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Research Article

NEUROMYELITIS OPTICA

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A R T I C L E I N F O

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A B S T R A C T

Background: Neuromyelitis Optica (NMO), also known as Devic's disease, is an inflammatory condition of young females leading to demyelination especially of the optic nerve (ON) and spinal cord. Clinical features vary too widely that they form neuromyelitis disease spectrum. Some consider it to be a variant of multiple sclerosis (MS). It is caused by autoantibodies against aquaporin 4 (APQ4) channels. As a result, NMO is considered as an autoimmune disease.
Objective: To describe a patient which satisfied clinical criteria for NMO, however, lacked the laboratory confirmation of aquaporin autoantibodies.
Methods: Detailed history, physical examination and laboratory investigations.
Conclusion: As NMO has a disastrous clinical course, early identification of this disease is necessary, as with the possible treatment like rituximab, clinical course can be altered and the progression hampered.

INTRODUCTION

Neuromyelitis Optica (NMO), also known as Devic's disease, is an inflammatory condition of young females leading to demyelination especially of the optic nerve (ON) and spinal cord. Clinical features vary too widely that they form neuromyelitis disease spectrum. Some consider it to be a variant of multiple sclerosis (MS). It is caused by autoantibodies against aquaporin 4 (APQ4) channels. As a result, NMO is considered as an autoimmune disease. As the autoantibodies affect the astrocytes in the central nervous system, the disease is an astrocytopathy. Combining all these, it is an autoimmune astrocytic channelopathy. It is known to occur in connection with viral and bacterial infections. It also has associations with other autoimmune disorders like systemic lupus erythematosus (SLE) and Sjogren syndrome. The clinical onset comprises of an acute attack with affection generally of optic nerve or spinal cord. The course is highly variable. Some patients have a monophasic illness with varying degrees of recovery while other have a relapsing and remitting course. The exact nature and clinical course of the disease is unpredictable. Diagnosis is by clinical history, examination and neuroimaging showing demyelination as longitudinal segment (more than three segments) in spinal cord and either clinical blindness, loss of visual acuity or by evoked potentials. Treatment of acute attack is high dose methyl prednisolone.

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Treatment of the disease per se is usually supportive. However, rituximab has been found to be effective. The patients having associated other autoimmune disorders like SLE and Sjogren syndrome are treated with immunomodulator therapy.

Case Report

A 47 year old hindu female who was a housewife by name Veenaben Kantibhai Parmar, residing at Chhani, Vadodara, Gujarat, India presented to the outdoor patient department of Sir SayajiRao General Hospital, Vadodara with complaints of back pain for 10 months and difficulty in walking for 10 months. On detailed history, it was found that her back pain started all of a sudden in one morning in January of 2016, followed in a few hours by complaints of heaviness sensation in both lower limbs. By that day evening, she was unable to stand up on her own, and by night time, she was unable to move her legs. The next day morning she noticed swelling on the lower abdomen and difficulty in passage of urine. She went to a local doctor, who catheterized her and around 2000 ml of urine was evacuated. She had no complaints of dimness of vision or double vision. No sensory complaints. No complaints related to other cranial nerves. No recent fever, multiple joint pains or rash. She was not recently vaccinated. For her complaints, she went to a local doctor, who gave her methyl prednisolone injections for 5 days. She was investigated with an MRO of back and CSF examination. Over the next few months, she gained control of her bladder and had some improvement in lower limbs. However, she had some difficulty in walking since then and had back pain near

continuous in nature with occasional relief by pain medications since then.

She is a known case of hypothyroidism since last six years on tablet thyroxine 25 micrograms a day. She had past history of weakness of both lower limbs with bladder involvement and some weakness of left upper limb in 2012, which recovered near completely over a span of 20 days with methyl prednisolone.

Her family history is insignificant in terms of her current illness. She is vegetarian by diet and has adequate sleep. She occasionally has urgency, at night time, and wets her bed. She had normal menses and insignificant obstetric history.

On examination, she was vitally stable. She had normal light reflex, but hippus was present bilaterally. Marcus Gunn pupil was present in left eye.Her cranial nerves were normal. She had weakness on entire left side of body by a MRC power grade of 4/5 and weakness of both lower limbs by a power grade of 3/5. She was able to perform all tests of coordination normally with her upper limbs. On examination, she had impairment of pain and temperature sensations from just below knee downwards in both lower limbs. Vibration sense was lost at all bony prominences from knee below and fine touch was lost from just below knee in right lower limb and just above knee in left lower limb. Sensations in upper limbs were normal. Among the superficial reflexes, abdominal reflex was lost in all quadrants and plantar response was extensor. All deep reflexes were exaggerated with ankle reflex absent. There was no clonus. She had positive Rhomberg sign with eyes open which aggravated with eyes closed. There was paraspinal muscle spasm over the lumbar region on left side. Movement of back was restricted in all quadrants due to pain. Straight leg raising test was positive. Lhermitte sign was positive. She needed support while standing from sitting position. She walked slowly with a wide base, and small step length. Rest of the systemic examination was normal.

Investigations

Parameter	Value
Haemoglobin	10.60 gm%
Total Counts	8470 per cu. Mm
Differentials	74 / 22 / 02 / 02
Platelet counts	3.23 lakh per cu. Mm
ESR	42 mm in one hour
Smear	Microcytic hypochromic RBCs. Ovalocytes seen
Blood urea	21 mg/dl
Serum creatinine	1.15 mg/dl
Bilirubin Total	0.7 mg/dl

Chest X-Ray	Normal
RBS	92 mg%
ECG	Grossly within normal limits
Serum TSH	4.03
Serum ANA	Negative
VDRL	Negative
D-Fundus	No retinopathy present
HIV	Negative
Urine routine micro	In normal limits

MRI Dorsal Spine in 2012 showed long segment abnormal intramedullary hyperintensity on T2W images in spinal cord, extending from C6/C7 to D9/D10 levels, suggestive of long segment transverse myelitis.

MRI Spine with contrast in January 2016 showed long segment, abnormal, intramedullary, hyperintense signal on T2W images in cervico-dorsal spinal cord, extending from C6/C7 to D9 levels, with altered signal intensity area involving more than 50% area of spinal cord. On post contrast study, heterogenous enhancement is seen in spinal cord, extending from D2 to D7. Faint restricted diffusion is seen in spinal cord from C7 to C8 levels.

MRI DL Spine with contrast with whole spine screening in September 2016 showed conus ending at D12-L1 level. Subtle abnormal signal intensity, mildly hyperintense on T2W images, is seen in central part of spinal cord at D1 to D8 levels with decrease in volume of spinal cord. Possibility of myelomalcia is likely. Subtle hyperintensity is seen in spinal cord from C3 to C6 levels without expansion of the cord. Possibility of demyelination lesion is likely.

MRI Brain with contrast was normal.

Cerebrospinal Fluid Analysis	
Total cells	2 per cu. Mm
Differentials	0 / 100 %
Total proteins	94 gm/dl
Sugars	50 mg/dl
NMO Antibody in CSF	**Weak Positive**
NMO Antibody in Serum	Detected +1

Visually evoked potential study (VEP study) was suggestive of bilateral optic neuropathy.

Patient was treated with oral steroids with slow tapering with azathioprine as immunomodulator. She was given pain killers and physiotherapy and rehabilitation. She continues to live and fight with her disease. She continues to have pain in the back.

DISCUSSION

Wingerchuk classification criteria for NMO are as follows:
REQUIRED CRITERIA:

- Optic Neuritis
- Acute transverse myelitis
- SUPPORTIVE CRITERIA: (two of the three)
- Longitudinal extensive cord involvement of more than three consecutive vertebral segments
- MRI Brain not meeting the criteria for MS
- Aquaporin 4 seropositivity

This patient clearly has both the required criteria and all three of the supportive criteria. There are no clinical case papers available from the 2012 attack, and considering that as the first attack and the attack in 2016 as the second, the patient would definitely have had a better quality of life if she was diagnosed early and treated for the same, either by steroids or by immunomodulator therapy, since then. Thus, the bottomline message is, for any chronic debilitating disease, it is better to early identify the disease process and treat them at its inception to prevent long term complications and improve the quality of life of the patients.

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