ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 11 | November - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

NEUROMYELITIS OPTICA SPECTRUM DISORDER (NMOSD) PRESENTING AS BRAINSTEM AND MULTIPLE LOWER CRANIAL NEUROPATHIES

Neurology

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ABSTRACT

Neuromyelitis Optica Spectrum Disorder (NMOSD) now recognized as autoimmune antibody mediated astrocytopathy is characterized by frequent relapses and disability. Previously thought to involve optic nerves and spinal cord only, now has an expanding spectrum of Neuraxis involvement. However brainstem, cranial nerves, peripheral nerves involvement has rarely been reported. We report a case of AQP-4 positive NMOSD presenting with Area postrema and bilateral CN IX, X, and XII palsies.

KEYWORDS

CASE REPORT

21year male without any comorbidity , addiction or any preceding illness presented with acute onset dysphagia,dyspnea and dysarthria which was more for liquids then solids, intractable hiccups and vomiting of 20 days duration.Patient was earlier admitted in medicine department and was reffered to us in view of intractable hiccups and vomiting after normal gastrointestinal work up for the same.On examination patient was drowsy but oriented to time place and person with Ryles tube insitu with drooling of saliva. General physical examination including blood pressure,pulse rate was normal except for tachypnea (RR =28) and bilateral crepitations in lower lobes of lungs on auscultation. Neurological examination depicted absent gag reflex, decreased sensation in uvula and posterior part of tongue, unable to protrude his tongue suggestive IX,X,XII palsies without any other focal neurological deficit.

Diagnostic work up of baseline investigations showed neutrophillic leucocytosis, rest of the parameters RFT/ LFT/ TSH/ B12/ HBA1C/ VIRAL MARKERS/ HIV were normal.Contrast MRI Brain with orbit suggested superoinferiorly elongated FLAIR/T2W hyperintense signal of dorsal aspect of medulla oblongata/area postrema without diffusion restriction or gradient susceptibility or odema or post contrast enchancement.MRI spine was normal. CSF examination was normal.VEP was normal.EMG of tongue suggested fibrillations and positive sharp waves suggestive of active dennervation changes Treatment with IV Antibiotics for aspiration pneumonia and Inj I/V methylprednisolone 1gm/day given for 5 days followed by oral prednisolone and symptomatic treatment. Patient showed improvement in his swallowing (able to take orally), speech and decreased frequency of hiccups. Antibody panel for Aquaporin 4 came to be strongly positive on day 5 of admission.For long term immunosupression patient was given Rituximab after treating aspiration pneumonia .Nature ,prognosis of illness and precautions were explained to patient.

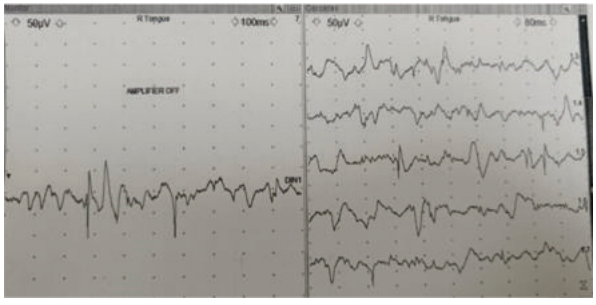
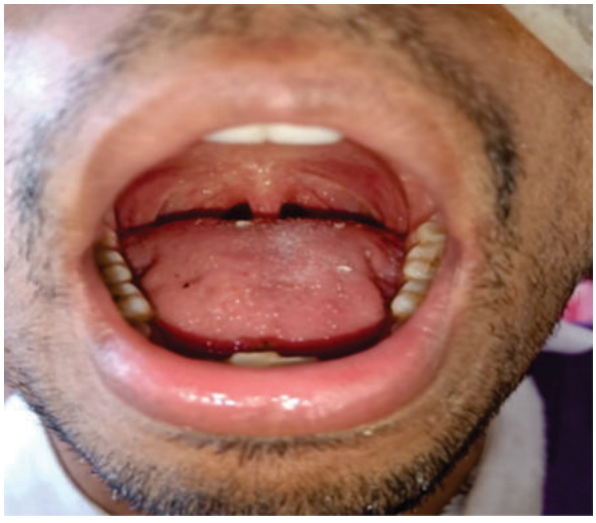


Figure 1 EMG Of Right Tongue Demonstrating Fasiculations And Positive Sharp Waves.



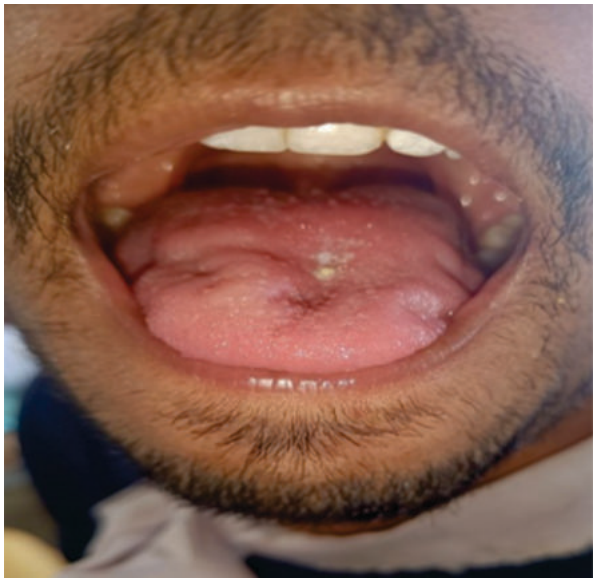


Figure 2 Demonstrating Downward Displaced Bilateral Uvula And Tongue Atrophy.

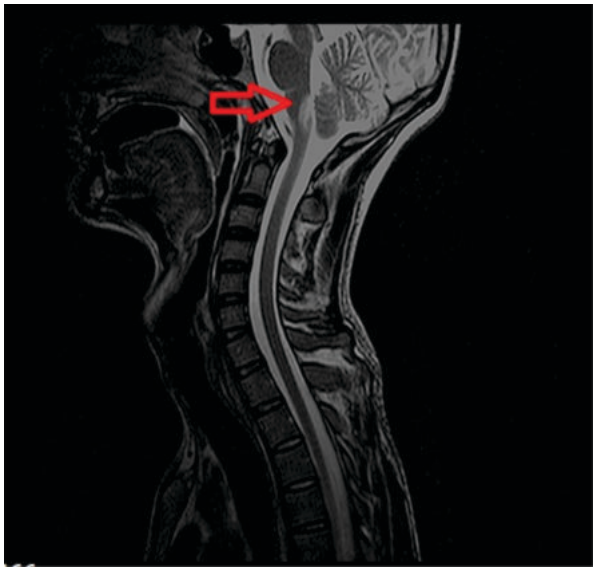


Figure 3 T2 MRI Saggital Section Demonstrating Dorsal Medullary/Area Postrema Hyperintensity

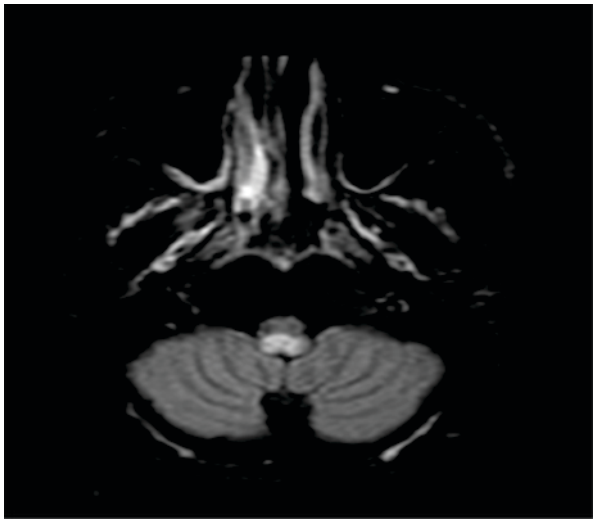


Figure3 T2 MRI Axial Section Of Brainstem Demonstrating Area Postrema Hyperintensity

DISCUSSION

Neuromyelitis optica spectrum disorders (NMOSD), previously referred to as neuromyelitis optica (NMO), was first described more than a century ago by Eugene Devic, for whom it was originally named. NMOSD is now recognized as autoimmune astrocytopathy that targets the waterchannel aquaporin-4 (AQP4)expressed on end-feet of astrocytes.AQP4-IgG plays a pathogenic role in astrocyticinjury. Complement and cell-mediated antibody-dependent astrocytedestruction is a primary event inNMOSD, which causes secondary loss ofoligodendrocytes and demyelination.[1,2,3]

NMOSD may manifest at any age,but predominantly affects middle-aged women.Recurrent attacks and event-related disability are common in NMOSD, which has an overall relapse rate between 60% and 98%.[4,5]85% of patients present with transverse myelitis (50%) or optic neuritis (35%) (10% with both)as their initial event, with a minority (4%) of patients manifesting other clinicalsyndromes at disease onset. The spectrum of neurologic, ophthalmic,systemic, and indeed psychiatric features that typify NMOSD continues toexpand.Optic neuritis,Transverse myelitis,pruritis; Area postrema syndrome,Brainstem syndromesDiencephalic syndromes,Cerebral syndromes Encephalopathy (acute disseminated encephalomyelitis [ADEM]-like), posterior reversibleencephalopathy syndrome (PRES),Acute myopathy withhyper Ckemia, Myeloradiculopathy, Neuropsychiatric Psychiatric manifestations,Uveitis.[6]Brainstem manifestations have rarely been described.In 2014 L kremer et.al conducted a multicenter study of 258 patients with NMO .Brainstem signs were observed in 81 patients (31.4%) like vomiting, hiccups, oculomotor dysfunction and other cranial nerve signs.[7].Cranial neuropathies like 9,10,12 have rarely been described except few case reports.[8]Our patient presented with dysphagia,dysarthria, intractable vomiting, hiccups and 9,10,12 cranial neuropathy

CONCLUSION

We present a case of 21 year male with symmetric bilateral 9,10,12 cranial nerve palsies, area postrema syndrome and strongly positive for NMO AQ4 antibodies.This case highlights the rare isolated presentation of NMOSD as multiple lower cranial neuropathy.Thus clinicians should keep in mind the varied spectrum of this disease for early suspicion,diagnosis and treatment.Hence avoiding the long term morbidity as early treatment has a better long term prognosis.Further longer observations and case reports will be worthwhile along this direction.

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