

Bilateral Synchronous Retrobulbar Optic Neuritis Inaugural of Behçet's Disease

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Abstract

Optic neuritis is exceptionally reported in the course of Behçet's disease. Its frequency is estimated at 0.6 to 4.7% in large series. It is often associated with the neurological involvement of this disease as part of neuro-Behçet.

Optic neuritis complicating Behçet's disease can result from several pathogenic mechanisms: ischemic, inflammatory or stasis lesion and is conventionally present in the form of an acute anterior neuritis. Retrobulbar optic neuritis remains unusual and usually occurs during the course of Behçet's disease.

We report the case of bilateral synchronous and isolated retrobulbar optic neuritis (without neurological impairment or other associated ocular lesions) inaugurating Behçet's disease in a 32-year-old woman.

Keywords: *Retrobulbar Optic Neuritis; Behçet's Disease; Optic Neuritis; Vasculitis*

Introduction

Optic neuritis (ON) is exceptionally reported in the course of Behçet's disease (BD). Its frequency is estimated to be 0.6 to 4.7% in large series: 1/168 patients with ocular involvement for kaçmaz, *et al.* [1], 2/200 patients for Akman-Demir, *et al.* [2] and 18 cases/376 for Frigui, *et al.* [3]. However, it is underestimated because of the frequent existence of uveitis and/or its complications making it difficult to examine the fundus [2,3].

It is often integrated within the framework of a complex neurological involvement of the disease defining the central neuro-Behçet of the parenchymatous type [3,4]. Similarly, its association with other ocular lesions, particularly retinal vasculitis, posterior uveitis, and inflammatory pseudotumors of the orbit is known [3,5].

It usually occurs during the evolution of BD, but inaugural forms are also described [3,6]. This possibility is rare: only 7% of ON were inaugural of BD in the series of Lamari, *et al.* [7]. Retrobulbar optic neuritis (RON) remains unusual, and bilateral involvement is also exceptional.

We report here an original observation of bilateral synchronous and isolated RON (without neurological impairment or other associated ocular lesions) as the first inaugural manifestation of BD.

Case Report

Mrs. B.F., a 32-year-old Tunisian woman with no pathological medical history, consulted the ophthalmology clinic for a significant

decrease in the visual acuity of the two eyes, which had evolved over the last two weeks, associated with intense holocranial headaches. Clinical examination and ophthalmological investigations (measure of visual acuity, fundus of the eye, visual fields, retinal angiography, and visual evoked potential) objectified bilateral retrobulbar optic neuritis. The etiological assessment of this RON performed in ophthalmology (viral serology, bacterial sero-diagnosis, phosphocalcic balance, immunological tests, tumor markers, sectional imaging) was negative; eliminating systemic disease, vasculitis, neoplasia, granulomatosis, viral and bacterial infections.

During her hospitalization in ophthalmology department, the patient developed vulvar and buccal ulcers with diffuse arthralgia requiring her transfer to the department of internal medicine for etiologic diagnosis.

The clinical examination noted bipolar aphthous ulcerations (internal faces of the lips, free edges of the tongue, internal faces of the cheeks, and labium majus). The Pathergy test was positive and the biopsy performed at the injection site showed a leukocytoclastic vasculitis. HLA B51 typing was positive. The biological explorations of the cerebrospinal fluid did not show abnormalities. Cerebro-medullary MRI was normal. Thus the diagnosis of Behçet's disease with bilateral RON was made. The patient was treated with Colchicine®: 1 mg/day, Aspegic®: 100 mg/day and systemic glucocorticosteroids: intravenous methylprednisolone at a dose of 1 g/daily for three days relayed by prednisone at a dose of 1 mg/kg/day for one month followed by a gradual decrease until stopped after 8 months. The evolution was favorable with disappearance of the cutaneo-mucous signs and progressive recovery of a normal visual acuity. Eye examinations and ophthalmological investigations were strictly normal at the three- and six-month follow-ups.

Discussion

Optic neuritis is exceptionally reported during Behçet's disease. It is rarely isolated and often precedes the central nervous system damage: Filali-Ansary reports only two cases in his series of 162 patients (82 patients with ocular lesions and 70 patients with neurological involvement) [8]; similarly Siva reported a frequency of 0.6% (only one case in his series of 164 patients with neuro-Behçet) [9].

The involvement of the optic nerve, isolated or associated with the neuro-Behçet, has been demonstrated both by magnetic resonance imaging [10] and by the post mortem histological examination showing a chronic and diffuse, predominantly lymphocytic inflammatory infiltrate of the neuronal tissue associated with necrosis foci and leukocytoclastic vasculitis of vasa nervorum [11].

ON complicating BD may result from several pathogenic mechanisms: ischemic, inflammatory or stasis lesion [3]. It is classically presented as an acute anterior neuritis (the most frequent: 8/13 cases of ON in the Frigui, *et al.* series) [3,4,7,12]. More rare are retrobulbar optic neuritis [1]. In some cases, the diagnosis of these ON is carried out only at the late stage of sequellar optic atrophy, where it is difficult to specify the type or the mechanism [3].

This RON may be uni or bilateral, acute or chronic, regressive or recurrent, and can precede the meningoencephalitis by several months [13].

ON associated to BD respond habitually to high doses of systemic steroids with or without additional immunosuppression, particularly cyclosporine [14]. Rarely biotherapy or therapeutic plasma exchange can be used for severe and resistant forms [15].

Its prognosis remains unfavourable even after the appropriate treatment: final visual acuity of 1/10th in 25% of the patients treated in the series of Frigui, *et al.* with blindness in 1/3 of the cases [3]. An early diagnosis is important especially for acute ON which may be transient and reversible under corticosteroid therapy [4] as was the case for our patient. This indicates the optic vasculitis: acute vasculitis of the arterioles of the optical disc [15].

Our case is distinguished by its inaugural, bilateral synchronous and isolated presentation (not associated with neurological damage or other specific ocular lesions of the disease).

- Indeed, the epidemiological study of Kneifel, *et al.* demonstrated that 56% of patients with BD developed ocular complications, and only for 8.6% of them ocular involvement was the first manifestation inaugurating the disease [17].
- Kidd DP, in a recent review published in 2013, noted that 4/20 cases of BD with inflammatory optic neuropathy had bilateral and synchronous involvement [14], and to the best of our knowledge, only six cases of bilateral and synchronous optic neuropathy in BD had been reported previously [14,15].

Conclusion

The RON is very exceptional during BD. It should be systematically sought to improve the prognosis of the disease as it often indicates the central neurological involvement. On the other hand the BD must be suspected in front of RON in young subjects. Our case is distinguished by its inaugural, bilateral and isolated presentation (not associated with neurological damage or other specific ocular lesions of the disease).

Conflicts of Interest

No conflicts of interest.

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