Early Presentation of Neuromyelitis Optica

NIKOLA DIMITRIJEVIC, DRAGANA BOGICEVIC, *ALEKSANDAR DIMITRIJEVIC AND DIMITRIJE NIKOLIC

*From the Department of Neurology; and *Educational Department, University Children's Hospital, and Medical Faculty University of Belgrade, 11000 Belgrade, Serbia.*

Correspondence to:
Dr Nikola Dimitrijevic,
University Children's Hospital,
Neurology Department,
Tirsova 10, 11000 Belgrade, Serbia.
ndimitrijevic@beotel.net.
Received: February 22, 2012;
Initial review: March 15, 2012;
*Accepted: July 06, 2012.*Abstract

Neuromyelitis optica is a rare autoimmune demyelinating disease of the central nervous system in childhood. Its relapsing form is usually reported in adults. We report a 3-year-old girl with relapsing, IgG seropositive neuromyelitis optica. Initially she presented with optic neuritis, followed by three relapses with deterioration of optic neuritis and developing transverse myelitis. With each relapse, the treatment was less effective. Four years after the onset of the disease, the patient was blind, had paraplegia associated with urinary and bowel incontinence and short stature.

Key words: *Childhood, Neuromyelitis optica, Relapsing form.*

Neuromyelitis optica is an inflammatory demyelinating disease of the central nervous system clinicaly presenting as optic neuritis and transverse myelitis [1,2]. The onset ranges from early childhood to late adulthood with the mean age in the forties. Pediatric case series data are insufficient, especially for patients younger than six years [3,4].

CASE REPORT

A previously healthy 3-year-old girl presented with sudden visual loss preceded by pain while moving the left eye. No recent history of fever, respiratory or gastro-intestinal symptoms, infections, trauma, or vaccination was reported. The family history was negative for neurological and autoimmune disorders. Neurological evaluation revealed divergent strabismus, mydriasis, and diminished pupillary reflex of the left eye. Fundoscopy showed left optic disc pallor. Visual evoked potentials were absent on the left eye, and significantly prolonged with diminished amplitudes on the right eye. Brain MRI revealed slight enlargement of the optic nerves, predominantly on the left side. Complete blood count and biochemical testing was normal. Analysis of the cerebrospinal fluid showed normal findings with absent oligoclonal bands. Serological tests for Toxoplasma, *Borrellia burgdorferi*, *Mycoplasma pneumoniae*, Herpes simplex virus, Epstein-Bar virus, cytomegalovirus, adenoviruses, influenza A and B viruses, varicella zoster virus and HIV were negative. Antinuclear, antiphospholipid and anti-thyroid antibodies, LE cells and rheumatoid factor were negative. Levels of complement were within normal ranges. The patient was treated with intravenous methylprednisolone 20 mg/kg/day for 5 consecutive days, followed by oral prednisone. Clinical improvement was noticed 2 weeks later with visual function recovery on visual evoked potentials (complete on the right and partial on the left eye).

The first relapse occurred 17 months later. The patient developed flaccid paraparesis with increased tendon reflexes and loss of sphincter control, preceded by back pain. This was followed by bilateral loss of vision in the next five days. Brain MRI was normal. Spine MRI showed a signal increase in spinal cord between T8 and T12 levels on T2–weighted images. Intravenous methylprednisolone therapy was repeated. Ten days after the initiation of the steroid therapy the patient regained vision of the right eye, with improved muscle strength and nearly normal muscle tone of lower extremities. A month later sphincter control was established. Spinal cord lesion on the control MRI disappeared completely.

Second relapse occurred at the age of 5 years and 10 months, with loss of vision in the right eye and weakness in the arms and legs. There was moderate hypertonia in legs with Babinski sign and increased tendon reflexes. Visual evoked potentials showed no cortical responses. Brain MRI was normal. Spine MRI revealed enhanced signal on T2 weighted images between C1 and T2 levels (***Fig.*** **1a**). Intravenous methylprednisolone was administered, with significant improvement after two weeks. Patient was able to walk, climb the stairs and jump. Muscle tone of the arms and legs was normalized, but the left eye blindness persisted.

The third relapse occurred at the age of 6 years and 4 months with total loss of vision and inability to sit, stand and walk. Vibratory sensory loss was noted below T10 level. Brain MRI showed atrophy of both optic nerves, optic chiasm and optic tracts . Spine MRI revealed diffuse cervical and thoracic spinal cord atrophy with subsequent dilatation of the central canal (hydromyelia) (***Fig.*** **1b**). Intravenous methylprednisolone and immunoglobulin treatement was unsuccessful. Parents of the patient refused any further therapy.

An year later, neuromyelitis optica testing revealed

high titer of antibodies.

At the age of nine years the patient was blind with spastic paraparesis, sensory loss below T10 level, urinary and bowel incontinence, neurogenic bladder with recurrent urinary infections, and height below third percentile. Mental development was normal. MRI revealed severe atrophy of the spinal cord (***Fig.* 1c**).

DISCUSSION

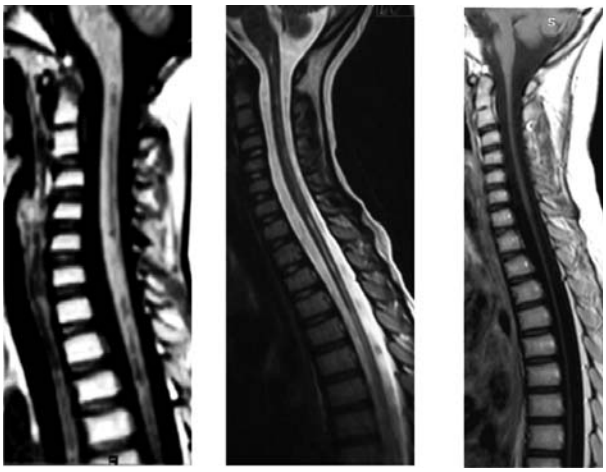
Relapsing neuromyelitis optica is rare in children. The youngest case previously reported in literature was a boy of 23 months [3]. Our patient presented with isolated optic neuritis at the age of 3 years, transverse myelitis occurred 17 months later. At the time of the first relapse, our patient fulfilled all absolute criteria, two out of three major supportive criteria and all minor supportive criteria for the diagnosis of neuromyelitis optica. NMO-IgG antibodies were confirmed later [5-7].

The diagnosis of neuromyelitis optica has been considerably facilitated by the discovery of highly specific serum autoantibody biomarker - NMO-IgG [6]. The high titer of NMO-IgG correlates well with frequent relapses [8]. Patients with the relapsing form of NMO have demonstrated progressive disability.

There are no scientifically proven guidelines and treatment strategies either in the acute attacks or on a long-term base. The outcome depends on early effective immunosuppression prior to accumulation of severe neurological damage.

Our patient is one of the youngest cases with relapsing NMO and NMO-IgG seropositivity reported in the literature. Although rare, pediatric NMO needs specific attention due to an unpredictable clinical course and possible poor outcome with severe disability . It is very important to identify risk factors that predict a relapsing course and to determine optimal treatment, especially for long-term preventive immunotherapy.

Acknowledgments: Professor Robert Marignier and Laboratory in Bron (France) for performing NMO-IgG analysis. Professor Borivoj Marjanovic from Insitute for Mother and Child, Belgrade, for useful clinical suggestions.



(*a*) (*b*) (*c*)

FIG. 1 *Progression of spinal cord atrophy shown on MRI (a) at the second relapse; (b) at the third relapse; and (c) after third relapse.*

REFERENCES

1. Wingerchuk DM, Weinshenker BG. Neuromyelitis optica. Curr Treatment Options Neurol. 2008;10:55-66.
2. Weinshenker BG. Neuromyelitis optica is a distinct from multiple sclerosis. Arch Neurol. 2007;64:899-901.
3. Yuksel D, Senbil N, Yilmaz D, Yavuz Gurer YK. Devic's neuromyelitis optica in an infant. J Child Neurol. 2007;22:1143.
4. Loma IP, Asato MR, Filipink RA, Alper G. Neuromyelitis optica in young child with positive serum autoantibody. Pediatr Neurol. 2008;39:209-12.
5. Wingerchuk DM, Lennon VA, Pittock SJ, Lucchinetti CF, Weinshenker BG. Revised diagnostic criteria for neuromyelitis optica. Neurology. 2006;66: 1485-9.
6. Lennon VA, Wingerchuk DM, Kryzer TJ, Pittock SJ, Lucchinetti CF, Fujihara K, *et al*. A serum autoantibody marker of neuromyelitis optica: distinction from multiple sclerosis. Lancet. 2004;364:2106-12.
7. Krupp LB, Banwell B, Tenembaum S. Consensus definitions proposed for pediatric multiple sclerosis and related disorders. Neurology. 2007;68:S7-12.
8. Banwell B, Tenembaum S, Lennon VA, Ursell E, Kennedy J, Bar-Or A, *et al*. Neuromyelitis optica-IgG in childhood inflammatory demyelinating CNS disordes. Neurology. 2008;70:344-52.