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Original Research Paper

Neurology

A SHORT JOURNEY OF N M O S D AND UNCOMMON PRESENTATIONS

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ABSTRACT The two uncommon manifestations APS and Brainstem syndrome overlap with APS along with common presentations of LETM and Optic neuritis of N M O Spectrum are described with their presentation and management.

KEYWORDS : N M O S D : Neuro Myelitis Optica Spectrum Diseases, APS : Area Postrema Syndrome, LETM : Long Extensive Trensverse Myelitis, AQ4: Aqua Porin 4 Antibodies, MOG : Myelin Oligo Dendrocyte Glycoprotein

INTRODUCTION:

(1) NMOSD are antibody mediated CNS Diseases. These diseases are diagnosed after discovery of positive antibodies to AQ 4 in 2004 and MOG Antibodies in 2012. AQ4 is located in the foot processes of astrocytes at blood brain barrier and MOG Antibodies are located on the surface of oligodendrocytes. AQ4 receptor antibodies destroy astrocytes and further damage oligodendrocytes and damage myelin which is a secondary process unlike MS. F:M =3:1 TO 9:1.

The spectrum consists of

1. Long segment myelopathy which involves more than 3 segments mostly central producing paraplegia or quadriplegia.
2. Optic neuritis predominantly posterior part and optic chiasma with acute diminution of vision.
3 Area postrema syndrome with presentation of vomitings and Hiccups with involvement of dorsal medulla.This is quite rare involving 10% of N M O S D**(2)**
4. Brain stem syndrome with an overlap of APS With other cranial nerve involvement.
5. Cerebral syndromes along with ADEM where MOG antibodies are mostly positive.
6. Acute diencephalic syndrome with thalamus and hypothalamus involvement with symptoms of Narcolepsy and diminished hypocretin levels.

75% of the N M O S D are AQ4 positive and 40 % of N M O S D With AQ4 negative show MOG positive.

Recurrence is more in AQ4 positive. Brain involvement may be seen in 60 % Of these cases. CSF May show mixed pleocytosis. These patients are well managed with pulse therapy of Methylprednisolone 1 gm per day for 5 days, followed by oral steroid in tappering dose over months.

Some patients respond well to plasmapheresis. We can treat with immuno suppressants like Mycophenalate and Azathioprine to prevent the recurrence.The second line drug if the patient doesn't respond is Rituximab to prevent recurrence of the disease.

Case Vignettes:

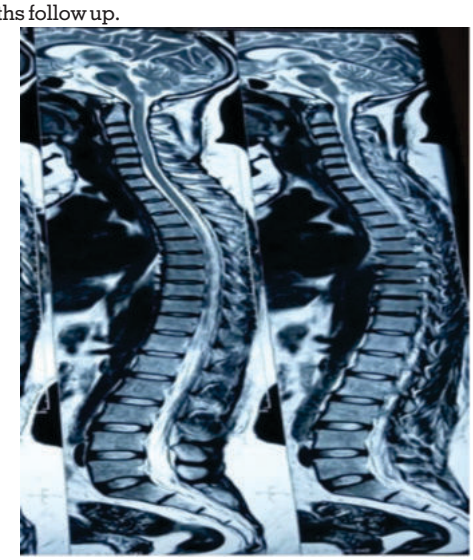
1.A 43 yrs old female presented with Reeling, vomitings and Hiccups since 4 days.History of swaying while walking and left sided numbness present.

Clinical examination showed Nystagmus,Hemiparesis 4/5 and hemihypaesthesia Lt side ,with extensor plantar response.

Her haematological, cardiac ,renal and Hepatic parameters and viral markers were normal.

Her MRI brain showed dorsal medullary hyper intense focus indicating Area postrema syndrome. Her CSF examination showed mild mixed pleocytosis. Her AQP4 is positive.

With the MRI, and Positive AQ 4 positivity the diagnosis of Area postrema syndrome was considered and treated with Inj Methylprednisolone 1 gm per day pulse therapy for 5 days. Patient showed good improvement and walked out of the hospital. She was kept on oral prednisolone in tappering dose along with Azathioprine. Patient is doing well during the last 5 months follow up.



AREA POSTREMA SYNDROME

Hyper Intense Lesion In The Dorsal Medulla - Area Postrema

2.A 30 yrs old female presented with acute onset of quadriparesis progressed over a period of 5 days.History of vomitings present.

Clinical examination showed

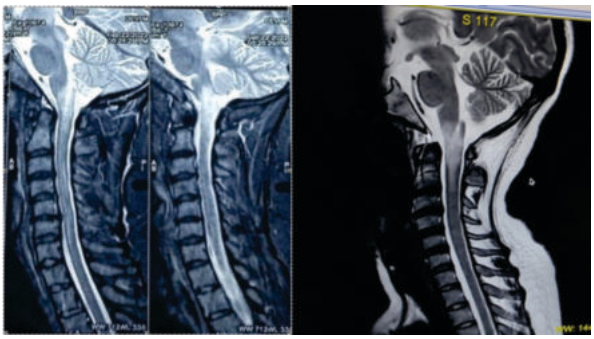
Dysarthria,9th,10th nerve palsy and quadriplegia with plantar bilateral extensor and impaired sensory below C3 C4. We considered the possibility of cervical myelopathy with extension to brain stem and evaluated.

Her routine investigations are normal. cardiac,renal , Hepatic and viral markers were normal.

MRI Brain and cervical spine showed Brainstem and upper cervical hyper intense lesion which suggested the possibility of Acute Brainstem syndrome with APS overlap. Her CSF examination showed mild pleocytosis.

She was started on Inj Methylprednisolone 1 gm per day for 5 days. She showed good improvement and could walk with out support in 2 weeks and walked out the hospital. She was kept

on Tappering dose of oral prednisolone and Azathioprine. Follow up after 1 month showed good improvement and walking alone without any support.



Cervico Medullary Syndrome With Super Added Aps.

Hyper Intense Lesion In The Medulla And Spinalcord And More Hyper Intense Lesion In The Dorsal Medulla.

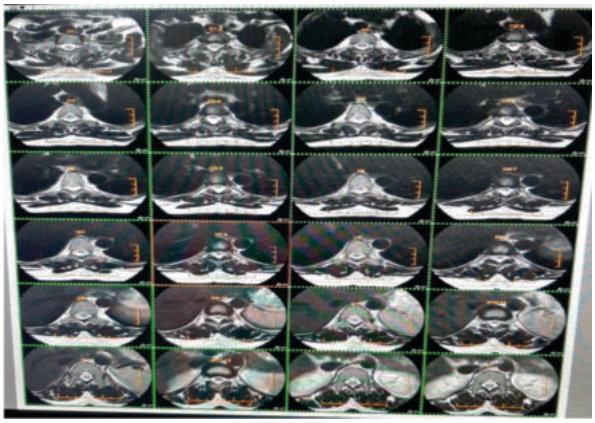
3.A 25 yrs old female brought with H/O weakness of both lower limbs since 3 days and numbness below mid chest Clinical examination showed normal cranial nerves and normal upper limbs.Lower limbs power 3/5 with Hyper reflexia ,extensor plantar bilateral and impaired sensation below D4.

Diagnosis of dorsal myelopathy was considered and evaluated. Routine investigations were normal. Her RFT ,LFT, Xray chest ,2DECHO and viral markers were normal. MRI Dorsal spine showed Hyperintense lesion extending from D1 to D8.

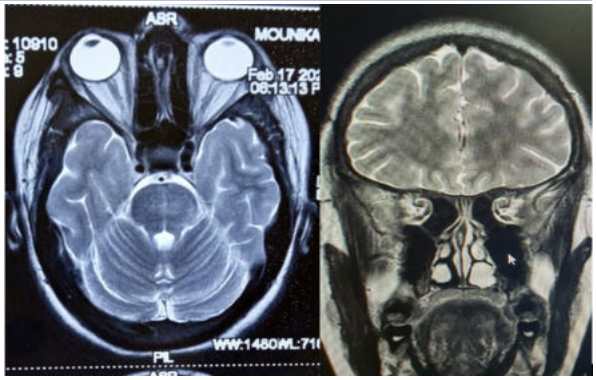
Her MOG antibodies were positive and AQ4 was negative. We considered the diagnosis of Long Extensive transverse myelitis and kept her on pulse therapy of Methylprednisolone 1gm per day for 5days. Patient showed good improvement in 1 week and able to walk without support in 2 weeks. Patient was discharged with Prednisolone tappering dose. She didn't come for follow up.

After 3 months the same Patient presented with diminished vision of acute onset in the left eye.Mild pain in the left eye was present .she stopped medication after her discharge. Her vision was counting fingers in the left eye and right eye was normal. Fundi were normal.MRI brain and orbits showed hyperintense focus in the left optic nerve.In view of the previous diagnosis of LETM we considered the diagnosis of Optic neuritis and started on IV Methylprednisolone 1gm per day for 5days. Patient showed progressive improvement and discharged with tappering oral cortico steroid along with Azathioprine.Patient regained near normal vision in 3 weeks. She is on regular follow up for the past 5 months and there is no recurrence of any symptoms.

LETM



HYPER INTENSE FOCUS FROM D1-D8.



HYPER INTENSE FOCUS IN THE LEFT OPTIC NERVE

DISCUSSION:

N M O S D are a group of diseases which are being diagnosed since the discovery of AQ4 in 2004 and anti MOG antibodies in 2012.Several case reports have been published since that time There are about 6 syndromes described in N M O S D. Out of these Optic neuritis and Long extensive transverse myelitis (LETM) are common. The other syndromes described are rare. We describe here AREA POSTREMA SYNDROME which accounts for 10% of N M O S D.The other one is still rare that is MEDULLARY SYNDROME WITH SUPER ADDED APS. The importance of the later syndrome is that there is an extension of the lesion to the cervical cord,making it CERVICO MEDULLARY SYNDROME WITH super added APS Which was not described earlier. These two cases were presented with all clinical manifestations and the treatment. The other common presentations ON and LETM were also presented with various clinical manifestations and management. The problem in last case is that the patient is lost for follow up and discontinued the medication and developed ON.During the second presentation also patient improved with treatment and there is no recurrence of the disease.The positivity of the antibodies is important in the diagnosis. The first two are AQ4 positive and the later is MOG antibody positive.MOG antibodies are positive in the involvement of anterior optic nerve contrary to AQ4 positivity in the posterior part and optic chiasma .It's known that the recurrence rate is high in the patients positive for AQ4 and response is better in MOG positive cases.We followed the criteria laid down by Winger Chuk etal for the diagnosis of N M O S D With positive antibodies .

Message:

The early diagnosis and management of the N M O S D gives good results and recurrence can be prevented.

REFERENCES ;

1. Royal college of physicians, clinical medicine
2. Neuro myelitis optica spectrum disorders ,Saif Huda etal
3. N MO Spectrum disorders
4. Dean M Winger Chuk and Claudia F Luchinetti MD , Aug 18 2022 387: 631-639