Spasmodic torticollis due to neurovascular compression of the 11th nerve

Case report

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An unusual case of spasmodic torticollis caused by posteroinferior cerebellar artery compression of the spinal accessory nerve is reported. The spasmodic torticollis was cured by abolishing the neurovascular compression.

KEY WORDS • spasmodic torticollis • neurovascular compression • spinal accessory nerve • postero-inferior cerebellar artery

Spasmodic torticollis is a neuromuscular disorder characterized by involuntary spasm of various muscles. The symptoms usually begin on one side. It has been defined as a rotated position of the head caused by clonic or tonic contractions of the cervical muscles, and is a symptom of either an organic disease of the nervous system or hysteria. Both clonic and tonic contractions may occur in the same patient.10,21

Patients suffering from spasmodic torticollis have always presented a problem for the psychiatrist, neurologist, or neurosurgeon, especially when they do not respond to treatment. Conservative therapy usually offers little or no improvement. Although surgical therapy gives better results,1 some side effects must be expected. Two operative procedures are currently used: rhizotomy of the cervical anterior roots, combined with section of the spinal accessory rootlets (the Förster-Dandy operation),7,10,20 and a stereotaxic ventrolateral thalamotomy and subthalamotomy.2,9,11,19 Both methods are frequently combined with peripheral operations, such as section of the accessory nerve or the sternocleidomastoid muscle.

The pioneer work of Dandy6 and Gardner,8 relating to compression at the root entry zone of the trigeminal nerve by vascular loops as a cause of trigeminal neuralgia,13 formed the basis for current concepts of neurovascular compression in other cranial nerve disorders.14-18 To our knowledge, spinal accessory nerve dysfunction due to neurovascular compression as a cause of spasmodic torticollis has not been previously reported.

Case Report

This 68-year-old woman had for 5 years suffered from painful spasmodic tilting of the neck to the left and elevation of the left shoulder from arrhythmic paroxysmal contractions of the left trapezius, sternocleidomastoid, and splenius capitis muscles. During the spasms, the head rotated to the right and the left shoulder was elevated to the ipsilateral mastoid process. At first, the spasms were short-lived, but later they became more frequent and lasted longer (30 to 90 seconds). To treat these symptoms, the patient underwent surgery elsewhere, consisting of an anterior interbody fusion at the C5–6 level and resection of the left sternocleidomastoid muscle. This procedure did not afford relief.

Examination. On admission, the patient’s spasms were very frequent, occurring every few minutes. The neurological examination was normal. Plain x-ray films of the skull and computerized tomography scans with contrast enhancement were noncontributory. X-ray films of the cervical spine revealed diffuse spondylosis and the C5–6 interbody fusion. Vertebral angiography was not performed because of the patient’s age. Electromyography with the patient at rest revealed motor activity occurring paroxysmally and simultaneously with the spasms in the left trapezius, sternocleidomas-
toideus, and splenius capitis muscles. Rhizotomies of the 11th nerve and anterior cervical roots, according to the technique of Förster and Dandy, were planned.

Operation. A loop of the left posterior inferior cerebellar artery (PICA), which was almost as large as the vertebral artery, was found impinging on the left accessory nerve, compressing and distorting its rootlets. To separate the nerve from the artery it was necessary to cut some of the lower rootlets of the left accessory nerve and some of the C-2 and C-3 rootlets (Fig. 1). Other rootlets were left intact.

Postoperative Course. The patient's postoperative course was uneventful and her symptoms were fully relieved. On examination 24 months later, she was left with rare sporadic contractions of the lower part of the left trapezius muscle, with a hardly visible elevation of the shoulder and slight bending of the neck.

Discussion

It has been suggested that chronic changes of root nerve fibers by vascular compression may be the cause of pain paroxysms of tic douloureux. Vascular root compression has been reported as a cause of paroxysmal sensory or motor disturbances in other cranial nerves, including the seventh nerve (hemifacial spasm), vestibular branch of the eighth nerve (Ménière's disease), auditory branch of the eighth nerve (tinnitus), and the ninth and 10th nerves (glossopharyngeal neuralgia). It has been demonstrated that, in a peripheral nerve trunk, an ectopic (ephaptic) impulse generator may develop at the level of lesion in continuity from local demyelination.

Alterations in the accommodative and adaptive properties of the repetitive firing mechanism at a site of chronic injury create the opportunity for the region to serve more effectively as a mechanoreceptor or chemoreceptor, and also enable antecedent activity in the axon or adjacent axons to serve as a stimulus for afterdischarge. Two types of discharge are seen in lesions in continuity. In the simpler case, brief stimulation of an isolated segment of peripheral nerve may cause the axon to exhibit repetitive firing for several minutes following the cessation of the stimulus train. Another form of discharge is the reflected impulse; an impulse propagated through the lesion in continuity may give rise to a second impulse that is directed backward, as if "reflected." Moreover, prolonged discharges might be triggered by a reverberating short circuit between afferent and efferent fibers in damaged nerve roots. In other words, afferent activity could trigger sustained discharges from a region of chronic injury. Regions of patchy demyelination and hypermyelination have been observed in the trigeminal nerve root of patients with trigeminal neuralgia. These are thought to be likely reexcitation sites triggering sustained afterdischarge that is responsible for pain paroxysms.

There are similarities between hemifacial spasm and spasmodic torticollis. The neurophysiological mechanism of both disorders might be summarized in the afterdischarge phenomenon, where the impulse trains outlast the stimulus. Jannetta stressed that, in cases of hemifacial spasm or trigeminal neuralgia, the vascular compression must be at a right angle to the nerve, and the artery must compress the root entry zone of the nerve near the brain stem. He added that a junctional zone defect, which exists at the root entry zone, may predispose to conduction disorders when compression exists at that point.

The close contact between a large vessel and the accessory nerve rootlets found at the operation in our patient could have caused a focal demyelination with self-sustaining impulse generation as suggested for tic douloureux and hemifacial spasm. In fact, once the neurovascular compression was alleviated, the spasmodic torticollis subsided. Thus, in spite of the fact that the vascular compression was not near the bulb or spinal cord, but far from the root entry zone, it is conceivable that it was the cause of paroxysmal activity. Unfortunately, to avoid further contact between the PICA and the spinal accessory nerve, it was necessary to cut some of the lower rootlets of the 11th nerve and some C-2 and C-3 rootlets, but most of them were left intact.
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To our knowledge this is the first case of spasmodic torticollis in which neurovascular compression was observed at the operation. The disorder was cured by abolishing the compression, without complete section of the accessory nerve or anterior cervical roots. Thus, the etiology may have been the neurovascular conflict between the PICA and the spinal accessory nerve. Further clinical observation and anatomical and electrophysiological investigations are necessary to confirm this hypothesis.

References


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