Movement Disorders
CLINICAL PRACTICE

CLINICAL VIGNETTE

Cervical Dystonia Following Injury to the Cerebellar Pontine Angle: An Instructive Case

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A 38-year-old woman presented with cervical dystonia in the context of a recent surgery to remove a vestibular schwannoma. She initially presented to neurology with pain in the right arm, and MRI of the brain showed an incidental right-sided vestibular schwannoma (Video 1, Segment 1). An elective gamma-knife procedure was performed, which failed. Hydrocephalus requiring ventriculoperitoneal shunt insertion developed, and 3 years following the initial procedure the lesion was surgically excised. Surgery was further complicated by right middle cerebellar peduncle injury, extending to the cerebellopontine angle and marginally to the right pontine tegmentum, with subsequent mass effect on cerebellum displayed on follow-up MRI (Video 1, Segment 2). Six months later, the patient experienced forced head deviation to the right, with difficulty moving from this position. Examination revealed clear right-sided torticollis, with hypertrophy of the left sternocleidomastoid muscle. Cervical dystonia worsened with action and nearly resolved with the patient lying down. A clear *geste antagoniste*, where symptoms improved with the patient touching the side of her head, was present (Video 1, Segment 3). Findings consistent with injury to the cerebellar pathways were additionally exhibited. She demonstrated clear dysarthria, bilateral dysmetria, dysdiadochokinesia (worse on the right), and prominent gait ataxia (Video 1, Segment 4). Although a possible role of the schwannoma itself in the cervical dystonia pathogenesis cannot be entirely ruled out, the timing of signs, occurring soon after the postsurgical injury, suggest a prominent involvement of structures lying within the cerebellar pontine angle.

Cervical dystonia is the most common focal dystonia, characterized by sustained involuntary contractions of cervical musculature.[1] Traditional views of dystonia pathophysiology focus predominantly on the basal ganglia.[2] However, recent literature and imaging studies suggest that cerebellar and brainstem pathways are prominently involved and dystonia is now considered a "network" disorder, within which several structures have been implicated.[3,4]

Dystonic features are also shown to improve with lesions of thalamic relay nuclei linking the cerebellum and striatum.[3] This suggests aberrant inputs from cerebellar pathways to the basal ganglia contribute to dystonia pathophysiology. Cervical dystonia, in particular, has been associated with cerebellar pathway lesions more than other focal dystonias.[4] In fact, a case series of 25 patients showed that secondary cervical dystonia was more often attributed to lesions in the cerebellum or structures within the brainstem, such as the inferior olive and cerebellar peduncles, instead of the basal ganglia.[4] Moreover, defects in transmission from the inferior olive to Purkinje cells are central to dystonic pathophysiology in the genetic rat model.[4]

The midbrain has additionally been implicated in dystonia, with animal studies demonstrating its role as a head neural integrator.[5] Muscimol injections into the interstitial nucleus of Cajal of subhuman primates resulted in fixed, dystonic posturing of the neck. Cerebellar and basal ganglia inputs to this region of the midbrain are also well known.[5]

Our case lends credence to the theory that cerebellar and brainstem structures are involved in the dystonia pathophysiology.

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We provide a clear visual representation of cervical dystonia, which, in this case, is associated with dysfunction of structures lying within the cerebellopontine angle.

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Author Roles

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Supporting Information

Supporting information may be found in the online version of this article.

Supplemental Video 1. Segment 1: Brain MRI T1—axial plane—showing a right-sided vestibular schwannoma. Segment 2: Postsurgical brain MRI—axial, coronal, and sagittal planes—showing a right middle cerebellar peduncle lesion, extended to the cerebellopontine angle and marginally to the right pontine tegmentum with a subsequent mass effect on cerebellum. Segment 3: Right-sided torticollis, worse with action, and a clear *geste antagoniste*, consistent with cervical dystonia. Segment 4: Cerebellar findings, including dysmetria and dysdiadochokinesia, worse on the right, with prominent gait ataxia.