Brain stem compression by a giant vertebrobasilar aneurysm mimicking seronegative myasthenia

J Frisby, A Wills, T Jaspan

Abstract
A patient is described with a vertebrobasilar aneurysm who was erroneously thought to have myasthenia gravis on the basis of the clinical presentation and investigations, which were interpreted as supportive of a disorder of the neuromuscular junction. Despite the correct diagnosis being made at a late stage the patient made a full recovery after radiological intervention.

Keywords: myasthenia gravis; aneurysm

Case report
A 19 year old man presented with a 12 month history of intermittent double vision, dysphagia, limb weakness, and weight loss. He had previously been fit and well. On examination he had a full range of eye movements but variable diplopia in horizontal gaze. Horizontal nystagmus was noted at the extremes of gaze. He had bilateral lower motor neuron facial weakness with a bulbar dysarthria and normal palatal and facial sensation. Tongue movements and shoulder shrug were normal. His limbs were not demonstrably weak at the bedside. Tone and reflexes were normal and both plantar responses were flexor. There was no evidence of limb or gait ataxia.

The symptoms and signs were thought to be consistent with myasthenia gravis. A tension test (10 mg edrophonium preceded by 0.6 mg atropine intravenously) resulted in resolution of his diplopia and was interpreted as positive. This was not performed in a double blind fashion. He was started on pyridostigmine and reported moderate symptomatic improvement.

Subsequently, EMG of the right abductor digiti minimi with repetitive stimulation showed a 2% decrement at rest, increasing to a 9% decrement 3 minutes after isometric exercise. The patient had taken a dose of pyridostigmine 2 hours before the procedure. Single fibre EMG was not performed. Acetylcholine receptor antibodies were absent. Thorax CT was normal.

His symptoms deteriorated and he was started on an increasing dose of steroids. He reported symptomatic improvement on 60 mg prednisolone daily lasting for 2 weeks; however, bulbar function again became impaired. By this stage he had developed nystagmus, tongue fasciculation, ataxia, and sustained clonus at both ankles. Plantar responses were still flexor. A cranial MRI disclosed a large vascular lesion producing high grade brain stem compression (fig 1). Cerebral catheter angiography showed a fusiform vertebrobasilar aneurysm, involving the left posterior inferior cerebellar artery (PICA) origin and extending cranially to the level of the anterior inferior cerebellar arteries.

His bulbar function continued to deteriorate and he was intubated, ventilated, and a feeding gastrostomy inserted. Six weeks later he was well enough for radiological intervention. Under local anaesthesia, full heparinisation, and close neurological assessment, the left vertebral artery was temporarily occluded. Retrograde filling of the left PICA via the basilar artery was confirmed on internal carotid angiography. The trial occlusion was well tolerated and after 30 minutes, the vessel was permanently sacrificed using two detachable silicone balloons placed immediately below the aneurysm and in the distal cervical segment of the vessel.

The patient made a rapid recovery and 6 months after the radiological procedure he was asymptomatic and neurologically normal.

Discussion
Seronegative myasthenia gravis occurs in about 15% of patients and can lead to diagnostic confusion. The tensilon test has been used as a diagnostic “gold standard” in myasthenia.
myasthenia gravis but concerns about safety combined with increasing recognition that false positive results occur have led to a decline in its use. Performing the tension test in a blinded fashion may protect against false positive results but is relatively time consuming. In addition the common associated side effects (eye watering and twitching, abdominal cramp) make this impractical in many settings. Our patient is extraordinary in that he showed a response to pyridostigmine as well as steroids. Presumably the response to pyridostigmine was a placebo effect and the prednisolone led to a decrease in oedema around the aneurysm. The EMG abnormalities on repetitive stimulation were interpreted as consistent with myasthenia gravis. Decrements of 10% are usually considered pathological but this is a relatively non-specific phenomenon that can occur in many other conditions. Where possible, neurophysiological studies should be performed with the patient off pyridostigmine as this can lead to diagnostic confusion. Single fibre EMG may be a useful ancillary investigation to repetitive stimulation where diagnostic doubt exists. It is also of interest that despite marked brain stem compression our patient did not develop palatal sensory abnormalities in the context of severe bulbar dysfunction.

Permanent occlusion of a vertebral artery (or arteries) is recognised as the optimal mode of treatment for vertebrobasilar aneurysms that are not amenable to endovascular parent vessel sparing techniques (such as coil occlusion or stenting), or are deemed inoperable. In our case the aneurysm incorporated the PICA origin. Thus, treatment was aimed at occluding the vertebral artery proximal to the aneurysm (eliminating antegrade flow into the aneurysm), inducing aneurysmal thrombosis but maintaining flow into the PICA by reversing the flow in the distal segment of the vertebral artery. The sum effect created by a large vascular demand in the PICA territory ensures preservation of flow in the vessel via retrograde supply from the internal carotid artery.

Despite the correct diagnosis being made at a late stage, the patient made a full recovery after parent vessel sacrifice, which is now the treatment of choice in vascular lesions of this nature.

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