Bilateral and Simultaneous Retrobulbar Optic Neuritis Revealing Multiple Sclerosis

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Abstract

Optic neuritis is a common manifestation of multiple sclerosis. It occurs in two thirds of patients at some point in the course of this disease and is usually unilateral, acute and often recurrent. However, optic neuritis can be the first manifestation of this demyelinating disease in 15 to 20% of cases.

Bilateral, simultaneous and retrobulbar forms of optic neuritis inaugurating multiple sclerosis remain exceptional and unusual. They represent a real diagnostic challenge and require special attention from the clinician.

Herein we report the case of bilateral simultaneous and isolated retrobulbar optic neuritis inaugurating multiple sclerosis in a 24 year old woman.

Introduction

Optic neuritis is a relatively common reason for consultation in routine medical practice [1]. Its age-adjusted incidence in the Nordic countries is estimated at 2.2 to 2.5 per 100,000 [2]. It is usually acute, unilateral and often recurrent [3]. The etiological diagnosis of this optic neuropathy represents a real challenge for the clinician given the multitude of possible causes. Among the demyelinating neuropathies most frequently involved are multiple sclerosis, neuromyelitis optica (previously known as Devic’s disease), neuromyelitis optica spectrum disorders (NMOSD, specifically associated with anti-aquaporin-4 antibodies) and anti-myelin oligodendrocyte glycoprotein (MOG) autoantibodies-related demyelinating diseases [1,4]. The so-called “idiopathic” optical neuritis has become increasingly rare since the detection of these specific entities associated with the auto-antibodies mentioned above [4].

Multiple sclerosis is the most common cause in typical forms of optic neuritis [1]. Indeed, in the large cohort of 1844 patients of Confavreux C and Vukusic S with confirmed multiple sclerosis; isolated optic neuritis was the first symptom of the disease in 335 patients (18%) [5].

However, bilateral, simultaneous and revealing presentations of the disease remain exceptional and unusual [3,4]. We report an observation.

Case Report

Ms. A.N., a 24 year old Tunisian woman with no significant pathological history consulted the ophthalmology clinic for a significant decrease in the visual acuity of the two eyes evolving for a week. The physical examination was without anomalies as well as the eye examination by the slit-lamp. In particular, there was no evidence of eye redness or eye pain (spontaneous or caused by the mobilization of the eyeball). The examination of the fundus of the eye showed a bilateral papillitis. Specific ophthalmological explorations (visual fields, retinal angiography and visual evoked potential) confirmed the diagnosis of acute and bilateral retrobulbar optic neuritis of inflammatory nature. The patient was transferred to the internal medicine department for etiologic assessment of this acute optic neuritis.

Physical examination showed no signs suggestive of connective tissue disease, primary vasculitis or systemic granulomatosis. Neurological signs and abnormalities of osteotendinous reflexes were not noted.

Routine biological tests were within the normal range: hemogram, erythrocyte sedimentation rate, C-reactive protein, serum protein electrophoresis, blood glucose, creatinine, ionogram, transaminases, lactate dehydrogenases, creatinine phosphokinase, alkaline phosphatases, calcium-phosphate balance and Lipid parameters.

Viral serologies (Epstein-Barr virus, HIV, Cytomegalovirus, Herpes, Measles, Mumps and Varicella-Zoster virus) were negative or showed an old immunization profile.

The immunological tests were also negative (anti-nuclear antibodies, native anti-DNA antibodies, anti-soluble antigens antibodies, anti-cardiolipin and anti-β2GP1 antibodies).

Chest X-ray showed no abnormalities and the angiotensin-converting enzyme rate was normal.

Lumbar puncture demonstrated a clear cerebrospinal fluid with normal pressure. Biochemical analysis showed normal glycocardichia and moderately high protein level at 0.65 g/l. The cytological study showed pleocytosis with 18 cells/mm³ predominantly lymphocytic. Direct bacteriological examination and culture were negative.
Conventional magnetic resonance imaging (MRI) with T1-weighted, T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences was performed. T2-weighted and FLAIR images revealed multiple hyperintense lesions in the centrum semiovale and periventricular white matter of both hemispheres with moderate global cortico-subcortical atrophy (Figures 1 and 2). After gadolinium injection, lesions showed punctuate enhancement on T1-weighted sequences suggesting acute MS lesions. MRI of the cervical and dorsal spine was without abnormalities.

At the end of these explorations, the diagnosis of a bilateral inflammatory retrobulbar optic neuritis revealing a multiple sclerosis was retained. The patient was treated with methylprednisolone in intravenous bolus at a dose of 1000 mg/day for five days and then relayed by prednisone at a dose of 1 mg/kg/day with a favorable evolution.

**Discussion**

Multiple sclerosis is the most frequent demyelinating central neuropathy with an estimated global incidence of 3.6/100,000 for women and 2/100,000 for men [6,7]. Tunisia is currently qualified as an area of average prevalence for multiple sclerosis with an incidence of 1.34/100,000 and the disease is characterized by a monosymptomatic initial presentation in the majority of cases (96% of cases) [8,9].

Optic neuritis, classically acute, unilateral, and typically recurrent, is the most common clinical manifestation associated with multiple sclerosis in both adult and pediatric forms [10]. It is seen in 2/3 of patients at some point in the course of the disease [7].

However, this optic neuropathy may be the initial symptom of the disease in approximately 15 to 20% of cases [11]. In the Tunisian series of Sidhom et al. 20% of adult patients with multiple sclerosis began their disease with isolated optic neuritis [9].

The risk of secondary transformation to multiple sclerosis of an isolated and apparently “idiopathic” optic neuritis is estimated at 10.8%; this risk is somewhat higher for women compared to men: 13.9% versus 7.7% [12]. The period between optic neuritis and the emergence of other specific clinical signs of multiple sclerosis varies from a few days to several years [12,13].

In the more recent series and due to the recognition of new specific entities (neuromyelitis optica spectrum disorder and anti-myelin oligodendrocyte glycoprotein autoantibody-related demyelinating diseases) and their association with specific auto-antibodies (anti-aquaporin 4 and anti-MOG); the transformation of initially isolated optic neuritis to multiple sclerosis becomes significantly less, particularly in subjects negative for anti-aquaporin 4-autoantibodies: only four patients/83 (4.8%) after a five year follow-up in the Zhou et al. series [3].

Some factors are significantly predictors of secondary evolution of optic neuritis to multiple sclerosis (p<0.05), in particular: female sex, retrobulbar type of optic neuropathy, recurrence, elevation of the IgG index in cerebrospinal fluid and the existence of signal abnormalities in cerebral imaging [14]. The two year cumulative probability of secondary transformation from optic neuritis to multiple sclerosis is estimated to be 5.92% and that of five years to 14.28% [14].

Acute bilateral and simultaneous retrobulbar optic neuritis, as in our case, can exceptionally be the inaugural symptom of multiple sclerosis in the adult [13] and child (7-9%) [15]. This possibility remains unusual and often unrecognized, requiring special attention: in fact only two patients with bilateral and simultaneous retrobulbar optic neuritis subsequently evolved into multiple sclerosis in the series of Parkin et al. [13].
These forms require a complete assessment, special investigations as well as prolonged monitoring to diagnose the transformation into multiple sclerosis in time. Currently, optic coherence tomography (OCT) may play a determining role in the diagnosis of possible multiple sclerosis underlying optical neuropathy [16]. This exam is a reliable indicator of axonal loss in the central nervous system [1].

Therefore, recent experimental studies have shown a positive and significant correlation between low levels of sPECAM (Platelet-Endothelial-Cell-Adhesion-Molecule-1) and sVCAM-1 (Human-Vascular-CAM-1) and secondary progression of isolated optic neuritis to multiple sclerosis. This correlation was significant for patients with optic neuritis with positive anti-aquaporin 4 IgG (AQP4-IgG) antibodies as well as those with optic neuritis and negative AQP4-IgG [17]. These molecules (sPECAM-1 and sVCAM-1) could thus be useful biomarkers for the prediction of a progression from initially isolated optic neuritis to multiple sclerosis [17].

Such a presentation may further discuss the diagnosis of Leber’s hereditary optic neuropathy (LHON); a mitochondrial disorder which is one of the most common inherited cause of bilateral painless optic neuropathy and blindness occurring in young adults and predominantly in males [18,19]. It’s due to mutation in the mitochondrial genome (mtDNA) and occasionally associated with neurological, cardiac, and skeletal changes [18,19]. This disease may have a clinical features that mimics a multiple sclerosis (sclerosis-like syndrome) [20] or be associated with an authentic multiple sclerosis defining the “Harding’s syndrome” making the diagnosis even more challenging for the clinician [21,22]. Moreover, LHON can mimic not only MS, but also neuromyelitis optica [23] or Susac syndrome [19]. In these cases, mtDNA mutation screening is important to confirm the diagnosis of LHON.

Conclusion

Acute bilateral and simultaneous retrobulbar optic neuritis remains an exceptional and unusual presentation of multiple sclerosis. This possibility should be kept in mind by the clinician to ensure early diagnosis and treatment of the disease in order to preserve the visual prognosis. New imaging techniques (especially OCT) as well as some specific biological markers are of great benefit in this diagnostic approach. Regular and sometime prolonged monitoring is also recommended because the timeframe before the emergence of other clinical signs of multiple sclerosis may be too long.

References