Mutism after posterior fossa surgery in children

Report of three cases

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Three patients aged 5½ to 9 years old with mutism after posterior fossa surgery are presented. The entity is discussed with a review of 15 additional previously reported cases in children aged 2 to 11 years. In all 18 patients, a large midline tumor of the posterior fossa (medulloblastoma in nine cases, astrocytoma in five, and ependymoma in four), often attached to one or both lateral recesses of the fourth ventricle, was removed. Mutism developed 18 to 72 hours after the operation (mean 41.5 hours) in patients with no disturbance of consciousness and no deficits of the lower cranial nerves or of the organs of phonation. All of these children had spoken in the first hours after surgery. The disorder lasted from 3 to 16 weeks (mean 7.9 weeks). Speech was regained after a period of dysarthria in six of the 10 cases for whom this information was available. The various hypotheses advanced to explain the pathogenesis of this speech disorder are analyzed.

KEY WORDS: mutism, posterior fossa tumor, speech disorder, children

Speech disturbances occur frequently after surgical removal of posterior cranial fossa tumors, especially when the surgery involves a generous resection of a cerebellar lobe (particularly the left) and part or all of the vermis. The main problems are with articulation of syllables, scanning speech, frequent mispronunciation, prolongation of sounds, distortion of vowels, slow rhythm, and absence of stress, all of which are generally covered by the term "dysarthria."

An unusual finding after the same type of operation is mutism: that is, the complete absence of speech. We describe three personal cases and review 15 previously reported cases. We then discuss the possible pathogenesis of this form of mutism, which may be more frequent in neurosurgical practice than is recorded.

Case Reports

Case 1

This 9-year-old right-handed girl had a 1-year history of occipital headache of increasing severity, accompanied the week before admission by vomiting, diplopia, and clouding of vision. Her parents reported personality and mood changes during the 6 months before admission and a decline in school performance.

Examination. Neurological examination on admission showed slight truncal ataxia and bilateral papilledema. A computerized tomography (CT) brain scan revealed a large midline posterior fossa tumor of patchy and ill-defined limits, with heterogeneous enhancement and triventricular hydrocephalus (Fig. 1).

Operation. The tumor was approached through a midline suboccipital craniectomy using microsurgical techniques. When the dura was opened the cerebellar tonsils were found to have herniated to the level of C-1. Retraction of the tonsils and incision of the inferior vermis exposed a grayish tissue of rubbery consistency with areas of degeneration, attached to the right lateral recess of the fourth ventricle. The lesion, which was totally removed, proved on histological examination to be a pilocytic astrocytoma.

Postoperative Course. The child presented no new neurological deficits during the first 48 hours after the operation and talked to her parents and to the medical staff. On the 3rd day, while remaining alert, she refused food and ceased to utter articulate comprehensible sounds, although there were no deficits of the lower cranial nerves. On Day 4 an aseptic meningitis developed which lasted for 15 days. About a month after the operation, during which period she remained fully con-
FIG. 1. Case 1. Computerized tomography scan with contrast enhancement demonstrating a large midline tumor of inhomogeneous density and marked triventricular hydrocephalus.

conscious, the patient gradually began to eat and to utter a few comprehensible words. On discharge and return to the family environment she rapidly regained verbal expression, although at first it was severely dysarthric. At her 1-month follow-up examination, she still had mild cerebellar disturbances although her expressive language was almost normal. Five years later, she was neurologically intact.

Case 2

This 5½-year-old right-handed girl had a 7-month history of intermittent frontal headaches of increasing severity, accompanied for 5 months by morning vomiting. Ten days before admission she developed diplopia with convergent strabismus and an unsteady gait.

Examination. Neurological examination showed ataxic gait with leftward deviation; rapid finger-nose movements and bilateral dysdiadochokinesia (greater on the right than the left); bilateral first-degree nystagmus (greater on the right than the left); convergent strabismus (paralysis of both lateral rectus oculi muscles); and bilateral papilledema. A CT brain scan demonstrated a large enhancing midline cerebellar lesion, with irregular contours and considerable triventricular hydrocephalus (Fig. 2).

Operation. Midline suboccipital craniectomy and opening of the dura mater exposed a friable gelatinous tumor which herniated through the cerebellar tonsils. The inferior vermis was incised and the tumor completely removed by means of a Cavitron ultrasonic surgical aspirator (CUSA), with microsurgical technique. The tumor, which histologically was a pilocytic astrocytoma, was attached to the right lateral recess of the fourth ventricle.

Postoperative Course. During the first 48 hours the child was alert and cooperative, carried out complex commands, and had simple conversations with her parents and doctors. Her neurological disturbances, mostly of cerebellar origin, were unchanged from the preoperative status. She had no lower cranial nerve deficits and had begun to take soft food without dysphagia. On Day 3 after surgery she ceased to speak or eat. On Day 4 she developed an aseptic meningitis, which lasted for 12 days. The child remained normally conscious but mute during the next 25 days, when she began to answer questions with gestures, to watch television with apparent interest, and to eat soft food again. Within a week she began feeding herself without help. About 2 months after the operation she began to utter a few words, with severe dysarthria (scanned speech), and wrote her name, although with marked dysgraphia.

Back in her home environment the child gradually regained verbal expression and the ability to write. Within 6 months she was speaking almost normally. At her 2-year follow-up examination her speech was fluent and appropriate for her age and there were no cerebellar disturbances.

Case 3

This 6-year-old right-handed boy had a 1-year history of frequent bouts of headache and occasional vomiting. His mother reported that for about 1 month prior to admission his walking had deteriorated with frequent reeling, and that his movements were clumsy. His writing had also deteriorated.

Examination. Neurological examination revealed static and dynamic ataxia, bilateral dysmetria in the finger-nose test, dysgraphia, and bilateral papilledema. A CT brain scan showed a large midline lesion of patchy density with indistinct borders, which enhanced considerably with administration of contrast material. There was also significant triventricular hydrocephalus (Fig. 3).
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**Fig. 3.** Case 3. Computerized tomography scan with contrast enhancement demonstrating a large midline cerebellar tumor and triventricular hydrocephalus.

**Operation.** Midline suboccipital craniectomy and incision of the vermis cerebelli exposed a large friable grayish red mass in the fourth ventricle which was attached to the right lateral recess of the ventricle. The tumor was totally removed with microsurgical technique, using the CUSA. Histological examination showed the tumor to be a pilocytic astrocytoma.

**Postoperative Course.** During the first 36 hours after surgery the boy was alert and cooperative. He spoke to his parents and doctors, answering simple questions and asking for anything he needed. He had the same neurological deficits as preoperatively. He had no deficits of the lower cranial nerves and had begun to eat without dysphagia. On Day 2 the child suddenly ceased to speak to his parents and to staff and to eat, although his consciousness was in no way impaired. On Day 5 he developed aseptic meningitis which lasted for 10 days. A month after the operation, during which time the child remained alert, he answered with comprehensible gestures the questions put to him and began to feed himself.

On returning to his home environment the child gradually recovered his powers of verbal expression. After 2 months he was able to make himself understood, although his speech was severely dysarthric. The dysarthria and the other cerebellar disturbances slowly improved and at his 6-month follow-up examination he had no speech disorder and only mild ataxia and slight dysmetria.

**Discussion**

The term "mutism" is usually defined as the complete absence of speech in a conscious subject, with no organic lesions of the neuraxis (functional forms) or, more rarely, with such lesions (organic forms). The functional forms are for the most part the purview of the psychiatrist (psychosis, autism) while the organic forms concern the neurologist and neurosurgeon. These latter forms of mutism are, according to the zone of the nervous system affected, subdivided into six types: 1) lesions of Broca's area (motor aphasia); 2) lesions of the supplementary motor area of the dominant hemisphere; 3) lesions of the mesencephalic reticular formation (akineti...
### TABLE 1
Summary of 18 reported cases of mutism after posterior fossa tumor surgery

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Tumor Location</th>
<th>Size of Tumor</th>
<th>Histology</th>
<th>IV Vent</th>
<th>Postop Anorexia</th>
<th>Postop Hydrocephalus</th>
<th>Postop Meningitis</th>
<th>Interval (hrs)</th>
<th>Duration of Mutism</th>
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<tbody>
<tr>
<td>Rekate, et al., 1985</td>
<td>8, F</td>
<td>vermis</td>
<td>--</td>
<td>medulloblastoma</td>
<td></td>
<td>yes</td>
<td>no</td>
<td>no</td>
<td>48</td>
<td>6 days</td>
</tr>
<tr>
<td></td>
<td>6, M</td>
<td>vermis, rt hemisphere</td>
<td>large</td>
<td>cystic astrocytoma</td>
<td></td>
<td></td>
<td></td>
<td>yes</td>
<td>72</td>
<td>4 days</td>
</tr>
<tr>
<td></td>
<td>2, --</td>
<td>--</td>
<td>--</td>
<td>ependymoma</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>8 wks</td>
</tr>
<tr>
<td></td>
<td>10, --</td>
<td>--</td>
<td>--</td>
<td>medulloblastoma</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>8 wks</td>
</tr>
<tr>
<td></td>
<td>9, --</td>
<td>--</td>
<td>--</td>
<td>medulloblastoma</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>12 wks</td>
</tr>
<tr>
<td></td>
<td>11, --</td>
<td>--</td>
<td>--</td>
<td>medulloblastoma</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>3 wks</td>
</tr>
<tr>
<td>Yonemasu, 1985†</td>
<td>--</td>
<td>bilat</td>
<td>large</td>
<td>ependymoma</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>18-72</td>
<td>4-12 wks</td>
</tr>
<tr>
<td></td>
<td>--</td>
<td>bilat</td>
<td>large</td>
<td>medulloblastoma</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>18-72</td>
<td>4-12 wks</td>
</tr>
<tr>
<td></td>
<td>--</td>
<td>bilat</td>
<td>large</td>
<td>medulloblastoma</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>18-72</td>
<td>4-12 wks</td>
</tr>
<tr>
<td>Humphreys, 1988‡</td>
<td>7, M</td>
<td>vermis, rt hemisphere</td>
<td>very large</td>
<td>medulloblastoma</td>
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<td>yes</td>
<td>no</td>
<td>yes</td>
<td>24</td>
<td>16 wks</td>
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<tr>
<td></td>
<td>3, M</td>
<td>IV vent</td>
<td>large</td>
<td>medulloblastoma</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>yes</td>
<td>7 wks</td>
<td></td>
</tr>
<tr>
<td></td>
<td>7, M</td>
<td>IV vent</td>
<td>large</td>
<td>medulloblastoma</td>
<td>yes</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>10 wks</td>
<td></td>
</tr>
<tr>
<td></td>
<td>4½, M</td>
<td>IV vent, vermis</td>
<td>large + cyst</td>
<td>fibrillar astrocytoma</td>
<td>no</td>
<td>no</td>
<td>preop</td>
<td>no</td>
<td>72</td>
<td>7 wks</td>
</tr>
<tr>
<td>Ferrante, et al., 1990</td>
<td>10, F</td>
<td>IV vent, vermis</td>
<td>--</td>
<td>ependymoma</td>
<td>yes</td>
<td>—</td>
<td>yes</td>
<td>no</td>
<td>24</td>
<td>10 days</td>
</tr>
<tr>
<td></td>
<td>9, F</td>
<td>vermis, MCH, IV vent</td>
<td>large</td>
<td>pilocytic astrocytoma</td>
<td>yes</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>48</td>
<td>4 days</td>
</tr>
<tr>
<td></td>
<td>5½, F</td>
<td>vermis, MCH, IV vent</td>
<td>very large</td>
<td>pilocytic astrocytoma</td>
<td>yes</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>48</td>
<td>8 days</td>
</tr>
<tr>
<td></td>
<td>6, M</td>
<td>IV vent, vermis, rt hemisphere</td>
<td>large</td>
<td>pilocytic astrocytoma</td>
<td>yes</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>36</td>
<td>8 days</td>
</tr>
</tbody>
</table>

* IV vent = fourth ventricle; MCH = medial cerebellar hemispheres; — = data not available.
† Data presented at the 13th annual meeting of the Japanese Society for Pediatric Neurosurgery in Tsukuba, Japan, in 1985.
‡ Data presented at the 16th annual meeting of the International Society for Pediatric Neurosurgery in Rome, Italy, in 1988.

Owing to the part of the child who feels betrayed by his parents and doctors, has some factual support. First, there is recovery of verbal expression as soon as the child goes home, as noted by Humphreys¹ and in our three cases. Other points in favor of this hypothesis are the absence of cranial nerve deficits and of the apparatus of phonation, the integrity of the supratentorial speech centers, and the ability to understand spoken language and to communicate with gestures. However, the functional interpretation does not explain why mutism occurs in children operated on only for posterior fossa tumors and is at variance with the fact that the intermediate phase of speech recovery is usually marked by dysarthria, which implies an organic basis in the cerebellum. It is highly probable that the emotional stress and the prolonged hospital stay⁸ may retard the recovery of the faculty of speech and perhaps explain the refusal of food noted in six of eight cases.

The possible organic factors underlying the loss of speech in children operated on for posterior fossa tumors include: 1) extensive damage to the cerebellar parenchyma;⁸,¹⁰ 2) vascular disturbances either of ischemic origin (such as arterial spasm) or due to edema; 3) disturbances of the CSF circulation (hydrocephalus); and 4) postoperative meningitis.

Extensive destruction of the median and paramedian cerebellar substance, often bilateral, involving the vermis and hemispheric cortex, cerebellar peduncles, fibers, and part of the deep nuclei is probably the most important anatomical substrate of mutism.⁵,¹⁰ A temporary but complete loss of speech has been observed after bilateral stereotactic lesioning of the dentate nucleus performed in patients with dyskinesias,⁵ and a similar disturbance has been reported after accidental stereotactic damage to the nucleus interpositus.¹² In this “cerebellar” organic interpretation of mutism, the disturbance is seen as the extreme form of dysarthria (that is, anarthria), which is the state of the conscious patient who understands what is said to him but is quite unable to articulate (“phonetic disintegration”).³ The loss of verbal expression in children who have undergone posterior fossa surgery might thus be traced to a language apraxia, an inability to coordinate the movements of the organs of phonation (vocal cords, tongue, facial and respiratory muscles). The cerebellar damage, although most probably the major factor, does not seem to be the only cause of this form of mutism. The almost constant finding of an 18- to 72-hour interval in which the patient has normal verbal expression suggests that other organic factors may trigger the cerebellar disturbance.

Segarra¹¹ described a “syndrome of the mesencephalic artery” consisting of a clinical pattern of akinetic mutism that arises after occlusion of this vessel. It is thus possible that spasm of one or more vessels supplying the nuclei or other cerebellar structures and/or

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vasogenic edema of the same parenchymal areas may contribute to the onset of “cerebellar” mutism.

In the 18 reported cases of mutism after posterior fossa surgery, a postoperative meningeal reaction and disturbance of the CSF circulation have often been described. These two factors might have a precipitating action. The organic factors described combined with the far from negligible psychic factor might account for loss of speech lasting as long as 3 weeks to 4 months. The dysarthria that marks the process of speech recovery suggests that the cerebellar damage is undoubtedly the most important of the possible organic factors underlying mutism.

Acknowledgment

We thank Professor R. P. Humphreys for encouragement and for providing reference material.

References