

Isolated Papilledema Revealing Behçet's Disease: Case Report and Literature Review

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Abstract**Case Report**

Neuro-Behçet's disease (NBD) is defined as the primary neurological involvement seen in patients with Behçet's disease (BD). It can be classified as parenchymal or non-parenchymal involvement, with variable manifestation that can be confused with other neuroinflammatory conditions of the nervous system. Papillitis is an inflammation of the optic disk that most often manifests itself with a papilledema. This condition seems to be exceptional during BD, especially as an initial presentation. We report the observation of a neuro-Behçet in a 30-year-old woman who presented with an isolated bilateral papillary edema.

Keywords: Behcet Syndrome, Neuro-Behcet's disease, Papilledema, Optic neuropathy.

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INTRODUCTION

Behçet's disease (BD) is a multisystemic vasculitis of unknown origin, affecting vessels of all calibers, mainly veins, characterized with recurrent oral and genital ulcer as well as uveitis, with possible other systemic involvement. Neuro-Behçet is defined as the primary neurological involvement seen in these patients. It can be due to either vascular or parenchymal damage, and classified as central or peripheral involvement, resulting in a wide spectrum of motor, sensory and psycho-cognitive syndromes. Papillitis is defined as an inflammation of the optic disk, associated with a range of autoimmune and infectious diseases that most often manifests itself with a papilledema. During BD, it is most often associated with bilateral posterior uveitis, but it can also be indicative of the optic nerve involvement in neuro-Behçet [1]. This seems to be exceptional in BD, especially as an initial presentation. We report the observation of a 30-year-old woman with neuro-Behçet revealed by isolated bilateral papillary edema on ophthalmologic examination.

CASE REPORT

A 30-year-old woman presented to the emergency department with occipital headache, vomiting, bilateral decrease of visual acuity (DVA), and

pain on abrupt ocular mobilization one week prior to admission.

She referred a history of recurrent oral ulcers with recurrent episodes of spontaneous resolving DVA. Physical examination on admission revealed two mouth ulcers, Pseudofolliculitis and genital ulcers. Capillary blood glucose and blood pressure were normal.

At admission, visual acuity was 6/10 in the left eye, counting the fingers with the right one. Fundoscopy revealed stage III bilateral papillary edema with blurred margins (Figure 1). Optical coherence tomography was normal. Brain-MRI and blood workup showed no abnormalities. The lumbar puncture revealed normal opening pressure and normal cerebrospinal fluid (CSF) composition.

The patient was diagnosed with bilateral neuropapillitis secondary to BD. She was treated with Intravenous pulse steroid therapy (methylprednisolone 1 g daily for 3 days), followed by oral prednisolone (1 mg/kg/day), and colchicine was initiated.

Within 1 month, the patient's condition gradually improved with the disappearance of headaches, increase of visual acuity and regression of the papillary oedema in the follow-up eye examination.

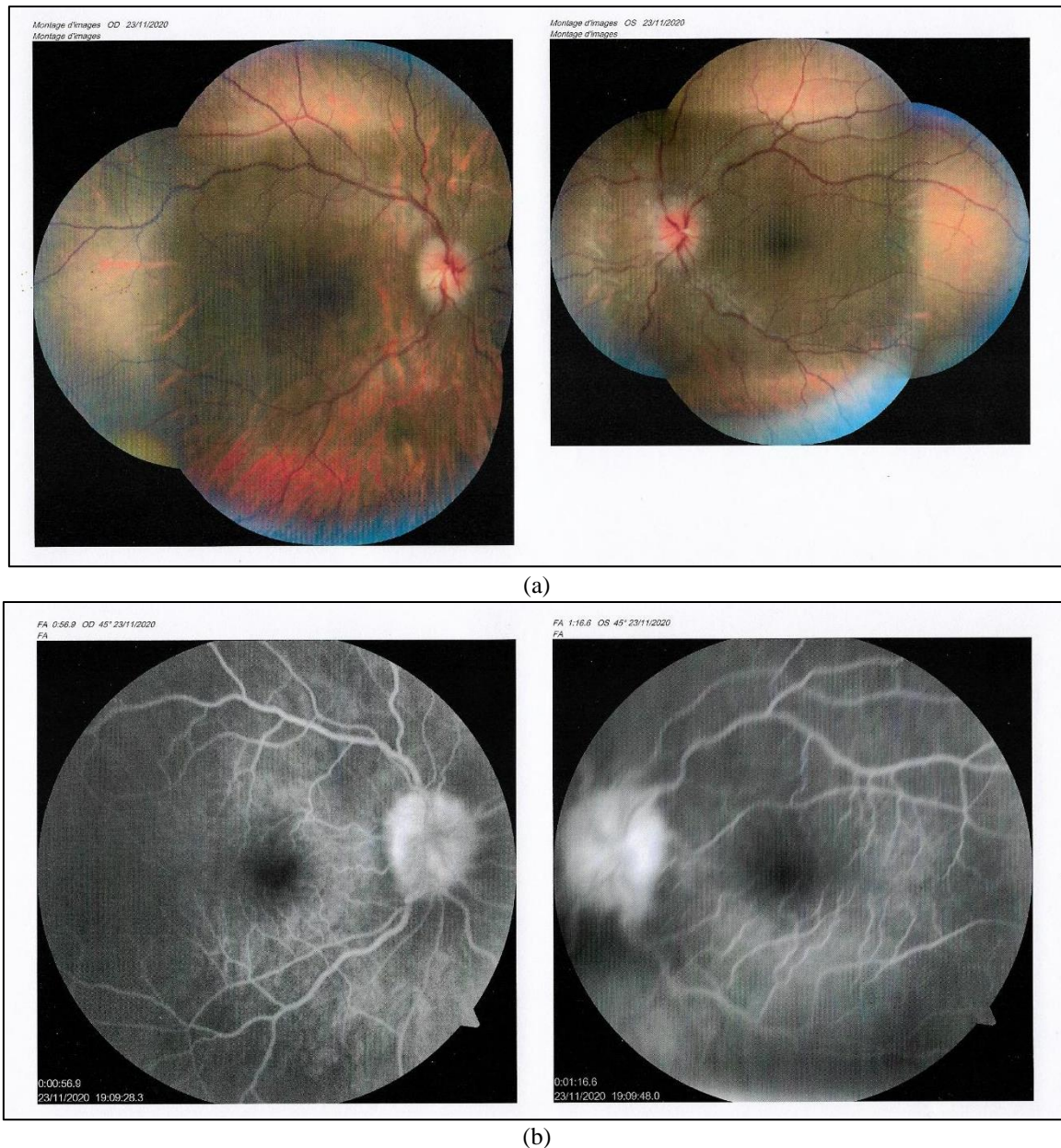


Figure 1: Retinography (a) and fluorescein angiography (b) showing isolated bilateral papilledema with blurred margins in our patient

DISCUSSION

Bilateral neuropapillitis - without the typical signs of intraocular inflammation - has rarely been reported as a manifestation of NBD. It is even more exceptional in its isolated form or as an initial presentation of the disease [2]. The pathogenesis of papillitis in BD remains poorly elucidated, however it is likely that this disease is related to demyelination or axonal necrosis of the optic nerve secondary to recurrent ischemic episodes (vascular involvement), as it may be secondary to an infectious origin given the immunocompromised status of these patients [3]. On the other hand, it would seem that NBD-related damage is associated with the HLA-B51 antigen, much like the Posterior segment involvement or the development of thrombophlebitis [4].

Neuropapillitis usually occurs during systemic flare-ups of the disease [5]. Papillitis may initially manifest itself as a decrease in visual acuity, usually unilateral, visual fog, headaches and retrobulbar pain that may occur during these episodes. An ophthalmologic examination could quantify the decrease in visual acuity and objectify a papilledema which is often associated with posterior ocular damage characteristic of BD. However, cerebral thrombophlebitis and an infection of the central nervous (particularly in immunocompromised patients) must first be excluded in the presence of an isolated and bilateral papilledema especially when intracranial hypertension is suspected [2, 6, 7]. Both of these diagnoses can be ruled out by performing cerebral magnetic resonance imaging (MRI) and lumbar puncture. In these patients, MRI may reveal optic nerve

enhancement, acute optic neuropathy or show no abnormalities. Lumbar puncture in these patients is often normal, however hyperproteinorachia and a predominance of lymphocytes in the cerebrospinal fluid have also been reported [5]. What characterizes our case was the initial presentation of isolated bilateral papilledema, without other inflammatory signs characteristic of the ocular involvement of seen in BD, but also in the absence of intracranial hypertension or cerebral MRI abnormalities that could explain its bilateral nature, leading to the diagnosis of isolated neuropapillosis related to BD.

In general, the treatment of BD includes two components: a flare-up treatment (corticosteroid

therapy) and a background treatment (immunosuppressants and biotherapy). The goal is to control inflammation, reduce the frequency and severity of relapses in order to minimize the sequelae and prevent the progression to blindness. Given the rarity of this condition, there is no consensus regarding the treatment of neuropapillitis in Behçet's disease. Most authors recommend an Intravenous pulse steroid therapy with methylprednisolone 1 g daily for 3 days followed by prednisolone at 1mg/kg/day for a few weeks and gradual tapering (Table 1). This protocol appears to be more effective than oral corticosteroid therapy alone for the control of an acute episode of neuropapillitis [5], however a good response was obtained following this protocol in 1 case [8].

Table 1: Neuropapillitis during Behçet's disease

Authors	Patients	Type of damage	Treatment and evolution
Nanke <i>et al.</i> , [8]	Male 27 years old	Two episodes of isolated papilledema involving the right then left eye	- 45mg/day of prednisolone - Favorable evolution
Shokoohi <i>et al.</i> , [11]	Male, 28 years old	Isolated bilateral papilledema	- Intravenous pulse steroid therapy - Favorable evolution
Tarzi <i>et al.</i> , [2]	Male, 32 years old	Isolated bilateral papilledema	- Intravenous pulse steroid therapy (methylprednisolone) with oral prednisolone combined with Mycophenolate 500mg twice a day - Favorable evolution at 6 months
Berker <i>et al.</i> , [3]	Male 23 years old	Papilledema with blurred margins and retinal vasculitis of the left eye	-Intravenous pulse steroid therapy (methylprednisolone) with oral prednisolone (1 mg/kg/day) for 2 months with progressive decrease. - Favorable evolution within 1 month. - No recurrences over a 6-month follow-up period.
	Male, 33 years old	Severe edema of the optic papilla with blurred margins in the right eye, and disc pallor in the left eye.	- Same treatment - Favorable evolution within 1 month. - recurrent vitritis and iridocyclitis during 1 year of follow-up, treated with local corticosteroid therapy and cyclosporine A (5mg/kg/day) with no recurrence of papillitis
	Female 44 years old	- Severe edema of the optic papilla with blurred margins and occlusive retinal vasculitis of the left eye - Pale papilla of the right eye (former involvement)	- same Treatment - Favorable evolution within 1 month with no recurrences over a follow-up period of 1.5 years.
Yalçındag <i>et al.</i> , [12]	Male, 47 years old	Edema of the optic papilla with blurred margins of the left eye	- High dose of corticosteroids - Favorable evolution at 2 weeks
Erdogan <i>et al.</i> , [9]	Female, 50 years old	Isolated bilateral papilledema	- - Intravenous pulse steroid therapy (1 g/day for 10 days) followed by worsening of visual acuity - Relay by plasma exchanges (5 sessions in total) - Favorable evolution at 3 months

In the case of visual acuity worsening under corticosteroid therapy, plasma exchanges have been proposed as an alternative due to their efficacy in the control of several autoimmune diseases including Behçet's disease. However, given their initial association with high dose corticosteroids in the reported cases, it is difficult to assess their therapeutic effect in these patients [9].

Immunosuppressants are an important mainstay in the treatment of BD, primarily in patients with associated posterior ocular injury or frequent relapses [3]. These disorders call for the prescription of cyclosporin A, infliximab in combination with azathioprine and corticosteroids, or interferon α -2a with or without corticosteroids [10].

CONCLUSION

Neuropapillitis is an exceptional manifestation of NBD, characterized by inflammation and papillary edema which can be bilateral. The presence of the latter requires a cerebral MRI with an LP to rule out sagittal sinus thrombosis or central nervous system infection. The association of papilledema with posterior ocular involvement with decreased visual acuity is quite characteristic, and should alert the examiner to search for BD. The recurrent nature is a major prognostic factor and warrants the association of an immunosuppressive therapy. Only an early diagnosis and adapted management can guarantee a favorable visual outcome.

REFERENCES

- Rougier, M.-B. (2017). Œdème papillaire bilatéral. *Journal Français d'Ophthalmologie*, 40(5), 422-9.
- Tarzi, M. D., Lightman, S., & Longhurst, H. J. (2005). An exacerbation of Behçet's syndrome presenting with bilateral papillitis. *Rheumatology*, 44(7), 953-954.
- Berker, N., Elgin, U., Ozdamar, Y., Acaroglu, G., Soykan, E., & Ozkan, S. S. (2007). Papillitis as the initial presentation of Behçet disease. *Neuro-Ophthalmology*, 31(3), 63-67.
- Sakane, T., Takeno, M., Suzuki, N., & Inaba, G. (1999). Behçet's disease. *New England Journal of Medicine*, 341(17), 1284-1291.
- Voros, G. M., Sandhu, S. S., & Pandit, R. (2006). Acute optic neuropathy in patients with Behçet's disease. *Ophthalmologica*, 220(6), 400-405.
- Kurna, S. A., Altun, A., Cakir, N., & Arsan, A. K. (2018). Neuro-Behçet's disease in a case with papilledema and intracranial hypertension as the initial presentation. *Northern Clinics of Istanbul*, 5(2), 153-156.
- Pamir, M. N., Kansu, T., Erbençi, A., & Zileli, T. (1981). Papilledema in Behçet's syndrome. *Archives of Neurology*, 38(10), 643-645.
- Nanke, Y., Kotake, S., Goto, M., Matsubara, M., & Ujihara, H. (2009). A Japanese case of Behçet's disease complicated by recurrent optic neuropathy involving both eyes: a third case in the English literature. *Modern rheumatology*, 19(3), 334-337.
- Erdoğan, S., Sorgun, M. H., Yalçındağ, N., Atilla, H., & Yücesan, C. (2015). Bilateral optic neuritis in a patient with Behçet's disease who respond to therapeutic plasma exchange. *Saudi Journal of Ophthalmology*, 29(4), 298-300.
- Khairallah, M., Yahia, S. B., Kahloun, R., Khairallah-Ksaa, I., & Messaoud, R. (2012). Œil et maladie de Behçet. *Journal français d'ophtalmologie*, 35(10), 826-837.
- Shokoohi, S., & Yaghoobi, G. (2014, March). A rare case of Behçet's syndrome presenting with bilateral papillitis. In *Seminars in Ophthalmology* (Vol. 29, No. 2, pp. 77-79). Taylor & Francis.
- Yalcindag, N., Yilmaz, N., Tekeli, O., & Özdemir, Ö. (2004). Acute optic neuropathy in Behçet disease. *Eur J Ophthalmol*, 14(6), 578-80.