Scientific Research Publishing

Open Journal of Pediatrics, 2023, 13, 432-436
https://www.scirp.org/journal/ojped
ISSN Online: 2160-8776
ISSN Print: 2160-8741

Neuromyelitis Optica Revealed by Headache in a Child: A Case Report

Amal Hamami*, Maria Rkain, Madiha Benhachem, Ayyad Ghannam, Aziza Elouali, Abdeladim Babakhoua

Department of Pediatrics, Mohamed VI University Hospital Center, Mohamed 1 University Oujda, Oujda, Morocco
Email: *hamami.amal16@gmail.com

How to cite this paper: Hamami, A., Rkain, M., Benhachem, M., Ghannam, A., Elouali, A. and Babakhoua, A. (2023) Neuromyelitis Optica Revealed by Headache in a Child: A Case Report. *Open Journal of Pediatrics*, **13**, 432-436.
https://doi.org/10.4236/ojped.2023.133048

Received: April 10, 2023
Accepted: May 26, 2023
Published: May 29, 2023

Abstract

Devic's neuromyelitis optica (NMO) is a rare inflammatory disease of the central nervous system that results in optic neuropathy and myelitis. Optic neuritis represents the mode of entry into the disease in more than two thirds of cases. It is a rare entity in children. There is no effective treatment at present, but some molecules can be used, such as corticosteroids, immunosuppressants and plasma exchange. The prognosis in children is generally favorable. Devic's neuromyelitis is a condition of unknown etiopathogeny which is functionally critical and requires early and appropriate treatment. We report the case of a 12-year-old girl who presented to emergency with a headache and decreased visual acuity, whose investigations led to the diagnosis of Devic's neuromyelitis optica.

Keywords

Neuromyelitis Optica, Devic Syndrome, Child

1. Introduction

Devic's neuromyelitis optica (NMO) or Devic's syndrome combines transverse myelitis and unilateral (9%) or bilateral (91%) optic neuropathy occurring 8 weeks apart. It is often preceded by a non-specific viral syndrome but may also be associated with varicella, infectious mononucleosis, tuberculosis, rubella vaccination or lupus. Many authors consider it an exceptional manifestation of multiple sclerosis (MS) or infectious encephalomyelitis. It is often reported in adults, rarely in children population [1], in whom it is a distinct clinical entity with an excellent visual and neurological prognosis. We report the case of a 12-year-old girl who presented to emergency with a headache and decreased visual acuity, whose investigations led to the diagnosis of Devic's neuromyelitis optica.

2. Observation

We report the case of a 12-year-old girl who presented to emergency with a headache and decreased visual acuity for one week. The clinical examination found meningeal stiffness and a quadri pyramidal syndrome.

The blood tests showed:

CBC: WBC 18,040/mm^3 predominantly PNN; Hb 14 g/dl; Platelets 465,000 elements/mm^3. CRP: 5.5 mg/l; Sedimentation speed: 30 mm/s; Procalcitonin negative. EPP: inflammatory syndrome with alpha1, alpha2 and elevated gamma globulin. Renal, hepatic and haemostasis tests were normal.

An ophthalmological examination was carried out, which showed a decrease in visual acuity to 2/10 for left eye and 2/10 for right eye, bilateral papilledema on the fundus and bilateral dry eye as confirmed by the Schirmer test.

Brain CT scan ray returned normal. A lumbar puncture was performed and an intracranial pressure of 15 to 19 mmHg was noted. Cerebrospinal fluid was found to be 155 cells/mm^3 and the culture was sterile.

Brain MRI showed infiltration of the left optic nerve (**Figure 1**), while spinal cord MRI showed extensive cervical hypersignal from C1 to C5 consistent with acute demyelinating disease (**Figure 2**).

Devic's syndrome was evoked in front of the association of optic neuritis and acute transverse myelitis. Serologies for Lyme disease, syphilis and HIV were negative. The tuberculin TST and the BK test in the sputum and gastric tube were negative. Anti-NMO antibodies were negative, anti-nuclear antibodies were positive. Devic's syndrome associated with a dry syndrome was suspected due to the presence of bilateral dry eyes as confirmed by the Schirmer test.

The patient was put on a three-day injectable bolus of methylprednisolone followed by long-term oral treatment with prednisone 2 mg/kg/day and azathioprine (disappearance of headache, progressive regression of papilledema and recovery of visual acuity), the patient is actually followed up in consultation with good clinical evolution thereafter.

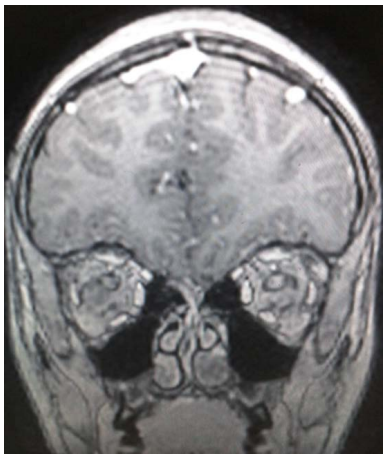

Figure 1. T1-weighted thin slice MRI of the brain with gadolinium injection showing infiltration of the left optic nerve (in favour of left optic neuritis).

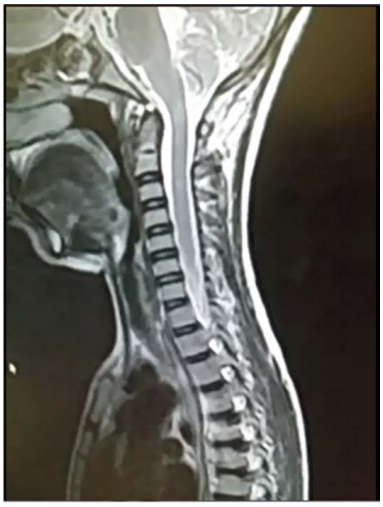

Figure 2. T2-weighted sagittal MRI of the spinal cord showing extensive hypersignal from C1 to C5.

3. Discussion

Neuromyelitis optica or "Devic's syndrome" is an autoimmune demyelinating disorder that combines:

- Unilateral or bilateral optic neuritis.
- Acute transverse myelitis.
- In most cases, optic damage precedes spinal cord damage by a few days to 3 months.

This rare condition can be seen at any age (from 4 to 73 years). However, it appears to be rarer in children population.It is more common in Japan and East Asia, but does not exceed a prevalence of 5 cases/100,000 population [1]. It is thought to represent about 1% of all bone marrow inflammatory diseases [2]. The average age of onset of monophasic forms is 28 years, while the relapsing form occurs later at an average age of 41 years.

The diagnosis is made according to the Wingerchuk criteria [3] which include absolute criteria (acute myelopathy, absence of neurological involvement other than optic neuritis and myelitis), major associated criteria (normal magnetic resonance imaging at onset, hyper signal spread over 3 or more vertebral segments, hyperleucorrhachia > 50 WBC/mm^3) and minor associated criteria (bilateral optic neuritis, severe optic neuritis with visual acuity < 20/200, severe motor deficit in at least one limb < 2/5).

In 70% of cases, positive anti-NMO antibodies are found, which are specific for neuromyelitis optica. Their presence is a predictive factor for recurrence/exacerbation.

The etiology of NMO remains unclear. This clinical picture has been described after infectious episodes, mainly viral (EBV, CMV, HSV-1 and 2, varicella-zoster, rubella, HIV…), bacterial (tuberculosis, mycoplasma, syphilis), in the post-vaccination period (flu vaccine) and in association with many autoimmune diseases such as hypothyroidism, Gougerot-Sjogren's syndrome or sys-

temic lupus erythematosus. Some authors have even suggested that NMO itself is a true autoimmune disease [1].

The association of primary Gougerot-Sjögren's syndrome (PGS) with genuine Devic syndrome has been reported in most series of NMO. In the series by Wingerchuk *et al.* [3], NMO was associated with autoimmune disease in 15 cases (30%) of which four cases were PGS. In the series by De Sèze *et al.* [4], five cases had associated autoimmune disease (38%), one of which was lupus and four of which were PGS. Conversely, in a series of myelopathies associated with PGS, four cases had a typical NMO picture [5].

The time between the onset of neurological signs and the diagnosis of PGS can be several years. For example, Gökcay *et al.* [6] reported a twenty-year-old girl who had been followed for Devic syndrome since the age of 10 years, but was not diagnosed with associated PGS until 10 years later. In the series by De Sèze *et al.* [7], Devic syndrome preceded the diagnosis of PGS in all four cases with delays ranging from two to 14 years.

In practice, the combination with a PGS does not change the "classical" treatment of Devic syndrome. Corticosteroids bolus for three to five days are used during relapses, and plasma exchange has also been proposed [5] [8]. To prevent relapses, high-dose oral corticosteroids combined with immunosuppressive therapy are indicated; the combination of prednisone and azathioprine and the use of an intravenous anti-lymphocyte B monoclonal antibody (rituximab) have led to a significant reduction in the frequency of relapses, but with an insufficient follow-up time of 18 and 12 months respectively [9] [10]. Cyclophosphamide is used in monthly courses for one year by some authors [5] [11].

Generally, the presence of an autoimmune disease associated with Devic syndrome is a predictor of relapse.

4. Conclusion

NMO in children is rare. It is a distinct clinical entitý with an excellent visual and neurological prognosis. Recurrence is rare and appears to be without long-term sequelae. In contrast, in adults, the disease can be fatal and often leaves ophthalmological and neurological sequelae. Despite its rarity, GSS should be part of the etiological work-up of a Devic syndrome even in the absence of a previously known dry syndrome.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Goetz, C.G. (1999) Demyelinating Disorders of the Central Nervous System (Chapter 48). In: Hogancamp, W.E. and Noseworthy, J.H., Eds., *Textbook of Clinical Neurology*, 970-986.

[2] Jouhadi, Z., Ouazzani, I., Abid, A., El Moutawakil, B., Rafai, M.A. and Slassi, I.

(2004) Neuromyelitis optica de Devic and viral hepatitis AA propos d'une observation pédiatrique. *Revue Neurologique*, **160**, 1198-1120. https://doi.org/10.1016/S0035-3787(04)71167-4

[3] Wingerchuk, D.M., Lennon, V.A., Pittock, S.J., Luccinetti, C.F. and Weinshenker, B.G. (2006) Revised Diagnosis for Neuromyelitisoptica. *American Journal of Ophthalmology*, **142**, 715-716. https://doi.org/10.1016/j.ajo.2006.08.010

[4] Lafitte, C. (2002) Neurological Manifestations of Primary Sjögren's Syndrome. *Revue Neurologique*, **10**, 959-965.

[5] De Seze, J., Stojkovic, T. and Hachulla, E. (2001) Myelopathies and Gougerot-Sjögren Syndrome: Clinical, Radiological and Evolutionary Profile. *Revue Neurologique*, **157**, 669-678.

[6] Gökcay, F., Celebisoy, N., Gökcay, A., Kabasakal, Y. and Oder, G. (2007) Primary Sjogren's Syndrome Presenting as Neuromyelitis Optica. *Pediatric Neurology*, **36**, 58-60. https://doi.org/10.1016/j.pediatrneurol.2006.07.008

[7] De Seze, J., Stojkovic, T., Ferriby, D., Gauvrit, J.Y., Montagne, C. and Mounier-Vehier, F. (2002) Devic's Neuromyelitis Optica: Clinical, Laboratory, MRI and Outcome Profile. *Journal of Neuroscience*, **197**, 57-61. https://doi.org/10.1016/S0022-510X(02)00043-6

[8] Mochizuki, A., Hayashi, A., Hisahara, S. and Shoji, S. (2000) Steroid-Responsive Devic's Variantin Sjögren's Syndrome. *Neurology*, **54**, 1391-1392. https://doi.org/10.1212/WNL.54.6.1391

[9] Mandler, R., Ahmed, W. and Dencoff, J. (1998) Devic's Neuromyelitis Optica: A Prospective Study of Seven Patients Treated with Prednisone and Azathioprine. *Neurology*, **51**, 1219-1220. https://doi.org/10.1212/WNL.51.4.1219

[10] Cree, B., Lamb, S., Morgan, K., Chen, A., Waubant, E. and Genain, C. (2005) An Open Label Study of the Effects of Rituximab in Neuromyelitisoptica. *Neurology*, **64**, 1270-1272. https://doi.org/10.1212/01.WNL.0000159399.81861.D5

[11] Williams, C.S., Butler, E. and Roman, G.C. (2001) Treatment of Myelopathy in Sjögren's Syndrome with a Combination of Prednisone and Cyclophosphamide. *Archives of Neurology*, **58**, 815-819. https://doi.org/10.1001/archneur.58.5.815