Pakistan Journal of Neurological Sciences (PJNS)

Volume 14 | Issue 4 Article 2

12-2019

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NEUROMYELITIS OPTICA SPECTRUM DISORDER: A CASE REPORT FROM REHMAN MEDICAL INSTITUTE, PESHAWAR.

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***Date of submission:** September 13, 2019 **Date of revision:** October 14, 2019 **Date of acceptance:** October 21, 2019*

ABSTRACT:

Devic's disease or Neuromyelitis Optica is a serious, rare idiopathic neurological disorder which affects the optic nerve and the spinal cord. A 15-year-old girl was admitted in the neurology was with a previous history of 4 episodes of weakness in both the lower limbs and urinary retention. The patient gradually lost her eyesight in both eyes at the age of 8. Initially, she was diagnosed as a case of Multiple Sclerosis at the age of 11 when she presented with first episode of lower limb weakness. Later, Seropositivity for Aquaporin 4 IgG and clear MRI scan of the brain confirmed the diagnosis of Neuromyelitis Optica.

Case Report:

Devic's disease or Neuromyelitis Optica is a serious, a rare idiopathic inflammatory neurological disorder which affects the optic nerve and the spinal cord. It is a severe demyelinating disorder of the nervous system characterized by recurrent attacks on the optic nerve and spinal cord with or without recovery and maybe fatal [(1)] In most cases, the etiology of Neuromyelitis Optica isn't specific and it is believed to be an immunological disorder [(2)]. A few studies have detected high titers of anti-Epstein-Barr virus [(3)] antibody in patients suffering from the disease and a possible association with tuberculosis [(4)] Neuromyelitis Optica has worldwide distribution and poor prognosis. It causes a sudden or gradual loss of vision in one or both eyes, varying degrees of weakness or paralysis of legs or arms, loss of sensation and bladder and bowel dysfunction. Originally the disorder was considered to be a variant of multiple sclerosis (MS), but clinical, pathological, radiological and immunological data of this disease have permitted clearer differences between both the diseases and lead to a novel definition of this clinical entity [(5)] The course of NMO is more severe and treatment strategies to prevent further attacks are different, therefore, it is important to differentiate between NMO and MS. Immunotherapy to prevent relapses of MS does not effectively prevent recurrent NMO attacks [(6)]. The advent of NMO antibody, the specific biomarker AQP4-IgG, is an additional criterion to support the diagnosis of NMO (sensitivity and specificity is 91% and 100%) [(7)]

CASE PRESENTATION:

We present here a case of a 15-year-old female patient, coming from a poor socioeconomic background, complaining of weakness in the lower limbs and urine retention. The medical history of our patient includes a gradual loss of vision at the age of 8 over the course of 3 years, losing her vision completely at the age of 11. No neurological diseases were reported in the family. The patient was hospitalized before in 2015 due to weakness in the lower limbs and facial droop. MRI scan of the brain showed no lesions. Fundus examination showed bilateral optic atrophy. (Figure. 1.0) Intermittent generalized slow waves were seen throughout the EEG record. She was treated with methylprednisolone and diagnosed with multiple sclerosis at the time and discharged. At this time she wasn't on an disease modifying therapy. The patient's financial constraints have been a great hurdle in the diagnosis and management of her condition.
with fever, headache, joint and muscle pain, and decreased platelets (thrombocytopenia). It is caused by

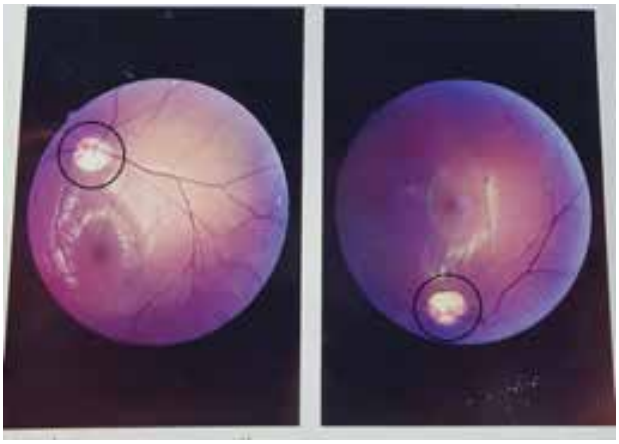

Figure 1.0: Fundus examination showed bilateral optic atrophy (encircled)

Her symptoms improved gradually, but ever since then she has experienced varying degrees of paralysis in the upper and lower limb, urinary retention and constipation every year and has been hospitalized to treat these symptoms Before being admitted here, she has had admission 4 times previously in Government Hospitals with no definitive diagnosis. The patient has been prescribed carbamazepine, azathioprine, and prednisolone to treat her spastic paraparesis episodes. 15 days ago, she presented to the hospital complaining of progressive weakness in her legs, spasticity, hyperreflexia and urine retention. On ophthalmological examination, the absent pupillary light reflex was observed. Ocular examination revealed the patient had bilateral nystagmus (horizontal and coarse).

Her visual acuity had no perception of light (NPL) in both eyes and the pupils were non-reactive to light. Anterior segment was normal in both eyes. Neurological exam showed grade 5 strength in upper limbs and grade 3 strength in the lower limbs. The patient was vitally and hemodynamically stable but her. T2-weighted MRI of the thoracic spine revealed high-intensity signal changes in the spinal cord involving both halves extending from D2 to D9. Similar changes were noted in the cord at L1 and L2. Diffuse edematous signals in entire cervico-dorsal cord with mild post-contrast enhancement was observed on T2 weighted MRI of the thoracic spine. (Figure. 2 There was no cord swelling and focal thinning was seen in the lower cervical cord.

The patient was diagnosed with acute transverse myelitis after observing the MRI results and Neuromyelitis Optica was suspected. Advanced immunological tests performed at Shifa International Hospital confirmed the presence of aquaporin-4-immunoglobulin G antibodies and the patient was diagnosed with Neuromyelitis Optica as she fulfilled both the major and minor diagnostic criterion of the disease. Improvement in her symptoms was seen after she was started on 5mg of prednisolone and went through 5 plasmapheresis sessions.

The patient's electrolytes were monitored regularly as her lab reports showed low levels of potassium and magnesium. Neurological examination before discharge showed grade 4 strength in her lower limbs and she could walk with assistance. No improvement in vision was observed.

Figure 2: MRI - Spine showing the radiologic finding of the patient T2-weighted MRI of the thoracic spine revealed high-intensity signal changes in the spinal cord involving both halves extending from D2 to D9.

DISCUSSION

Through this study, we highlight the complexity of diagnosing neuromyelitis optica and the treatment challenges when socioeconomic factors are a hurdle. Because of lack of funding in public hospitals, plasmapheresis and the advanced immunological test of NMO was delayed. Patients' family was also reluctant to go through these tests and procedures due to financial constraints. The patient presented with typical symptoms of NMO but was diagnosed as a case of MS before she was presented to us. Differential diagnosis between Longitudinal Extensive Transverse Myelitis (LETM) that occurs in MS and NMO and SLE is important since different therapeutic regimens are required and interferon treatment may cause a flare-up of the diseases [5,7,8]. NMO is quite similar to the optico-spinal form of MS hence it can be misdiagnosed, however, the course of NMO is more acute. Optic neuritis is severe with poor prognosis [9], our patient had a very severe case of optic neuritis and lost her vision irreversibly.

NMO Ig antibody is a highly specific diagnostic marker for NMO. NMO is a rare pathological entity with neurological manifestations. Previous literature suggests that in patients' with SLE or Sjogren's syndrome positive NMO IgG is not a secondary effect but possibly these patients have two autoimmune disorders [10,11]. In other studies, the presence of NMO IgG has been considered as a predictor for the manifestation of NMO spectrum disorder and predicts

relapse of the disease. Co-existence of NMO is other autoimmune disorders either organ-specific or non-specific results in a poorer prognosis [12].

The current diagnostic criteria for NMO requires the presence of optic neuritis and acute transverse myelitis along with supportive criteria which include aquaporin 4 seropositivity normal brain magnetic resonance imaging or not meeting the criteria for multiple sclerosis and longitudinally extensive cord lesion extending over 3 or more vertebral segments [7]. Our patients radiological and serums results fulfilled the diagnostic criteria and all 3 supportive criteria were present.

The common initial therapy for acute attacks of NMO is intravenous corticosteroids. In some studies, total remission was obtained after prednisolone treatment. Patients who do not benefit from corticosteroid therapy may benefit from plasmapheresis [7]. Our patient went through 5 plasmapheresis sessions and was given 5 mg of prednisolone during her stay. The first line therapy suggested for NMO is azathioprine combined with prednisolone and rituximab whereas cyclophosphamide, mitoxantrone, and mycophenolate mofetil are the second line [14]. However, it is unclear that if NMO with other autoimmune disorders requires different treatment regimens.

The prognosis for NMO varies, studies have shown that 20% of the patient are functionally blind in one eye and 31% developed monoplegia/paraplegia. The 5-year survival rate in patients with paraplegia was 91% [13]

CONCLUSION:

NMO is a rare but severe devastating disease affecting vision and nervous system resulting in blindness and paraplegia in children. Hence an early intervention with the appropriate treatment modality in patients suspected with NMO decides on the favorable outcome from an acute episode.

CONSENT:

Informed consent was taken prior to the writing of this case report from the patient.

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Conflict of interest: Author declares no conflict of interest.
Funding disclosure: Nil

Author's contribution:
Omer Nasim: Study concept and design, protocol writing, data collection, data analysis, manuscript writing, manuscript review
Zainab Rustam: data collection, data analysis, manuscript writing, manuscript review
Muhammad Shahid Iqbal: Study concept and design, data analysis, manuscript writing, manuscript review