Case Report

Jaw clonus in neuromyelitis optica spectrum disorder with subsequent osmotic demyelination syndrome

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Abstract

Jaw clonus, a fascinating, yet uncommon clinical sign, is suggestive of supranuclear lesions of the trigeminal nerve. It has previously been reported in association with amyotrophic lateral sclerosis. Hereby, we report an index case of jaw clonus in a patient of neuromyelitis optica spectrum disorder with subsequent osmotic demyelination syndrome with pseudobulbar palsy due to the involvement of pontine corticobulbar fibres.

Keywords: Jaw clonus, neuromyelitis optica spectrum disorder, osmotic demyelination syndrome

Introduction

Jaw clonus is a series of rhythmic involuntary muscular contractions of the temporalis, masseter and medial pterygoid muscles occurring at a frequency of 5–7 Hz in response to an abruptly applied and sustained stretch stimulus. It typically indicates damage to the upper motor neurons in the corticopontine tracts. In patients with supranuclear lesions of the trigeminal nerve (e.g. amyotrophic lateral sclerosis [ALS], extensive multiple sclerosis [MS], bilateral corticobulbar infarction with pseudobulbar palsy) the jaw jerk is exaggerated and clonus may be observed.[1]

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Osmotic demyelination syndrome (ODS) is a life-threatening demyelinating syndrome, which usually occurs in the setting of rapid correction of severe chronic hyponatremia.[2] Central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM), a subset of ODS, most consistently presents with features of pseudobulbar palsy and spastic quadriplegia caused by demyelination of corticospinal and corticobulbar tracts either within the pons or extra-pontine brain tissue, respectively.[3]

Neuromyelitis optica spectrum disorders (NMOSD, previously known as Devic disease or neuromyelitis optica [NMO]) are inflammatory disorders of the central nervous system characterized by severe, immune-mediated demyelination and axonal damage predominantly targeting optic nerves and spinal cord. NMO may lead to hyponatremia due to severe vomiting resulting from lower brainstem involvement. Thereby, ODS remains a practical possibility in NMO.[4]

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Herein, we report an unusual case of NMO complicated by subsequent ODS that presented with jaw clonus due to pseudobulbar palsy.

Case Report

A 37-year-old rural male driver by occupation was referred to us from a block primary health centre (BPHC) with a diagnosis of an "unarousable coma with hyponatremia". History revealed that he had abrupt onset, persistent vomiting for the last 3 days, not responding to oral antiemetics. On the evening of the fourth day, he was found unconscious and seized in a tonic-clonic fashion in his own vehicle. After initial resuscitation at BPHC, he was found to have hyponatremia (Na 112 mEq/L) and the correction was attempted by hypertonic (3%) saline infusion. Initially, he showed some improvement in consciousness and complained of diminished vision in both eyes but gradually over the next few hours his sensorium further deteriorated and he was soon unable to move his limbs. He was referred to us in the morning on the fifth day of his illness.

Neurological examination revealed cloudy consciousness, florid emotional lability, dilated pupils with sluggish response to light and relative afferent papillary defect (RAPD), difficulty in deglutition and nasal regurgitation of liquids associated with cough and flaccid symmetric quadriplegia. All the reflexes were absent except jaw jerk and bilateral extensor plantar. Jaw jerk reveled jaw clonus [Video 1].

Initial laboratory investigations came out to be normal except mild hyponatremia (Na- 128 mEq/L). Magnetic resonance imaging (MRI) of the brain showed focal altered intensity lesions, hyperintense on T2, fluid-attenuated inversion recovery (FLAIR), diffusion-weighted image (DWI) and hypo- to isointense on apparent diffusion coefficient (ADC) with mild patchy contrast enhancement at pons, both middle cerebellar peduncles, crus of midbrain and bilateral basal ganglia [Figure 1]. MRI of orbit was suggestive of bilateral retrobulbar intraorbital optic nerve demyelination. Visual evoked potential (VEP) test revealed prolonged P100 wave latencies and normal P100-N70 wave amplitudes suggestive of the demyelinating type of bilateral retino-optic pathway dysfunction. Cerebrospinal fluid (CSF) study with paired sera ruled out infectious processes, autoimmune encephalitis. CSF anti-NMO/aquaporin 4 (AQP4) came back negative but serum anti-NMO antibody was found to be positive in high titers along with a few oligoclonal bands. MR spine, MR angiography and DSA were normal.

On the basis of classic clinical manifestations, lesions in MRI brain and AQP4 positivity, the patient was diagnosed as NMOSD.[5] We hypothesized that NMO flare-up caused the intractable vomiting due to brainstem affection without radiological involvement of area postrema, and hypothalamus (considering that 1.5 T MR may not always record the subtle signal changes). However, a history of rapid correction of hyponatremia, symmetrical pontine and extra-pontine signal changes without much contrast enhancement was strongly suggestive of concomitant ODS.

The patient was treated promptly with 5 days of 1 gm/day intravenous methylprednisolone followed by oral corticosteroids (1 mg/kg) and intravenous immunoglobulin (IVIG). The patient was treated for hypovolemic hyponatremia with very slow sodium

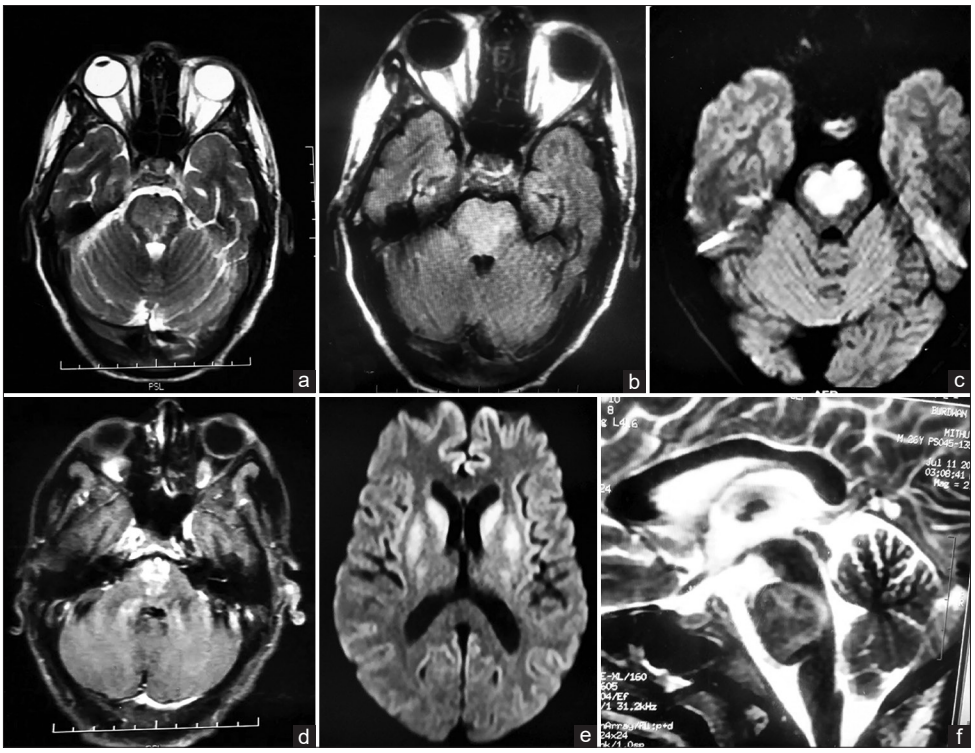

Figure 1: MRI brain reveals focal altered signal intensity lesions hyper on T2 (a and f), FLAIR (b), DWI (c and e) at pons, both middle cerebellar peduncles and crus of the midbrain and bilateral basal ganglia. Contrast imaging reveals patchy enhancement at the pons (d)

replenishment with fluid resuscitation until his serum sodium level normalised after 5 days. During the course of IVIG, his vision improved, emotional lability started abating and after 6 weeks he was able to identify colors correctly. Limb power also improved gradually and deep tendon reflexes returned. Azathioprine (50 mg twice daily) was started to prevent further relapses.

Discussion

AQP4 channel dysfunction in astrocytes mediated by autoantibodies targeting AQP4 channels has been the etiopathogenic mechanism for both NMO and ODS.[6] The destruction of AQP4 channels has been established in astrocytes in autopsy specimens of lesions of ODS.[7] There are at least two reasonably possible mechanisms by which ODS can occur in NMO: a) co-occurrence of ODS and NMO i.e. immune-mediated astrocyte dysfunction brought about by autoantibodies against AQP4 water channel in NMO as well as ODS, b) NMO leading to metabolic perturbation, fast change in serum osmoles along with hyponatremia because of either severe persistent and intractable vomiting due to area postrema involvement (hypovolemic hyponatremia) or the syndrome of inappropriate antidiuretic hormone (SIADH) due to hypothalamic lesions (euvolemic hyponatremia). Both of these can give rise to oligodendrocyte damage and thus demyelination resulting in ODS on their own or during treating them. ODS has also been described, even though rarely, in association with NMO.[4,8]

Jaw clonus, although a fascinating neurological sign, is scarcely reported in the medical literature. Beevor first described a case of amyotrophic lateral sclerosis with clonus of the lower jaw way back in 1886.[9] Since then few reports of jaw clonus have been reported in association with ALS.[1,10-12] In this rare case (probably the index one) of jaw clonus in a patient of NMO with subsequent ODS with the possible reason was pseudobulbar palsy due to demyelination of pontine corticobulbar fibres secondary to ODS.[13]

Conclusion

NMO should be kept in mind in all patients coming with a complaint of acute intractable vomiting. Hyponatremia can be the initial presenting feature of anti-AQP4 positive individuals with or without any concomitant optico-spinal involvement. These patients comprise that group of NMO-SD presenting with an acute diencephalic or brainstem syndrome. Treating hyponatremia can sometimes result in the rather formidable osmotic demyelination syndromes, EPM and CPM. It is well known that ODS presents with features of pseudobulbar palsy. However, jaw clonus even though being a manifestation of pseudobulbar palsy is an extremely rare clinical finding and such presentations of NMO-SD have not been reported previously in the literature. Jaw clonus occurs due to bilateral pyramidal tract involvement is primarily seen in MS, ALS, bilateral stroke and brain stem demyelination. This finding, although rare, is very much pathognomonic for suspicion of the above-mentioned conditions, and can easily be extrapolated in primary care level by a simple examination of jaw jerk and early referral to higher centre.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflict of interest

There is no conflict of interest.

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