patients was cystoid macular edema with a rate of 60.40% (figure 2). Macular degeneration comes in second with 32% of macular complications shares (figure 3). As for the other types, we accounted for macular hole with 5.30% and both serous retinal detachment (SRD) and epimacular membrane (EMM) for the remaining 2.30%. (Fig. 4 and 5).

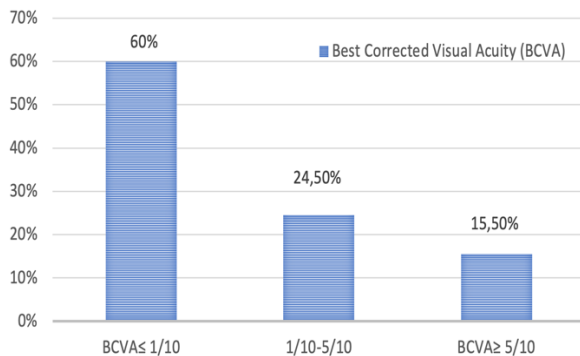


Fig. 1. Distribution of best corrected visual acuity of our patients.

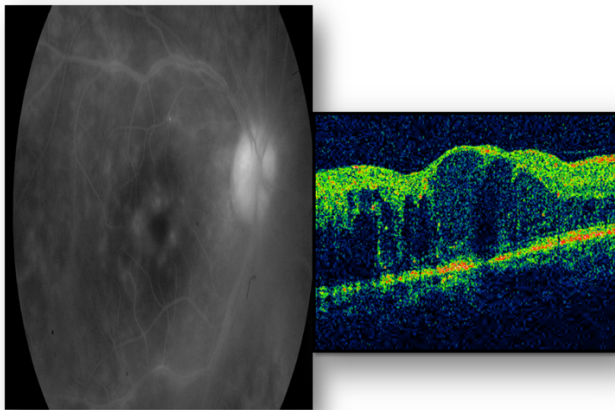


Fig. 2. Cystoid macular edema in a patient with Behcet disease, fluorescein angiography and OCT scan.

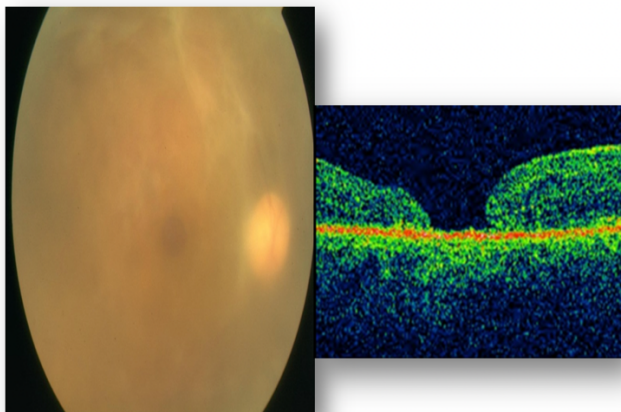


Fig. 3. Macular hole in patient with Behcet disease, Fundus retinography and OCT scan.

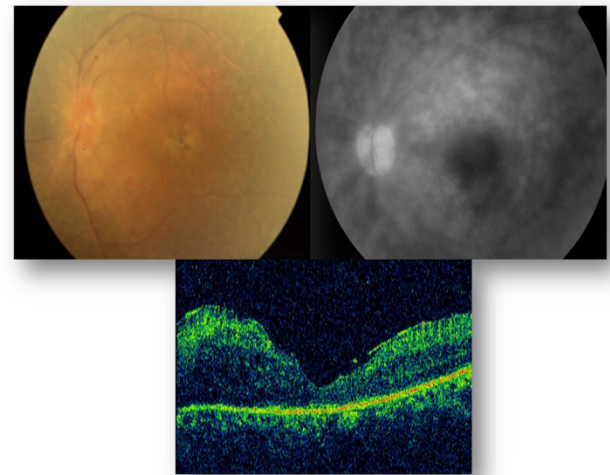


Fig. 4. Macular degeneration, fundus retinography, Behcet disease, fluorescein angiography and OCT scan.

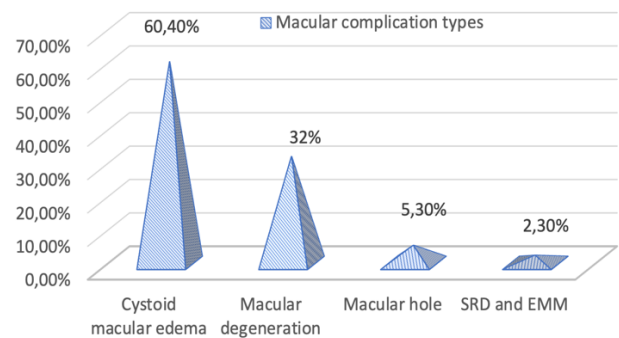


Fig. 5. Distribution of Macular complication's type in Behcet disease, SRD: Serous Retinal Detachment, EMM: Epimacular +membrane.

IV. DISCUSSION

Behcet disease is a relatively rare systemic multiorgan vascularitis that was formally described by dr. Hulusi Behcet in 1922, with a typical spread in the Middle and far East of the globe [2].

This syndrome is best known as a triad of symptoms associating oral and genital ulcers as well as ocular complications affecting 70% of BD patients [3]. However, major and minor criteria have now been well established by the International Study Group (ISG) in order to help include or exclude Behcet disease diagnosis [4].

The ocular manifestations of Behcet syndrome are variable and quite sight threatening. The most common manifestation of the disease is an explosive nongranulomatous uveitis associated to a hypopyon in most cases [5]. Alongside comes inflammatory complications due to the recurrent and relapsing nature of the disease such as: posterior synechiae, iris bombe, uveitic cataract and angle closure glaucoma [6]. However, as the entire uveal tract can be affected by this disorder, the posterior segment involvement can be considered as the most serious ocular manifestation of Behcet disease. Typically, it presents as a nongranulomatous obliterative vascularitis of the retina, responsible for necrotizing symptoms such as branch retinal vein occlusion [7], which consequently leads to neovascularization phenomenon and indefinite sight loss [8].

Macular involvement in Behcet disease is quite common.

Reference [9] found that among 300 patients diagnosed with Behcet disease 50% presented macular complications. [10] and [11] concluded to a medium rate of maculopathy incidence in patients with Behcet syndrome for a corresponding percentage of 16% and 24% of 92 patients and 108 patients accordingly. Another study, [12] that included 90 patients described a rate of incidence of macular complications of 19% among Behcet disease patients.

As for the occurrence of maculopathy during Behcet syndrome evolution, our study doesn't fall far from the literature's results with a rate of 36% among a total of 150 patients.

Macular complications in Behcet disease can be quite variable. The most common type is cystoid macular edema that is believed to be a direct result of posterior segment inflammation cascade. The disruption of the normal blood-retinal barrier leads to an accumulation of fluids in the outer plexiform layer of the macula which forms many cavities within the intracellular spaces of the retina [13]. Reference [14] observed the occurrence of different types of maculopathies in Behcet disease evolution and concluded, through their study, that the most frequent type of macular lesion was the cystoid macular edema. The same goes for our observation as 60.40% of our patients suffering from a macular complication of Behcet disease had a cystoid macular edema as a specific maculopathy.

Macular degeneration (MD) is the second common macular complication of Behcet disease. In a study conducted by [15] 19% of 111 eyes suffered MD. This type of maculopathy can be seen initially in patients with Behcet disease, as it can result from the chronic evolution of untreated cystoid macular edema. In our series, the share of patients with MD was of 32%.

As for macular hole, it remains a rare complication of Behcet disease panuveitis. It was reported in only 2.6% of 880 patients in a big study conducted by [16], and in 3.4% in [12]. As for our study, 5.3% of patients presented a macular hole during the follow up period. Macular hole can be a direct result of Behcet panuveitis inflammation that causes vitreoretinal interface changes leading to macular traction [17].

Furthermore, some studies reported serous retinal detachment (SRD) and epimacular membrane as a possible macular complication in Behcet disease evolution [18]. These lesions were also seen in 2.3% of our patients.

All of the macular complications quoted above are responsible for poor visual acuity and the poor visual prognosis of Behcet disease. As in our study 60% of patients presented with a BCVA of less than 1/10 which is solely consecutive to macular involvement.

V. CONCLUSION

In conclusion, an array of macular complications, especially macular edema, should be expected in patients with Behcet disease. These macular changes can be responsible for severe, often irreversible loss of vision. Therefore, it is mandatory to provide an early and proper treatment of Behcet syndrome as well as an appropriate follow-up to decrease the risk of visual impairment due to macular involvement.

CONFLICT OF INTEREST

Authors declare that they do not have any conflict of interest.

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