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RESEARCH ARTICLE

A CASE REPORT OF PARAPLEGIA IN A YOUNG FEMALE WITH DEVIC'S DISASE.

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ABSTRACT

This is a case of a younge female who presented with b/l lower limb weakness and then paralysis. Loss of sensation of both lower limbs and loss of bowel and bladder control. On examitaion and tests patient was found to have paraplegia and csf was positive for anti nmo antibody called as neuromyelitis optica. The patient was treated with iv steroids and then with oral steriods and supportive treatment

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INTRODUCTION

Neuromyelitis optica (NMO) or Devic's syndrome is an idiopathic, severe, inflammatory demyelinating disease of the central nervous system that selectively affects optic nerves and spinal cord. It tends to spare the brain early in the disease course. Early discrimination between multiple sclerosis (MS) and NMO is important, as optimum treatment for both diseases may differ considerably. In contrast to typical MS, clinical experience and case series suggest that NMO requires long-term immunosuppressive therapy. Diagnostic criteria for NMO from 1999 have been revised in 2006 by Wingerchuk et al. (2006); Wingerchuk, 2008. NMO-IgG has facilitated an appreciation that the spectrum of NMO is wider than previously recognized, and includes patients with recurrent longitudinally extensive transverse myelitis, recurrent isolated optic neuritis, and Japanese opticospinal MS (Lennon et al., 2004). A variety of encephalopathic presentations may occasionally be encountered in NMO-IgG seropositive patients, most commonly in children (McKeon et al., 2008).

Is Devic's disease a type of multiple sclerosis?: Until recently, Devic's disease was thought to be a kind of Multiple sclerosis (MS) that caused more severe problems with the optic nerves and spinal cord. Recent research has suggested that Devic's disease is probably a different disease in which there is a specific immune attack on a molecule known as aquaporin 4.

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Our understanding of Devic's disease is changing quickly at the present time due to new research in this important disorder.

Potential symptoms of this disease include:

- Loss of vision or blurred vision
- Weakness of the extremities
- Numbness of the extremities
- Incontinence of bladder and/or bowels
- Spasticity of the muscles

How is devics disease diagnosed?

Diagnosis: The Mayo Clinic proposed a revised set of criteria for diagnosis of Devic's disease in 2006. Those new guidelines require two absolute criteria plus at least two of three supportive criteria. In 2015 a new review was published by an international panel (Wingerchuk et al., 2003) refining the previous clinical case definition but leaving the main criteria unmodified:

Absolute criteria:

- Optic neuritis
- Acute myelitis

Supportive criteria:

- Brain MRI not meeting criteria for MS at disease onset
- Spinal cord MRI with continuous T2-weighted signal abnormality extending over three or

more vertebral segments, indicating a relatively large lesion in the spinal cord
- NMO-IgG seropositive status (The NMO-IgG test checks the existence of antibodies against the aquaporin 4 antigen.)

Testing for Devic's disease may include MRI scans, to show inflammation of the spinal cord. In a person with Devic's disease, the MRI scan may show inflammation in a long segment of the spinal cord. In MS, the findings tend to be in a short segment in the spinal cord. In Devic's disease, the MRI scan of the brain may be normal or show relatively mild changes. The optic nerve MRI may show areas of abnormality**.** Another difference in findings for Devic's disease as compared to Multiple sclerosis(MS) is that cerebrospinal fluid may show a greater increase in white blood cells than in MS patients, and may show a type of cell (neutrophil) that is not usually seen in MS.

In general, the test for oligoclonal bands (a test that is often positive in MS) is usually negative in the spinal fluid in Devic's disease. Oligoclonal bands are immunoglobulins (or antibodies), proteins produced by the immune system to fight off invaders like bacteria or viruses. A blood test known as the NMO-IgG blood test is positive in 70 percent of patients diagnosed with Devic's disease. This test, in general, is negative in patients with multiple sclerosis. This has become an important marker for Devic's disease and has helped improve our understanding of this disorder.

Case report

A 41 year old female came with chief complaints of inability to move both lower limbs , loss of sensation in both lower limbs, incontinence of urine and stools and lower back ache. To start with, patient 1st experienced weakness of both lower limbs just being able to manage her routine activities and subsequently developed paralysis of both lower limbs confining her to bed. Sensory symptoms developed from the 3rd day of weakness of her limbs and incontinence of urine and stools from 4th day of weakness of lower limbs.

On examination

Pulse 82/min ; Blood Pressure -110/80mmhg ; Temperature – afebrile.

Central nervous system examination

Tone: Upper (b/l)- normal lower (b/l)-hypotonia.

Nutrition: Upper (b/l)- good lower (b/l)-good.

Power: Upper (b/l)-5/5 lower (b/l)-0/5.

Sensory: Upper (b/l)-present lower (b/l)-absent.

Reflexes: (Deep tendor reflexes) upper (b/l)- present lower (b/l)- absent.

Plantars: Right-upgoing left-upgoing

Cardiovascular system – within normal limits.

Respiratory system- within normal limits.

Per Abdomen system- within normal limits.

INVESTIGATIONS

Haemaoglobin-8.3gm/dl WBC- 16600 PLATELETS-48300

CSF Examination-total cells – 2cmm
Lymphocytes – 100%
Oligoclonal Bands- not seen

Neuromyelitis optica antibody titre: 1:10 (positive)
ana blot assay: ro52 + , ribp-protein + fundoscopy of both eyes - within normal limits

MRI whole spine suggestive of Diffuse altered signal abnormality involving dorsal spinal cord from D2-D10 vertebral levels with heterogenous peripheral nodular enhancements within spinal cord extending from D4-D9 Levels suggestive of active demyelinating etiology.

Treatment: Her treatment included I/V Methyl prednisolone 1 gram TDS for 5 days along with oral methyl prednisolone and supportive treatment of i/v antibiotics and oral medications. Over a period of 15 days, she responded well to treatment and improved. She was discharged on oral wysolone and on follow up, wysolone was continued. She had no recurrence of symptoms over the last 3 months.

DISCUSSION

Neuromyelitis optica (NMO) was first identified in the 19th century, as a monophasic, destructive disorder affecting the spinal cord and both optic nerves, but sparing the remainder of the central nervous system (CNS). The position of NMO within the group of CNS demyelinating diseases has been long debated - whether this clinical entity is a severe variant of MS, a form of acute disseminated encephalomyelitis, or a distinct disease (Wingerchuk, 2003; Pittock, 2006). It has gradually become accepted that the spectrum of idiopathic NMO is broader than suggested by the historical definition. The recent discovery of serum autoantibody marker NMO-IgG in 2004 further advances the hypothesis that NMO is actually a distinct disease (Lennon et al., 2004). NMO-IgG is 73% sensitive and 91% specific for distinguishing NMO from optic-spinal presentations of classical MS. The target antigen of NMO-IgG is aquaporin 4 (AQP4), glial water channel protein, that facilitates water transport, especially in "stress situations" such as brain injury. It is component of the dystroglycan protein complex located in astrocytic foot processes at the blood-brain barrier. Data suggest that autoantibodies to aquaporin 4, derived from peripheral B cells, cause the activation of complement, inflammatory demyelination, and necrosis that is seen in neuromyelitis optica [7, 8]. It is also important to emphasize that patients presenting with a first-ever LETM event, who are found to be NMO-IgG seropositive, have a 56% risk of LETM recurrence or optic neuritis (conversion to NMO) during the subsequent 12 months (Wingerchuck, 2007). With the positive NMO-IgG finding we confirmed the diagnosis of NMO in our patient, who satisfied all absolute and supportive diagnostic criteria (Wingerchuk et al., 2006). According to the data from the literature, most patients with NMO, perhaps more than 90%, have relapsing rather than monophasic disease (Wingerchuck, 2007), which was also the case in our patient, but was not recognized as that in the beginning of the disease.

Our patient met the features of relapsing NMO, such as female dominance, older age at onset, and probably autoimmune disease - we could assume that steroid therapy unmasked autoimmune diabetes mellitus (Wingerchuck, 1999). For many neurologists, the current standard preventive approach for NMO includes oral azathioprine in a combination with oral prednisone, rituximab (chimeric anti-CD20 monoclonal antibody), mitoxantrone, IVIGs, and cyclophosphamide (Mandler, 1998; Birnbaum et al., 2008; Feingold, 2008; Jacob et al., 2008; Jarius et al., 2008; Schröder et al., 2009; Mok et al., 2008). The best evidence (although all from retrospective series) for long-term immunosuppression in NMO is with azathioprine and rituximab, that are considered first line therapies, while cyclophosphamide is generally considered as a second line agent in NMO. In a recently published retrospective multicenter case series of NMO patients treated with rituximab, that included 25 patients (including 2 children), 23 of whom experienced relapses despite use of other drugs before rituximab, treatment with rituximab appeared to reduce the frequency of attacks, with subsequent stabilization or improvement in disability (Jacob et al., 2008). Jarius et al reported that treatment with immunosuppressants such as rituximab, azathioprine and cyclophosphamide resulted in a marked reduction in antibody levels as well as in relapse rates, demonstrating a strong relationship between AQP4-Abs and clinical state. Cyclophosphamide resulted in a long-lasting relapse-free interval in one of their patients, with 10 relapses within 1295 days (2.82/year) prior to initiation of therapy but only one within 1610 days (0.23/year) under therapy (Jarius et al., 2008). Immunoablative cyclophosphamide was also successful in halting relapses in a patient with systemic lupus erythematosus-associated NMO who was unresponsive to high-dose oral and intravenous corticosteroids, intravenous immunoglobulin, mycophenolate mofetil, tacrolimus, low-dose daily oral cyclophosphamide and rituximab (Mok et al., 2008).

After treating diseases attacks with pulse corticosteroid therapy and IVIGs in our patient, we included oral azathioprine in a combination with oral prednisone in the therapy, but since there was no significant therapeutic response, we decided to use cyclophosphamide therapy. That resulted in good clinical improvement and gradual decrease in cord swelling and T2 signal hyperintensity. We were also considering to use rituximab, but at that time this agent was not approved for treating NMO in Croatia. It is important to point out that one does not need to wait for a second attack of transverse myelitis or optic neuritis prior to commencing immune suppression if NMO-IgG is positive, since NMO-IgG itself predicts a relapsing course. Unfortunately, at the time we received positive finding of NMO-IgG antibodies from Mayo Clinic, our patient experienced relapse. If immunosuppresive therapy had been started earlier in the course of the disease (patient's clinical worsening started to happen one year before admission to our institution), maybe the prognosis for this patient could be more favourable.

Conclusion

In this case report we wanted to emphasize the extensiveness of inflammatory spinal cord changes in our patient, from T4-T9, which was initially attributed to Longitiudnal Extensive Transverse Myelitis, but after investigatiosn evolved to neuromyelitis optica which improved symptomatically and clinically with conservative management. In the conclusion it is important to say that accurate, early diagnosis and distinction from multiple sclerosis is critical to facilitate initiation of immunosuppressive therapy for attack prevention.

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