Annals of Case Reports & Reviews

Case Report

doi: 10.39127/2574-5747/ACRR:1000223
Zouine M, et al. Annal Cas Rep Rev: ACRR-223

Seronegative Neuromyelitis Optica in Children: A Case Report

M. Zouine[13], S. Moussaoui[1,3], G. Draiss[1,3], N. Rada[1,3], M. Bouskraoui[1,3], S. Zouita[2,3], D. Besraoui[2,3], D. Jalal[2,3]

[1]Service de pédiatrie A, hôpital mère enfant, CHU Mohammed VI Marrakech
[2]Service de radiologie, hôpital mère enfant, CHU Mohamed VI Marrakech
[3]Faculté de médecine et de pharmacie Marrakech

***Coressponding author:** Mouna Zouine, Service de pédiatrie A, hôpital mère enfant, CHU Mohammed VI Marrakech. Email: mounazouine@gmail.com

Citation: Zouine M, Moussaoui S, Draiss G, Rada N, Bouskraoui M, et al. (2021) Seronegative Neuromyelitis Optica in Children: A Case Report. Annal Cas Rep Rev: ACRR-223.

Received Date: 09 April 2021; **Accepted Date:** 14 April 2021; **Published Date:** 20 April 2021

Abstract

Optic neuromyelitis, also called Devic's disease, is an inflammatory, demyelinating, rare and aggressive disease of the central nervous system that preferentially affects the optic nerves and the spinal cord, simultaneously or consecutively, with a monophasic or recurrent course.

***Observation:** we report the observation of a 13-year-old girl, with no particular pathological history, admitted for flaccid paraplegia with urinary retention and sudden onset blindness. The visual evoked potential showed an increase in lag time in relation to retrobulbar optic neuritis. Spinal magnetic resonance imaging (MRI) has shown an aspect of extensive myelitis. The dosage of anti-AQP4 Ab was normal. Devic syndrome was retained in this patient before the association of myelitis and retrobulbar optic neuritis. The evolution under treatment with a bolus of methylprednisolone at a dose of 1 g / 1.73 m2 per day for 5 days, relayed by prednisone at a dose of 2 mg / kg per day, was marked by the recovery of visual function with persistent functional impairment.*

***Conclusion:** Devic's NMO occurring in children is rare; it is a distinct clinical entity with a prognosis which can sometimes be reserved.*

Introduction

Optic neuromyelitis, also known as Devic disease or Devic syndrome, is a rare condition in children. It is characterized by acute or subacute optic neuritis and transverse myelitis. In the pediatric population, only a few cases of optic neuromyelitis (NMO) have been reported so far [1]. Here we report a case of NMO in a 13-year-old girl with bilateral optic neuritis with brain and spinal cord injury.

Reported case

It is a 13-year-old girl, with no particular pathological history, who developed 3 weeks before her hospitalization of paresthesia of the lower limbs, which rapidly developed into flaccid paraplegia of the lower limbs with sphincter-like disorders of the vesical retention. The neurological examination showed a motor deficit in the lower limbs with a deficit in surface sensitivity and a sensitive level C6. The osteotendinous reflexes of the 2 lower limbs as well as the skin-abdominal reflexes were abolished. On general examination, the child was apyretic, conscience and intelligence were preserved. The ophthalmological examination showed reduced visual acuity on the "count the fingers to 1 m" test, with bilateral papillary pallor at the back. The visual evoked potentials showed an extension of the latency time of the 2 eyes, while the auditory and somesthetic evoked potentials of the trunk were normal.

On the radiological level, the medullary MRI objectified an aspect of transverse longitudinal myelitis extended over more than 3 vertebrae of cervico-dorso-lumbar topography. Brain MRI showed signs of retrobulbar optic neuritis such as bilateral hyper signal signal of the predominant optic nerve at the optic chiasm associated with hyperintense nodules at the right posterior thalamic level, not specific for white matter.

The CSF study showed a pleiocytosis at 45 elements / mm^3 with lymphocytes at 70% and a proteinorrhage at 2.5 g / l and a glycorrhage at 0.45 g / l (glycemia / glycorrhage ratio: 0.5). The search for mycobacterium tuberculosis in the CSF by PCR and by GeneXpert was negative. The viral serologies and the immunological assessment were normal. The anti-NMO antibody assay was negative. Therapeutically, the patient received a bolus of methylprednisolone at a dose of

1g / 1.73m^2 / d for 5 days. In the absence of response to treatment, polyvalent immunoglobulins at a dose of 2 g / kg were administered with an oral relay by prednisone 2 mg / kg / day at a decreasing dose. A first course of rituximab had been carried out 1 month later in two injections 15 days apart at a dose of 375 mg / m^2. The evolution was marked by the improvement of the visual function on the right side, lowering of the level sensitive become D6 and regression of sphincter disorders but with persistence of functional symptomatology.

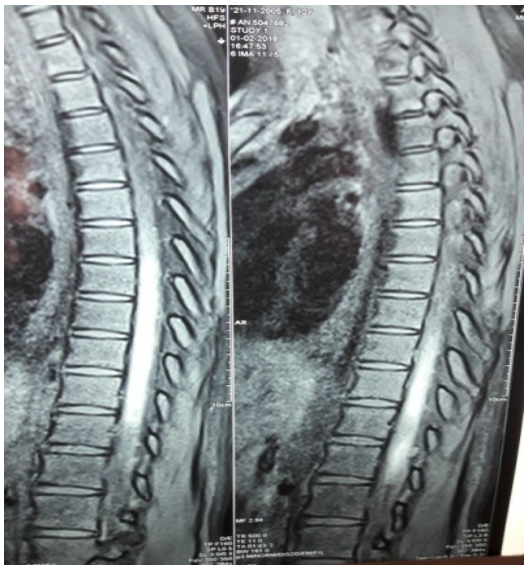

Image 1: extensive transverse cervical-dorsal-lumbar myelitis associated with a few leptomeningeal contrasts in the lumbar and cervical regions, hypersignals of the above-mentioned non-MS-like tendon type.

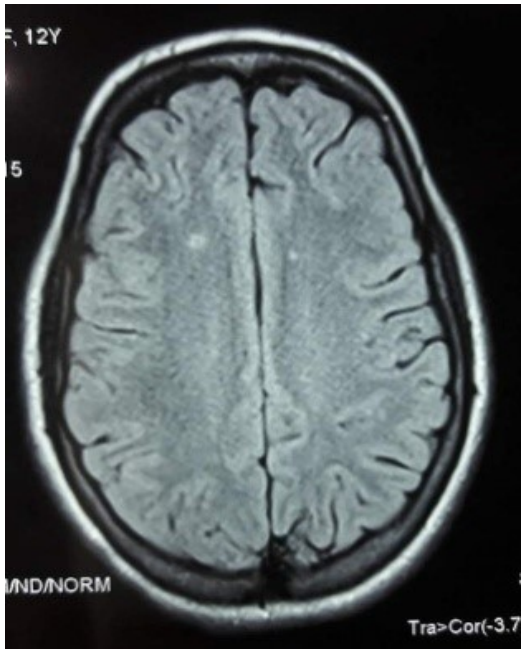

Image 2: multiple punctiform intra-parenchymal calcifications above tentorial sparse in iso-signal T1 and hyposignalT2 at the junction gray substance white substance with presence of nodular lesions in hypo T1, hypersignal T2 and T2 flair, some of which have a hyposignal center, visible at the upper right thalamic level and white matter.

Discussion

Devic optic neuromyelitis (NMO) is a rare inflammatory disease of the central nervous system, defined by attacks of transverse myelitis and optic neuritis. For a very long time, it was considered as a particular form of multiple sclerosis (MS) and treated as such [1]. However, recent work has highlighted clinical, epidemiological, immunological and anatomopathological differences between MS and NMO. The dysfunction of the optic nerve and that of the spinal cord can be concomitant, or successive. Our patient presented a bilateral optic neuritis followed by an acute myelitis and the diagnosis was retained according to the criteria of Wingerchuk [2] before the association of compulsory criteria which are optic neuritis, acute myelitis and the absence of signs in favor of damage other than that of the optic nerve and marrow, to a major secondary criterion which is a spinal cord injury extended over more

than 3 vertebrae, and to two minor secondary criteria which are bilateral optic neuropathy and optic neuropathy severe with visual acuity less than 1 / 10th. In most cases, the decrease in visual acuity is abrupt, bilateral, although frequently asymmetrical. Papillary edema is often moderate. Jeffrey and Buncic [3] reviewed all the pediatric cases of NMO reported in the English literature since 1900. Only 18 cases obeyed the current criteria of Devic's NMO. The mean age of onset was 11.9 years; there was a female prevalence with 11 girls and 7 boys. 2/3 (66%) initially had optic neuritis, 28% transverse myelitis and 6% concomitant involvement. Optic neuritis was bilateral in 88% of the cases, with an acute and severe visual loss which is the case in our patient too. All of the publications agree on the good specificity of anti-AQP4 antibodies (> 90%) with a sensitivity estimated at 73% [5]. The discovery of the positivity of anti-AQP4 antibodies is of less interest for the positive diagnosis if the disease is already advanced because the criteria of NMO are often already met [6]. It still allows us to have an additional objective criterion to establish the diagnosis definitively. By definition, cerebral MRI is normal apart from the extensive hypersignal of the two optic nerves going back to the chiasm in connection with a retrobulbar optic neuritis. However, most authors admit the presence of nonspecific hypersignals of the encephalic white matter on T2-weighted sequences, especially in correlation with the duration of evolution, without calling into question the diagnosis of NMO [7]. The cerebral MRI of our patient showed the presence of nodular lesions in hypo T1, hypersignal T2 and T2 flair, some of which have a hyposignal center, visible at the upper right thalamic level and white matter. The serologies carried out made it possible to eliminate the other etiologies which could explain these hypersignals. The handicap is a direct consequence of the attacks and not of a progressive phase, which is exceptional [8]. Recovery from the thrust can take several months and be of remarkable quality even if the initial disability was dramatic. This is particularly true for medullary thrusts but much less for visual thrusts. There may also be benign forms of development, but at present no specific prognostic marker has been identified [9; 10]. The prognosis in children generally seems favorable under corticosteroid therapy and correlated with early diagnosis and treatment, as well as the absence of relapses. However, in this observation, the child remained paraplegic despite the concomitant treatment with immunoglobulins and corticosteroids.

Conclusion

The clinical course of NMO is variable. It can be a fulminant and fatal monophasic disease or one associated with varying degrees of recovery. Pediatric cases generally have a monophasic course and many have complete neurological recovery. Multi-phase trajectories characterized by relapses and remissions also occur.

References

1. Seze de J. Histoire de la neuromyélite optique de Devic. PratNeurol FMC 2011; 2:41-4
2. Wingerchuk DM, Hogancamp WF, O'Brien PC, Weinshenker BG.The clinical course of neuromyelitis optica (Devic's syndrome).Neurology 1999;53:1107-14.
3. jeffery AR, Buncic JR. Pediatric Devic s neuromyelitis optica. J Pediatr Ophthalmol Strabismus 1996; 33:223–9
4. Papeix C. Maladie de Devic. Presse Med 2006; 35:11–2
5. P.J. Waters, A. McKeon et al. Serologic diagnosis of NMO: A multicenter comparison of aquaporin-4-IgG assays.
6. N. Collongues et al. Neuromyelitis optica in France: A multicenter study of 125 patients
7. DE SEZE J, STOJKOVIC T, FERRIBY D, GAUVRIT JY, MONTAGNE C, MOUNIER-VEHIER F et al. Devic's neuromyelitis optica: clinical, laboratory, MRI and outcome profile. JNeurol Sci. 2002; 197: 57 -61.
8. Wingerchuk DM, Pittock SJ, Lucchinetti CF, Lennon VA, Weinshenker BG. A secondary progressive clinical course is uncommon in neuromyelitis optica. Neurology 2007; 68: 603-5
9. Köller H, Neuen-Jacob E, Saleh A, Kieseier BC,Jander S, Hartung HP.A patient with a benign course of neuromyelitis optica (Devic's syndrome) over 12 years: MRI follow up and histological findings.J Neurol2006; 253:819-20.
10. Wingerchuk DM, Weinshenker BG. Neuromyelitis optica: clinical predictors of a relapsing course and survival. Neurology .2003; 60:848-53.