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Original Research Paper

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UNUSUAL PRESENTATION OF NEUROMYELITIS OPTICA – A CASE REPORT

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ABSTRACT Optic neuropathy and transverse myelitis (TM) are common symptoms of multiple sclerosis (MS) but may also be seen in association with the antibody-mediated autoimmune disorder, neuromyelitis optica (NMO). We report a female patient presenting with intractable vomiting and hiccups and TM. Serum NMO antibodies were positive. Serial MRI (Magnetic resonance imaging) abnormalities included longitudinally extensive TM of the cervical cord, focal T2-weighted hyperintensity in upper cervical cord. Clinical and MRI involvement of spine extending over three vertebral segments strongly favoured a diagnosis of NMO. She required several courses of intravenous methylprednisolone and plasmapheresis before receiving intravenous rituximab therapy. NMO spectrum disorder should be considered in the differential diagnosis of atypical central nervous system presentations such as intractable vomiting and hiccups. Recognition of this syndrome has significant implications as its treatment and prognosis differs from MS.

KEYWORDS : Neuromyelitis optica, Relative afferent pupillary defect, Aquaporin 4, Rituximab

INTRODUCTION:

Neuromyelitis optica is an autoimmune, inflammatory disorder of the central nervous system that has a prevalence of 0.5 to 10 persons (predominantly women) per 100,000 population. It is characterized by attacks of optic neuritis and transverse myelitis. Recent studies have identified intractable vomiting and hiccups and other central nervous system (CNS) manifestations including encephalopathy and hypothalamic dysfunction as additional symptoms.[1] NMO is caused by a pathogenic serum IgG antibody against the water channel aquaporin 4 (AQP4) in the majority of patients.[2] Here we report a 23 years old female presenting with intractable vomiting and hiccups and spastic quadriparesis.

CASE REPORT:

A 23 years old female patient came with chief complaints of weakness of all 4 limbs since 15 days. There was no history of preceding fever, trauma, vaccination, altered sensorium, syncope, loss of consciousness, bowel/bladder involvement, sensory involvement.

3 years before this admission she had history of multiples episodes of vomiting and intractable hiccups, for which she consulted a local doctor. She was advised upper gastro-intestinal scopy which revealed lax lower esophageal sphincter. Patient was given antiemetics and antacids which resolved her symptoms. She also noticed difficulty in swallowing for which she was advised MRI (Magnetic resonance imaging) brain but had lost to follow up.

2 years before this admission she had similar complaints of weakness of all 4 limbs which initially started in bilateral lower limbs soon progressed to involve upper limbs. It was associated with retention of urine and bowel incontinence. Patient also decreased pain and touch sensations over bilateral upper and lower limbs. Clinical examination revealed normal cranial nerves and pyramidal weakness of grade 3/5 over all 4 limbs, exaggerated reflexes and plantars extensor. Fundus examination was normal. MRI brain was normal, MRI cervical spine showed a large intramedullary hyperintensity from cervico-medullary junction to C7 level. CSF examination identified 18 mononuclear cells and normal concentrations of protein and glucose. Oligoclonal bands were not detected in the CSF. She was provisionally diagnosed as a case of Transverse myelitis and was given Injection Methylprednisolone (1gm/day) for 5 days. Patient didn't show any improvement so she underwent 5 cycles of plasmapheresis. Patient showed improvement in power which became 4/5 at the end of last cycle of plasmapheresis. Patient was advised physiotherapy and rehabilitation and was discharged on tapering dose of steroids for which she was non-compliant to medications.

4 months before admission, patient noticed decreased vision from left eye for which she didn't seek any medical consultation.

15 days before the admission patient noticed weakness of right upper limb and lower limb which progressed over 5 days to left upper limb and lower limb. Patient was admitted at private hospital where she developed breathlessness and desaturation 5 days after admission, so was intubated and was referred to our hospital for further management.

At presentation in our hospital she was vitally stable. On CNS examination, her power was 0/5 in all 4 limbs, reflexes exaggerated and plantar extensors. Her sensory system was intact and she was able to void urine on her own. Routine ophthalmological examination showed RAPD (relative afferent pupillary defect) and optic atrophy of left eye. MRI of the brain was normal. MRI cervical spine was suggestive of hyperdense lesion from C1 to T6 vertebra. CSF showed 13 lymphocytes and normal concentration of proteins, glucose and it was negative for oligoclonal bands. Her AQUAPORIN-4 antibodies were done, which were positive. MOG antibodies were negative. She was diagnosed as NMO and was started on Injection Methylprednisolone (1gm/day) for 5 days followed by 5 cycles of plasmapheresis. Patient didn't show satisfactory improvement, so was given Injection Rituximab (1gm). Her power has now improved to 3/5 in all 4 limbs.

Table 1: Blood Investigations

	Previous admission	This admission
Haemoglobin	12.8	11.4
TLC	13000	7800
PLATLET	3.8 lacs	2.96 lacs
Urea	30	28
Creatinine	0.9	0.8
Total bilirubin	0.8	1.1
AST	32	26
ALT	24	29
ALP	78	84
CSF – protein	23	32
CSF – glucose	54	70
CSF – cells	18- lymphocytes	13 lymphocytes
CSF – ADA	09	14

AST - aspartate transaminase,
ALT- alanine transaminase, ALP- alkaline phosphatase,
ADA- adenosine deaminase, TLC- total leukocyte count

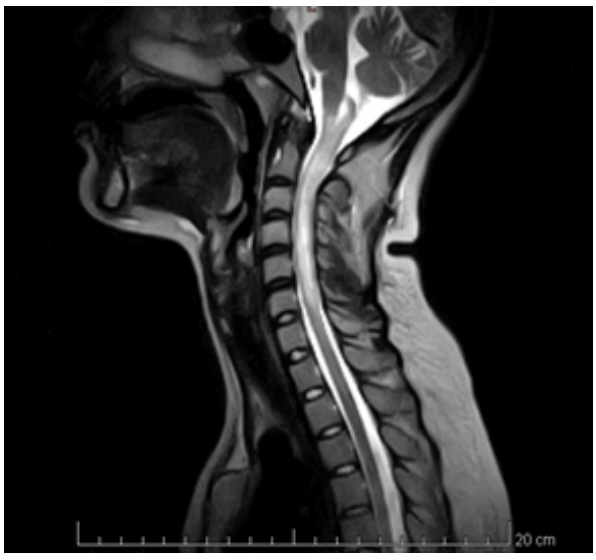

Fig 1: Saggital T2 Image - Long Segment Hyperintesity From C1-t6 (involvement > 3 Vertebra)

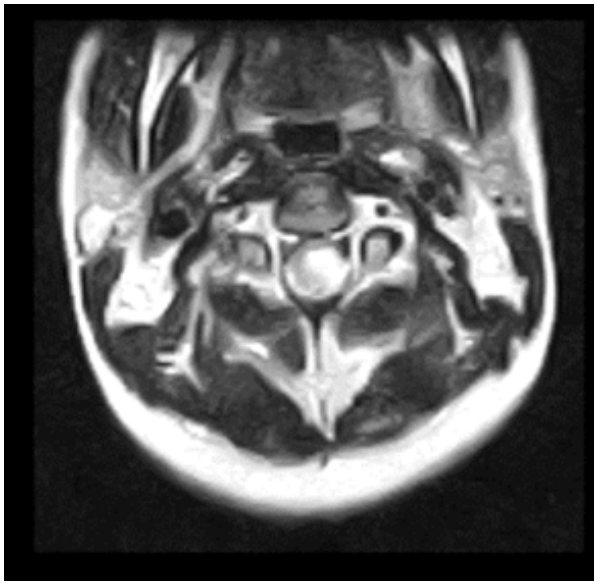

Fig 2: Axial T2w Image - Upper Cervical Cord Reveals Hyperintensity Occupying More Than 2/3 Of Cervical Cord

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, the advanced immunological test of NMO was delayed. Patient's 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 TM before she was presented to us.

During the first admission this female presented with signs and symptoms typically suggestive of transverse myelitis, for which she was treated. She was evaluated only for infective causes (history of fever) and multiple sclerosis (absent oligoclonal bands). On discharge she was advised tapering dose of steroids for which she was non-compliant.

In this admission she had similar complaints of weakness of all 4 limbs and MRI cervical spine (T2) was suggestive of hyperdensities in C1 to T6 (involvement of >/3 vertebral segments, s/o longitudinally extensive transverse myelitis) and ophthalmological examination revealed RAPD and optic atrophy of left eye, which pointed towards NMO spectrum disorder. Serum AQUAPORIN-4 antibodies were done which were positive. So the patient satisfied the criteria for NMOSD.

Diagnostic criteria for NMOSD with AQP4-IgG[3]:

- At least 1 core clinical characteristic
- Positive test for AQP4-IgG using best available detection method (cell-based assay strongly recommended)
- Exclusion of alternative diagnoses

Core clinical characteristics:

- Optic neuritis
- Acute myelitis
- Area postrema syndrome: episode of otherwise unexplained hiccups or nausea and vomiting
- Acute brainstem syndrome
- Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD typical diencephalic MRI lesions
- Symptomatic cerebral syndrome with NMOSD-typical brain lesions

Other possibilities were Multiple Sclerosis which was excluded by absence of oligoclonal bands in csf and MRI brain being normal. Next possibility was Post infectious causes which were excluded by no history of preceding fever or vaccination. ADEM was excluded as there was no evidence of encephalopathy.

CONCLUSION:

To conclude, we report a patient with a rapid succession of symptoms including intractable vomiting and hiccups, TM and ON, most probably due to NMO spectrum disorder. NMO should be considered in the differential diagnosis of atypical CNS presentations such as intractable vomiting and hiccups and not be ignored as just a gastro intestinal cause. Recognition of this syndrome has significant implications as its treatment and prognosis differs from MS.

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