

Myasthenia mimicry

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Background

Bickerstaff's encephalitis is a rare GQ1b-mediated polyneuropathy, considered to be a brainstem variant of Guillain-Barré syndrome and Miller Fisher syndrome. It is classically differentiated from Miller Fisher syndrome by an altered sensorium but similarly demonstrates ophthalmoplegia and ataxia. Other commonly described features include peripheral involvement and bulbar weakness. Myasthenia gravis is more common, a chronic autoimmune condition primarily characterised by muscle weakness and fatigability, and is an important cause of neuromuscular respiratory failure.

Case presentation

This case reports a man aged in his late 60s who presented with dysphagia to solids and headaches, double vision and an unsteady gait. He had a history of an antecedent viral upper respiratory tract illness a week prior to the onset of his symptoms. On arrival at the emergency department, he was assessed to be ataxic, ophthalmoplegic and was mildly confused (scoring 8/10 on AMT-10) but with no other neurological signs. Shortly after arrival, he became floridly confused and agitated with marked intermittent stridor. An arterial blood gas demonstrated severe respiratory acidosis secondary to hypercapnic respiratory failure. He was intubated for airway protection and subsequent investigations including direct visualisation of his vocal cords, oesophago-gastro-duodenoscopy, computed tomography and magnetic resonance imaging all did not find a cause for his symptoms. Cerebro-spinal fluid (CSF) analysis showed albuminocytological dissociation and nerve conduction studies demonstrated absent proximal motor responses and possible conduction block.

The combination of ataxia, altered sensorium, ophthalmoplegia, bulbar palsy, nerve conduction and CSF results prompted an initial working diagnosis of brainstem encephalitis, Bickerstaff's: a GQ1B-mediated Guillain-Barré variant. Autoantibodies were sent for GQ1B, myasthenia and voltage-gated calcium channels (VGCC).

He had a stormy clinical course with two failed primary extubations in his first week of presentation, and seeming improvement post-intravenous immunoglobulins (IVIG), with a recurrence of his symptoms 1 month later. A second IVIG course was restarted and the autoantibody results were found later positive for anti-acetylcholine receptor, confirming a diagnosis

of late onset myasthenia. Anti-GQ1B, VGCC and GM1 were all negative.

Key points

This case critically highlights the overlap of key GQ1B and myasthenia features and demonstrates the challenge of distinguishing between them in clinical practice. The concurrent presentation of both stridor and his postural worsening of symptoms is suggestive of both a diaphragmatic and bulbar palsy while his history of weight loss with negative investigations may relate to a more insidious course of tongue and palate muscle weakness. ■

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