MYASTHENIA MIMICRY

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CASE REPORT

A man in his late 60s with a medical history of hypertension, stage 3 chronic kidney disease and renal stones presented to the A&E department of a DGH reporting a 3 day history of a constant headache, an unsteady gait and double vision. He reported feeling unwell for a week prior to his presentation with difficulty swallowing and a cough, consulting his GP for an upper respiratory tract infection. He was otherwise usually well with a clinical frailty score of 3, although he reported an unintentional weight loss of 8 stone in the past year. Initial examination revealed mild confusion, AMT 8/10, gross opthalmoplegia, a wide based gait and a negative Romberg's test with no limb ataxia. He also reported pins and needles in his upper limbs. Shortly after arrival, he became floridly confused and agitated, with marked intermittent stridor particularly upon lying flat. An blood gas demonstrated severe hypercapnic respiratory failure and he rapidly deteriorated into a broad complex tachyarrthymia requiring x3 DC cardioversion and an IV antiarrthymic. He was then intubated for airway protection and to facilitate further investigations.

Initial differentials included obstructive upper airway/ upper GI lesions, myasthenia and brainstem pathology. Autoantibodies were sent for GQ1B , myasthenia and VGCC. A nasoendoscopy and OGD revealed normal anatomy. Brainstem lesions were excluded with MRI imaging, with no abnormal signals nor restricted diffusion seen. CSF analysis showed albuminocytological dissociation, with elevated protein but no white cells. An examination by the neurologist revealed ophthalmoplegia, bulbar failure, limb weakness with preserved reflexes and flexor plantar responses. Along with his history of possible viral prodrome and CSF results, his clinical presentation was felt to be in keeping with a working diagnosis of a brainstem encephalitis; a GQ1B mediated Guillain Barre variant. Subsequently, peripheral nerve conduction studies met the EDX 'probable' criterion for diagnosis of GBS[1] and also demonstrated low amplitude sensory action potentials. A 5-day course of intravenous immunoglobulins was commenced after which a full body CT later excluded thymic pathology and underlying malignancy.

Throughout his critical care admission, he required only minimal ventilatory support and responded appropriately with sedation breaks. Two trials of primary extubations were performed on days 3 and 6 post IVIG commencement; both of which he obstructed his airway within the first 10-15 minutes, receiving a course of dexamethasone to reduce airway oedema each time. He was considered for a tracheostomy wean but successfully maintained his airway on his final trial of extubation 11 days post IVIG commencement. He remained opthalmoplegic, dysphonic and failed swallow assessments after but made a steady improvement in the subsequent days; graduating to thickened fluids and yoghurts and regaining full extraocular eye movement.

10 days later, his bulbar symptoms regressed and he experienced a choking episode. A repeat nasoendoscopy demonstrated mild palsy and significant muscle tension dysphonia. He was re-referred back to neurology and was restarted on a second course of IVIG. His autoantibody results later returned grossly positive for anti-acetylcholinesterase receptor antibodies at 460 x10^-10mol/l and a repeat electrophysiology study of his facial muscles demonstrated severe neuromuscular transmission defect, confirming a diagnosis of late onset oculobulbar myasthenia. Anti GQ1B, VGCC and GM1 were all negative. He was started on prednisolone, pyridostigmine and propantheline bromide and was successfully discharged back home after 2 months inpatient stay.

DISCUSSION

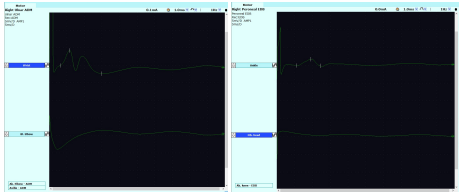
Bickerstaff's Brainstem Encephalitis (BBE) is a rare GQ1b-mediated polyneuropathy, considered to be a brainstem variant of Guillain Barre and Miller Fisher Syndrome (MFS). It is a condition with generally good outcomes, classically presenting with opthalmoplegia and ataxia but is differentiated from MFS by an altered sensorium with intact reflexes. In a cohort of patients with BBE, rates of bulbar palsy has been reported to be over 30% [2]. Other commonly described features include a monophasic illness course, history of an antecedent infection, and peripheral sensory involvement. Myasthenia gravis (MG) is a chronic autoimmune condition primarily characterised by muscle weakness and fatiguability. Subtle, fluctuating symptoms can result in a diagnostic delay. Myasthenia is an important cause of neuromuscular respiratory failure - termed myasthenic crisis. Signs of an imminent myasthenic crisis include rapid progression of myasthenic and bulbar symptoms[3], as seen in our patient. There were mimicry of several key features which such as ocular + bulbar symptoms but with a reported history of an illness prodrome and ataxia which were compatible with a brainstem pathology[4]. As both conditions respond to IVIG, the recurrence of symptoms was our first hint of an alternative diagnosis to BBE. In this case, dexamethasone courses given for airway oedema may have masked and slowed the recurrence of his symptoms.

The concurrent presentation of both stridor and his postural worsening of symptoms suggests of a concurrent diaphragmatic and bulbar palsy whilst his history of weight loss with subsequent negative investigations for malignancy may relate to a more insidious course of tongue and palatal weakness. Single fibre EMG was not initially performed due to working diagnosis of BBE, but is more sensitive for MG than standard NCS[5]. His low amplitude sensory action potentials could be coincidental neuropathy, due to critical illness/ immobility or immune-mediated, or due to technique. Similarly, his ataxic gait could have various confounding factors including his sensory and vision deficits.

This case highlights the overlap of key GQ1B and myasthenia features, demonstrating the challenges of distinguishing them in clinical practice. Both pathologies have a high rate of treatment success, with potentially fatal respiratory complications if not identified early.

Day 16 PERIPHERAL STANDARD NERVE CONDUCTION STUDIES (NCS)

Ulnar and peroneal motor response were normal distally but not seen on proximal stimulation, suggesting a degree of conduction block with normal conduction velocities.



Low amplitude sensory action potentials

Right Sensory Sural

Sites	Onset Lat [ms]	Peak Lat [ms]	Amp [µV]	Dur [ms]	Dist [mm]	CV [m/s]
Calf - Ankle	3.15	3.59	0.70	1.07	140	44.4

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