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Chiari I malformation mimicking myasthenia gravis

Chiari I malformation is accompanied by a variety of symptoms and signs suggesting brain stem, cerebellar, or cervical spinal cord lesions. The most common symptoms include headache, neck pain, sensory loss, and ataxia.[1] Dysphagia occurs in 5–15% of the patients and it may be the only presenting symptom.[2] Progressive dysphagia caused by Chiari I malformation, mimicking amyotrophic lateral sclerosis, has been reported in this journal in 1996 and 2002.[2 3] Dysphonia may occur rarely, but it has not been described as an early symptom.[2] Pain and stiffness in the posterior neck is a common feature, but severe neck extensor weakness leading to dropped head syndrome has not so far been reported in Chiari I malformation.

Case report

A 13 year old girl was admitted to our department of neurology four weeks after adenoidectomy under general anaesthesia, because of progressive difficulty in lifting her chin off her chest, together with dysphagia and dysphonia. There was no pain or stiffness in the posterior neck. Computed tomography of the head was reported as normal. There was mild fluctuation of the dysphagia and dysphonia during the day, with worsening of the dysphonia after prolonged conversation or after reading in a loud voice. There was no sleep disturbance.

Neurological examination revealed an increase in the deep tendon reflexes in all four limbs. Routine serum biochemistry and blood count were unremarkable.

On the basis of the history and clinical data, myasthenia gravis was suspected, so electrophysiological testing was undertaken to investigate the neuromuscular junction. Percutaneous 3 Hz repetitive nerve stimulation of the right accessory nerve along with recordings obtained from surface electrodes over the trapezius muscle did not show significant variations in compound muscle action potential amplitude under baseline conditions, or when the test was repeated three minutes after maximal voluntary effort for 30 seconds. Single fibre electromyography of the right extensor digitorum communis muscle during voluntary activity failed to show any abnormally increased jitter or neuromuscular block, and the mean jitter value was in the normal range. Serum antiacetylcholine receptor antibodies were absent. Standard concentric needle electromyography of proximal and distal muscles in all four limbs was normal. The cervical paraspinal muscles were not investigated. Motor and sensory conduction velocities were normal. A neostigmine test resulted in mild improvement in the dysphonia.

One week later, the patient experienced a worsening of dysphagia and dysphonia and she started to complain of gait disturbances, with instability in walking. Magnetic resonance imaging of the brain and cervical spine showed herniation of the cerebellar tonsils through the foramen magnum, reaching C2–C3 vertebral level (fig 1). No syrinx or hydrocephalus was demonstrated. A diagnosis of Chiari I malformation was made.

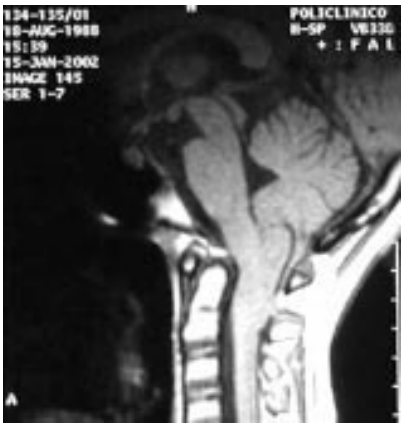

Figure 1 Brain and cervical spine magnetic resonance imaging: herniation of the cerebellar tonsils through the foramen magnum.

A posterior fossa craniectomy was undertaken. The anaesthesia and operative procedure were uncomplicated. Two days after the operation, the dysphagia and dysphonia improved and one month later there had been a remarkable improvement in the neck extensor muscle weakness.

Comment

Dropped head may be a part of a generalised neuromuscular disorder, such as myasthenia, polymyositis, amyotrophic lateral sclerosis, adult onset nemaline myopathy, or chronic inflammatory demyelinating polyneuropathy. Our patient had a dropped head "plus" syndrome secondary to Chiari I malformation. Strangely, the neck pain and stiffness were not referred. This case report suggests that one should suspect Chiari I malformation in patients with neck extensor muscle weakness, especially if this is associated with lower cranial nerve impairment. We postulate that the symptomatology in this girl may have been the result of brain stem dysfunction secondary to the compression caused by the malformation. Dysfunction of the lower cranial nerves and the higher cervicospinal roots by a retrograde effect of the compression may be the pathogenic explanatory mechanism. The rapid disappearance of the symptoms after posterior fossa decompression supports this hypothesis. Fluctuations of dysphonia and dysphagia may, on the other hand, reflect variations in intracranial pressure. Recently, a presentation of a previously asymptomatic Chiari I malformation was reported following a flexion injury to the neck by a trivial car accident.[4] In our patient, it is possible that slight cervical trauma during anaesthesia for her adenoidectomy may have brought to light the underlying congenital abnormality.

Moorty *et al* have reported eight cases, initially diagnosed as ocular myasthenia on the basis of clinical features and response of anticholinesterase agents, in which an intracranial mass lesion instead of or in addition to myasthenia gravis was later found.[5] Patients with dropped head and lower cranial nerve involvement, although presenting with a clinical history strongly suggestive of myasthenia, should be carefully evaluated and the diagnosis of Chiari I malformation considered. A response to anticholinesterase agents observed clinically or recorded electrically has been reported in a variety of disorders, including Eaton–Lambert syndrome, botulism, and transverse myelitis, and even in patients with intracranial mass lesions.[5] The partial response to anticholinesterase drugs in our case reinforces the view that it is unwise to base the diagnosis of myasthenia gravis purely on a positive pharmacological test.

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Expanding cerebral cysts (lacunae): a treatable cause of progressive midbrain syndrome

A progressive motor defect presenting in adulthood is an ominous sign, being often associated with either neoplasia or neurodegenerative diseases. Notable if very rare exceptions to this poor prognosis are cerebral expanding lacunae or, as they are sometimes called, benign intraparenchymal brain cysts.[1] These are intraparenchymal cavities without an epithelial lining, filled with cerebrospinal fluid (CSF), located in the thalamo-mesencephalic arterial territory.[1 2] Their expanding nature is demonstrated by their progressive clinical course and by the frequent complication of aqueduct stenosis and triventricular hydrocephalus.[2–4]

We present a case of progressive midbrain syndrome associated with expanding cysts, which was successfully treated by neuroendoscopy.

Case report

A 43 year old woman with an unremarkable clinical history presented in 1996 with progressive resting tremor and weakness of the left arm. The tremor persisted during posture maintenance and action. Within a year the motor problems extended to the left leg. Brain magnetic resonance imaging (MRI) showed large (10 to 20 mm) well defined lesions with signal intensities identical to CSF occupying most of the right thalamo-mesencephalic region. There was no contrast enhancement either in the lesions or in the surrounding tissue. The ventricular spaces were only mildly