Cerebellar mutism associated with a midbrain cavernous malformation

Case report and review of the literature

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The authors report a case of cerebellar mutism arising from a hemorrhagic midbrain cavernous malformation in a 14-year-old boy. No cerebellar lesion was identified; however, edema of the dorsal midbrain was noted on postoperative magnetic resonance images. Dysarthric speech spontaneously returned and then completely resolved to normal speech. This case provides further evidence for the theory that involvement of the dentatothalamic tracts, and not a cerebellar lesion per se, is the underlying cause of “cerebellar” mutism.

KEY WORDS • mutism • dysarthria • oropharyngeal apraxia • midbrain • cavernous malformation • vascular malformation

Mutism, or the inability to speak, has commonly been described after lesions in the Broca area, the supplementary motor area, and the mesencephalic reticular formation (akinet mutism). Holmes described speech disturbances following traumatic injury of the cerebellum, but did not specifically mention mutism. True mutism of a presumed cerebellar origin was first described in detail in 1985. Cerebellar mutism typically occurs after posterior fossa operations in the pediatric population, especially after resection of a posterior fossa tumor. Adult cases have also been reported. The clinical picture involves mutism that occurs hours to days after the operation. The mutism spontaneously gives way to dysarthric speech, which then completely resolves over time.

The anatomical and pathophysiological basis of cerebellar mutism has remained elusive despite numerous case reports. Several authors have analyzed the reported anatomical location of lesions giving rise to cerebellar mutism. Such topographic analysis has led most to suspect the deep cerebellar nuclei and, more specifically, the dentate nucleus and its output. Others have suggested involvement of the cerebellar hemispheres, particularly the right one and functional imaging studies have even been used to link cerebellar mutism with altered function in the cerebral cortex.

We present a case of mutism following surgery in a patient with a midbrain cavernous malformation. Analysis of this case further elucidates the anatomical basis for cerebellar mutism.

Case Report

History. This 14-year-old, right-handed, previously healthy boy presented with a severe headache. Several hours later, right arm and face numbness, a right hemiparesis, and a left-sided ptosis were noted. He had headaches since childhood, approximately once every 2 weeks, but they were never preceded by an aura or accompanied by a neurological deficit. The patient was awake, conversant, and normotensive. He had a left-sided oculomotor palsy and right-sided trigeminal and facial nerve palsies. Glossopharyngeal and hypoglossal nerve function was normal. He had a left-sided oculomotor palsy and right-sided trigeminal and facial nerve palsies. Glossopharyngeal and hypoglossal nerve function was normal. He had a right-sided pronator drift and right lower-extremity weakness. Deep tendon reflexes were hyperactive, with increased tone and a Babinski response on the right side. Hypalgesia was present in the right upper extremity and face, and he exhibited dysmetria in the right upper extremity. The patient’s family thought that his speech was not quite normal, and a speech therapist described slow, slurred speech with a monotone vocal quality and dysarthria; the patient thought his speech was “choppy.” He was able to follow complex commands accurately and responded to questions and conversation appropriately; however, he could speak only in short sentences. He had a flat affect but responded appropriately to occasional jokes. He was able to imitate all oral movements with only a mild impediment in initiation and speed.

Abbreviations used in this paper: MR = magnetic resonance; SPECT = single-photon emission computerized tomography.
Examination and Operation. An MR image revealed a lesion consistent with a brainstem cavernous malformation and an associated hematoma (Fig. 1 upper left). A posterior subtemporal craniotomy was performed 7 days after presentation. An image-guided system was used to plan the surgical trajectory. A $3 \times 4$-cm craniotomy was centered just superior to the lateral third of the left transverse sinus. The posterior temporal lobe was elevated and the tentorium was incised to increase exposure. A 3- to 4-mm incision was made in the dorsal midbrain, just rostral and lateral to the exit zone of the trochlear nerve. After traversing approximately 2 mm of brainstem, a clot was encountered. After evacuation of the clot, the malformation came into view and was resected. Histopathological analysis revealed a classic cavernous malformation and associated hemorrhage.

Fig. 1. Upper Left: Axial $T_1$-weighted MR image revealing a cavernous malformation (arrow) and associated midbrain hematoma. Upper Right: Axial $T_2$-weighted MR image obtained 1 day postoperatively, demonstrating the air–fluid level in the operative bed and some surrounding $T_2$ hyperintensity thought to represent edema. Lower Left: Axial $T_2$-weighted MR image obtained 14 days postoperatively, demonstrating increasing $T_2$ hyperintensity surrounding the operative bed and crossing the midline. Lower Right: Axial $T_2$-weighted MR image obtained 4 months postoperatively, demonstrating maturation of the operative bed and resolution of the edema.
Postoperative Course. Postoperatively, the patient was awake and followed commands. He suffered two short generalized seizures and a regimen of phenytoin was initiated. Results of his motor examination were unchanged and his speech was dysarthric but also unchanged. A postoperative MR image was obtained (Fig. 1 upper right). On the 2nd postoperative day, the patient had difficulty swallowing thin liquids and was placed on a diet of soft foods. On the 4th postoperative day, he became mute and his right hemiparesis worsened slightly. On the 6th postoperative day, he was able to make grunting noises. He was frustrated, but nonetheless made attempts to speak. An MR study obtained on postoperative Day 14 demonstrated increased edema relative to the immediate postoperative image (Fig. 1 lower right). Dysarthric speech was recovered on postoperative Day 16, and he was speaking in single words and short phrases in a low-volume monotone by postoperative Day 19. His vocal volume slowly improved, and he became able to imitate more complex oral movements.

Four months postsurgery, the patient still had dysarthric speech but was walking without a cane or brace. A repeated MR image obtained at that time showed maturation of the operative bed (Fig. 1 lower right). By 7 months, his dysarthria had resolved, his speech and affect had returned to normal, and his paresis and hemiparesis had almost completely resolved.

Discussion

Various theories have been proposed to explain cerebellar mutism. Rekate, et al.,27 suggested that bilateral involvement of the dentate nucleus was required. In a study by Fraioli and Guidetti,22 two of 50 patients, in whom stereotactically guided lesions had been placed bilaterally in the dentate nucleus for dyskinesias, exhibited temporary mutism that resolved after 1 to 3 months. Ammirati, et al.,7 noted resolution of bilateral hypodense areas in the dentate nucleus that coincided with improvement in their 14-year-old patient’s mutism. Lesions of the dentate nucleus and its associated tracts were cited by Dietze and Mickle7 and by Crutchfield, et al.,8 as a cause of cerebellar mutism. Other authors have implicated the paravermian structures that include the dentate nuclei.18 Interestingly, positron emission tomography studies and SPECT scanning have demonstrated bilateral activation of the paravermian areas during speech.26

Frim and Ogilvy,13 in their report of a case of mutism following resection of a pontine cavernous malformation, hypothesized involvement of the dentatothalamic tracts; however, the pontine lesion did not clearly involve these tracts.26 Nishikawa, et al.,27 reported cerebellar mutism occurring after a basilar artery occlusion; however, both cerebellar hemispheres were damaged, making it difficult to localize the anatomical substrate specifically.

Miyakita, et al.,22 reported on a 3-year-old girl who presented with a brainstem infarction in the left tegmentum of the midbrain, whose mutism was detected 1 week later, after extubation. Although the time course was not well documented and oropharyngeal apraxia was not noted, the similarity to our case is clear.

Some authors have specifically implicated the left cerebellar hemisphere.2 Lechtenberg and Gilman19 retrospectively reviewed 162 cases of cerebellar mutism associated with nondegenerative cerebellar disease and found that, in the majority of cases, the lesions were located in the cerebellar hemisphere, more on the left side than the right. These authors also noted that damage to the medial hemispheres was present in all patients who underwent vermis resections and later became mute. Some authors have reported cerebellar hemorrhages in the lateral hemispheres, more commonly on the left side, which resulted in speech difficulties.7 Others have attributed dysarthria to damage in the superior vermis.21 Also, SPECT scans have been interpreted to demonstrate bilateral edema in the cerebellar peduncle and in the left cerebellar hemisphere in patients with cerebellar mutism.16 Germano, et al.,14 reported bilateral frontotemporoparietal areas of reduced perfusion on SPECT scans that were associated with cerebellar mutism; however, their patients also had marked hydrocephalus. They suggested diaschisis as a possible origin, with changes in the posterior fossa causing perfusion changes in the cerebral cortex, but it is difficult to say whether hydrocephalus in and of itself could cause those changes. Other authors have suggested functional disturbances, hydrocephalus, or meningitis as possible causes.11

Vasospasm or edema is thought to account for the delay in onset of mutism.8 In our case we believe that edema was the causative factor, as evidenced by the postoperative MR image obtained after the onset of the patient’s mutism.

The presence of dysarthria on presentation is unusual in our case. Edema and hemorrhage in the left midbrain tegmentum was already present at that time. This may indicate some hemispheric dominance in relation to speech. In addition, our patient had oropharyngeal apraxia. This has previously been discussed by Bhatoe,4 who linked posterior fossa mutism with oropharyngeal apraxia, and by van Mourik, et al.,27 who also cited bilateral edema of the de- cussation of the superior cerebellar peduncles as a factor possibly involved in this clinical picture.

In a recent analysis of cerebellar mutism, Doxey, et al.,8 described the risk factors as including tumors, especially medulloblastomas, with brainstem invasion. Vermis invasion did not correlate well with the occurrence of mutism. These conclusions also support our findings.

One possible confounding factor in our patient is the surgical approach, in which the posterior left temporal lobe was elevated to obtain exposure. This could potentially have caused direct cortical injury to speech areas, especially given that the patient suffered postoperative seizures. Arguing against this is the fact that the patient exhibited a clinical picture consistent with cerebellar mutism and not aphasia. Additionally, postoperative T2-weighted imaging revealed abnormalities in the posterior temporal lobe (Figs. 1 upper right and lower left).

Conclusions

The majority of the evidence implicates involvement of the dentatothalamic tract in cerebellar mutism. Edema appears to be the most likely explanation for the delayed onset seen in most cases. Alterations in speech may appear in conjunction with unilateral lesions; however, bilateral involvement seems to be necessary for complete mutism. Cerebellar mutism appears to be an extreme form of dys-
arthria and oropharyngeal apraxia. We speculate that it may arise from lesions located at any site along the dentatothalamic tracts.

References


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