

Case Report

Multiphasic Neuromyelitis Optica and Life-threatening: A Case Report

Patrice Ntenga¹, Salaheddine Mourabit¹, Soumaila Boubacar¹, Ousmane Cissé¹,
Mouhamed Lelouma Mansare¹, Kamadore Touré^{1,2}, Ndiaye Moustapha¹,
Mouhamadou Mansour Ndiaye¹

¹Department of Neurology, Fann National Teaching Hospital, Dakar, Senegal

²Department of Preventive Medicine and Public Health, Faculty of Medicine, Pharmacy and Odontostomatology, University of Cheikh Anta Diop, Dakar, Senegal

Email address:

jacobntenga@gmail.com (P. Ntenga)

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Abstract: Neuromyelitisoptica is an inflammatory and demyelinating disease of the central nervous system that affects astrocytes in the optic nerve and the spinal cord. It is characterized by outbreaks of transverse myelitis and retro bulbar optic neuropathy with a pejorative aspect in terms of prognosis and prognosis in the short and medium term. We report the case of A S, 32 years old, male, Senegalese living in Dakar, hospitalized in June 2016 at the neurological clinic of the CHU of FANN, Dakar-Senegal for aneuromyelitisoptica. Magnetic resonance imaging showed cervical myelitis extended to the thoracic cord. The search for antibodies to aquaporin 4 was positive, the visual evoked potential showed P100 latency. Lumbar puncture performed, showed a protein content to 0.73 g/l and 10 elements (cells). The patient was placed under corticosteroid therapy. His clinical picture was stationary in the short term, with a recovery of the walk at 5 months of the appointment.

Keywords: Multiphasic, Neuromyelitis, Optica, Life-threatening

1. Introduction

Optic neuromyelitis (NMO) or Devic's disease is an inflammatory and demyelinating disease of the central nervous system (CNS) that affects astrocytes in the optic nerve and spinal cord. It is characterized by transverse myelitis and retro bulbar optic neuropathy (NORB), which leads to limb paralysis and blindness [1]. It is a rare disease with an unequal global distribution and a prevalence of 1-2 / 100,000 which varies from 0.51 per 100.000 in Cuba to 4.4 in southern Denmark, 0.12 / 100.000 in Ghana, 5/100.000 inhabitants in South Africa, and 0.05 / 100.000 inhabitants in Kenya [2]. This pathology is usually pejorative in terms of prognosis in the short and medium term [3].

Central nervous system (CNS) lesion analysis supports a humoral mediated immune-mediated mechanism due to complement activation, eosinophilicpolynuclear infiltration

and vascular fibrosis. In patients with NMO, loss of astrocytes was observed in lesions in the spinal cord and optic nerves. There are very few data on brain involvement in pathological examination [4]. Of the few cases analyzed, macrophage infiltration and infiltration of eosinophilicpolynuclear cells have been observed in the lesions as in the spinal cord. Magnetic resonance imaging is usually normal at the onset of the disease [4].

We report a case of multiphasic optic neuromyelitis in a young adult admitted to the Neurological Clinic of the University Hospital Center of FANN.

2. Medical Observation

Mr. AS 32 year old, living in Dakar, Senegal. Single, tailor, received for a motor deficit of the 4 limbs in June 2016. This table had evolved for 8 months by relapses and which had begun with right cervico-brachial neuralgia followed two days

later by a homolateral hemiplegia. This table will evolve for 4 months with a partial recovery on the motor plane. Then there was motor deficit on the contralateral side, accompanied by urinary incontinence and constipation. His antecedents indicated a decrease in left monocular vision, which had regressed without treatment. The clinical examination revealed a preservation of superior functions, a four-paresis, a bilateral Babinski, a C6 (sixth cervical vertebra) sensory level, left eye diplopia, urinary incontinence and constipation. In front of this table, medullary magnetic resonance imaging (MRI) was performed and revealed a stepped cervical spinal hyper signal with a large marrow aspect (Figure 1). Brain MRI showed no signal abnormalities. The search for autoantibodies to aquaporin 4 was positive. A lumbar puncture showed a sugar level of 0.4 g/l, proteins at 0.73 g/l, 10 cells (lymphocytes), culture and identification of germs were negative after 48 hours. A visual evoked potential (VEP), showed an elongation of latency P100. The diagnosis of an NMO was made according to the criteria of Wingerchuk DM *et al.* [5]. The patient had been given corticosteroid therapy (prednisone at a dose of 1mg/kg/day and an adjuvant treatment of calcium, potassium). Its short-term evolution was stationary on the motor plane.

3. Discussion

Most authors agree that NMO, an inflammatory disease of the central nervous system (CNS), is a rare disease globally and varies geographically [2]. In our environments, low prevalence is multifactorial, in particular the difficulty of measuring the antibodies to aquaporin 4 (AQP4) on the one hand, and the low socioeconomic level on the other hand, which limits the accessibility to the MRI for many patients.

Our observation concerned a man. However, the literature shows that in 80% of cases, this pathology favors the female sex, as shown by the studies of Samy and *al.* [6], J Seze and *al.* in French Caribbean [7, 8]. It is important to ask why this female predisposition, broader studies to determine relative risk would help to determine whether gender is a risk factor for the NMO. For Bernard-Valnet and *al.* [9], the sex ratio is variable according to the studies and especially according to the serological status of the patients. It is estimated to be greater than 5F/1M in seropositive patients and close to 1 in seronegative patients.

Studies by Bernard-Valnet and *al.* [9] demonstrated that the median age of NMO declaration was around 40 years, although it had early pediatric or early-onset forms after 80 years. Initially described as monophasic, it would appear that most NMO patients develop outbreaks if followed long enough. The frequency of these outbreaks is however difficult to estimate because it depends on the anti-AQP4 status.

The age of our patient is in the age range of some studies which stipulate that the average age of onset of the disease is around 30.9 years according to Cabre (10), 34.4 years for Samy [6] and 40 years for J Seze [7].

According to Déral-Stéphant and *al.* [11], the disease is characterized clinically by one or more episodes of optic

neuritis and myelitis. It evolves either in a monophasic form with a delay between optic neuritis and myelitis which is less than a month, or in a relapsed form. For this same study, it was shown that the average delay between the episode of optic neuropathy and myelitis was 166 days up to 730 days. In addition, it was shown the rarity or absence in the literature of the described case where optic neuritis and myelitis were spaced ten years.

Clinically the onset of the disease was marked by visual disturbances in our patient. A relative equilibrium between the optic, medullary and mixed beginnings was noted for some authors [11, 12]. On the other hand, Déral-Stéphant [11] had shown in his studies that the mode of onset of the disease in 72% of cases was marked by an optic neuritis which could be bilateral, but in the majority of cases it was unilateral and acute with severe prognosis.

The frequency of outbreaks is however difficult to estimate because it depends on the anti-AQP4 status. It was revealed by these cohort studies that the inaugural chart would seem to be associated with the age of onset of the disease. Before age 50, it is more common to start a NMO by a retro bulbar optic neuropathy outbreak while later forms more readily start with an episode of myelitis. This would explain the more rapid evolution towards an irreversible motor handicap of later onset forms. [9]

In a large literature, medullary MRI showed in more than 70% of cases a lesion extended to several metamers producing a clinical picture of extensive transverse myelopathy, as is the case in our patient and as also shown in studies of S. Bourmatte and *al.* [3], where MRI showed an extended signal abnormality on more than three vertebral segments in all patients.

The positivity of anti NMO antibodies in our patient reinforced the diagnosis. However, it was reported that it is a non-mandatory criterion because of its sensitivity which was only 50-70% according to the studies. [4] and therefore it is important only if it contribute to the diagnosis. [9] The classical and historical method of detection of antibodies specific to NMO, initially called NMO-IgGC, has several limitations. The main problem comes from its relatively low sensitivity. According to the published works, this sensitivity varies from 38 to 87%. The second problem is the subjective character which would make this test very dependent on the operator. This would promote a risk of false positivity, especially in the presence of other autoantibodies in the patient's serum, including anti-nuclear antibodies.

Some authors believe that the positivity of these antibodies is considered to be an element of poor prognosis because there is a strong positive correlation between the serum concentration of these antibodies and the severity of the disease as well as the degree of disability [13].

In our patient's magnetic resonance imaging (MRI) brain was normal. However, NMO tables with brain lesions on MRI, symptomatic or not, have been reported, mainly in Asian cohorts [9]. The most characteristic are clinico-radiological panels of acute disseminated encephalomyelitis. This raises the question of the search for anti-AQP4 in front of this clinical picture with specific brain lesions of the NMO. In

some rare cases, preferred MRI lesions in the hypothalamus has been described, with associated neurological symptoms such as hypersomnia or eating disorder [9;14].

The life expectancy in the acute and short-term phase in our patient was good, however, it is reported in some series that the acute phase mortality rate could reach 20%, especially when considering monophasic forms (Optic neuropathy and myelitis occurring at the same time without further thrust). The latter appear to have a darker prognosis in the short term, in particular because of the increased risk of death from associated dysautonomic impairment (cardiorespiratory arrest, major hypotension) [4].

Our patient had not recovered on the motor plane during his hospitalization, this corroborates the literature which shows that the long-term disabilities were more marked in the remittent forms [3].

4. Conclusion

Epidemiological studies on NMO are still rare and knowledge about the epidemiology of NMO in many parts of the world remains extremely limited. Without appropriate treatment, the development is rapidly pejorative and leads to blindness and / or a major motor handicap for half the patients after 5 years of evolution on average. Multiphasic forms are better prognostic for the patient compared to monophasic forms. This pathology of unknown etiology still poses problems on the functional and mental development of the patient, recurrences are possible making the person a socio-familial and economic charge.

Iconography



Figure 1. Medullary MRI, sagittal section, T2sequence, showing extensive hypersignal from the cervical cord to the thoracic cord.

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