The structures and pathways that control vertical eye movements are complex. The rare syndrome of downward gaze palsy has been described by several authors\(^5\)\(^,\)\(^7\)\(^,\)\(^9\)\(^,\)\(^10\)\(^,\)\(^12\)\(^,\)\(^15\)\(^,\)\(^19\)\(^,\)\(^24\)\(^,\)\(^25\) mainly in the context of midbrain infarction or hemorrhage, degenerative diseases such as progressive supranuclear palsy, and metabolic disorders such as Niemann-Pick disease. Downgaze paralysis caused by an intrinsic brainstem/midbrain tumor has also been infrequently reported.\(^4\)\(^,\)\(^11\)\(^,\)\(^14\)\(^,\)\(^16\)\(^,\)\(^19\) Lesion and stimulation studies in monkeys,\(^3\)\(^,\)\(^13\) as well as from autopsy findings in humans,\(^4\)\(^,\)\(^5\)\(^,\)\(^9\)\(^,\)\(^21\)\(^,\)\(^23\) suggest the localization of a bilateral lesion of the periaqueductal gray matter (riMLF and INC)—a butterfly-shaped area in the mesodiencephalic junction, dorsomedial to the red nucleus and adjacent to the caudal part of the third ventricle and the aqueduct.\(^1\)\(^,\)\(^8\)

To the best of our knowledge, there have been no previous studies describing transient selective downgaze paralysis as a complication after posterior fossa operations in children. The authors found downgaze palsy to be a transient complication after resection of large pediatric posterior fossa midline tumors reaching the aqueduct of Sylvius. They reviewed the cases of 2 children with large posterior fossa midline tumors who underwent resection via an inferior transventricular approach. They developed a hypothetical scheme to account for downward gaze paralysis based on anatomy and insight gained from experimental studies.

The authors describe potential risk factors for developing transient selective downward gaze paralysis with the hope of making more pediatric neurosurgeons aware of this complication following removal of lesions around the mesencephalic periaqueductal gray matter. Recognition and understanding of downward gaze palsy after posterior fossa surgery should improve preoperative counseling and promote postoperative family coping.

(DOI: 10.3171/2009.2.PEDS08446)

**Key Words** • down gaze palsy • downward gaze paralysis • pediatric brain tumor • posterior fossa tumor

**Case Reports**

**Case 1**

**History and Examination.** This previously healthy 3-year-old girl presented with a 2-week history of frequent falling, occasional headache, and intermittent vomiting. Neurological examination revealed mild truncal ataxia and otherwise unremarkable findings. There was no restriction in ocular movements. Workup with MR imaging showed a very large heterogeneously enhancing posterior fossa tumor centered in the fourth ventricle, bulging into the aqueduct of Sylvius and displacing the tectal plate superiorly (Fig. 1 upper).
Operation. We monitored facial nerve and lower cranial nerve electromyography data, brainstem auditory evoked potentials, motor evoked potentials, and somatosensory evoked potentials from the upper and lower extremities. A combined occipital/suboccipital craniotomy and C-1 laminectomy was performed. Through an inferior transventricular/transvermian approach, the tumor was visible between the tonsils of the cerebellum. The plane between the tumor and normal cerebellar tissue was distinct and further developed. The floor of the fourth ventricle was identified and carefully protected early in our dissection. The rostral-most portion of the tumor against the tectal plate was removed completely until the abnormally enlarged aqueduct was patent. However, a thin carpet of tumor subcentimeter in dimension was left on the floor of the fourth ventricle near the superior cerebellar peduncle. There was no electromyography activity from the facial nerve or lower cranial nerves; furthermore, brainstem auditory evoked potentials, motor evoked potentials, and somatosensory evoked potentials remained at their baseline levels throughout the procedure.

Postoperative Course. The patient was awakened from general anesthesia and extubated after the surgery. Although she was awake and following commands, she exhibited both spontaneous downward saccades and visual pursuit impairment. The downward component of the oculocephalic reflex was also impaired. Postoperative MR imaging showed no evidence of residual tumor. There was bilateral T2 signal abnormality in the periaqueductal region with a unilateral focus of restricted diffusion (Fig. 1 lower). By the 3rd postoperative day, the downgaze paralysis had resolved completely. The pathological diagnosis was low-grade ependymoma. At 5 months after surgery, the patient has finished a course of radiation therapy and there has been no MR imaging evidence of recurrent tumor. She has no neurological deficits and continues to do well clinically.

Case 2

History and Examination. This previously healthy 7-month-old girl presented with projectile vomiting and a 1-week history of progressive lethargy. Admission CT scanning demonstrated a hyperdense posterior fossa tumor, intraventricular metastatic lesions, and marked hydrocephalus with transependymal edema. An external ventricular drain was placed on the day of admission and the patient’s lethargy rapidly resolved. Examination showed global developmental delay, but there were no obvious focal neurological deficits. The spontaneous ocular movements were full. Further MR imaging workup of the brain revealed a large heterogeneously enhancing solid tumor obliterating the fourth ventricle with 2 metastatic intraventricular nodules (Fig. 2 upper). Spinal MR imaging showed distant leptomeningeal metastasis.

Operation. A combined occipital/suboccipital craniotomy for an inferior transventricular approach was performed. Intraoperative neurophysiological monitoring was discontinued shortly after starting the procedure due to technical difficulties. Because of the likelihood of a highly malignant tumor with distant metastatic disease, only a very judicious attempt at resection was undertaken. The primary goals of surgery were to establish tissue diagnosis and to decompress the brainstem. The floor of the fourth ventricle was identified and protected early in

![Fig. 1. Case 1. Upper: Preoperative sagittal contrast-enhanced T1-weighted MR image demonstrating an enhancing tumor extending from the aqueduct through the fourth ventricle. Lower: Postoperative axial T2-weighted MR image revealing abnormal signal in the bilateral periaqueductal area (arrow).](image)
the dissection. The tumor was removed in a piecemeal fashion. The tumor above the level of the enlarged aqueduct was difficult to reach from the transventricular approach below. Through the supracerebellar/infratentorial window, the tumor was seen encasing and densely adherent to the great veins. At this point the operation was terminated because it was not possible to resect more tumor without increasing the risk of surgical morbidity.

Postoperative Course. The patient was awakened from general anesthesia and extubated. She had no spontaneous downward gaze. The downward component of the oculocephalic reflex was also impaired. This limitation of downward gaze spontaneously resolved after 1 day. Postoperative MR imaging demonstrated a subtotal resection of the posterior fossa tumor with T2 signal changes in the periaqueductal region (Fig. 2 lower). The pathological diagnosis was disseminated atypical teratoid/rhabdoid tumor. At 2 months after surgery, the patient underwent ventriculoperitoneal shunt placement for persistent hydrocephalus and was then undergoing experimental chemotherapy.

Discussion

Anatomy

The midbrain includes centers and pathways that mediate supranuclear vertical gaze generation, neural integration in the vertical plane, and vergence movements (Fig. 3). In this region, there are 3 key structures that control vertical gaze: a group of medium-sized cells located medial and dorsal to the riMLF; the posterior commissure; and the INC.16,19 Slightly different lesions in these 3 structures are responsible for central up- versus downgaze paralysis.19,22 A downward saccadic deficit is the result of a bilateral lesion of the riMLF, which is considered to be the main premotor nucleus for the generation of vertical saccades.2,3 It is located dorsomedial to the INC and lateral to the nucleus of Darkschewitsch. There is a separation between the burst neurons in the riMLF for pathways of up- and downgaze movements.18,22 The dissociation of down- and upgaze palsy may be due to lesions of the efferent fibers of the riMLF neurons, which leave the riMLF at different sites to synapse with oculomotor nuclei.2,18,19 That is, upgaze prenuclear neurons send bilateral projections to the elevator muscles via the posterior commissure, with further crossing over within the oculomotor nuclear complex, while axons that mediate downgaze project ipsilaterally to the depressor muscles without traversing the posterior commissure.1,18 Thus, disturbance of the rostral floor of the fourth ventricle may differentially affect the downgaze fiber more so than the upgaze fiber. The INC provides a gaze-holding signal and projects to oculomotor nuclei via the posterior commissure. This hypothesis would be in accordance with localization of lesions found in humans and monkeys with down- and upgaze palsy.4,13,17,19,21

It is necessary to distinguish the difference between downbeat nystagmus and downgaze paralysis. The former is very familiar to neurosurgeons and is often present in cerebellar or cervicomedullary junction disease, whereas the latter is less frequently encountered and is associated with midbrain structures as discussed above. The vertical component of the vestibuloocular reflex modulates eye movement in relation to gravity. It has been hypothesized that the upward vestibular system is physiologically “stronger” than the downward vestibular system to counteract gravitational pull. Accordingly, the hyperactive upward vestibular system would require constant modulation/inhibition, which is supplied by the caudal medulla and cerebellar flocculus and converges on the oculomotor nuclei via the ventral tegmental tract. Compression at the cervi-
comedullary junction or cerebellar flocculus would interrupt this inhibitory pathway and lead to an upward slow drift and downbeat nystagmus.\textsuperscript{20}

**Clinical Features and Potential Risk Factors**

The most frequent postoperative findings after posterior fossa operations in children include cerebellar mutism, ataxia, cranial nerve VI and VII palsies, and hemiparesis.\textsuperscript{6} This report represents the first description of transient selective downward gaze paralysis in 2 children following surgery to resect large posterior fossa tumors. In these 2 children, the tumors were located in the midline with brainstem, fourth ventricular, and periaqueductal involvement, and they were associated with hydrocephalus and dilation of the aqueduct. There were significant compression and distortion of the mesencephalic periaqueductal gray matter. Chronic hydrocephalus and dilation of the aqueduct of Sylvius probably make the periaqueductal gray matter even more susceptible to irritation and injury through additional mechanical distortion and stretch.

The limitations of surgical approaches may increase the likelihood of this uncommon complication. The aqueduct defines the superior extent of the operative field when using inferior transventricular, transvermian, and telovelar approaches; the periaqueductal region usually represents the longest reach for the surgeon and harbors the last remnants of tumor. In addition, an aggressive attempt at gross-total resection predisposes the periaqueductal gray matter to overzealous manipulation and trauma.

Susceptibility of fibers to injury may also be related to the degree of myelination. Myelination begins early in the third trimester with completion of myelination after the first 2 years of life. It would seem to follow that those children 2 years of age or less would be more susceptible to injury of unmyelinated efferent fibers of the riMLF; however, 1 of our patients was a 3-year-old child. Further observation would be needed to determine if there is an age-related difference in susceptibility.

Both patients had downgaze palsy immediately after surgery, which resolved within 24–48 hours. There was no specific, directed intervention except the passage of time. Spontaneous resolution of this neurological deficit occurred by the 2nd postoperative day.
Conclusions

Transient selective downward gaze paralysis is a rare, probably underreported complication of posterior fossa surgery in children. The exact cause of and anatomical basis for this complication remain unclear; however, strong experimental evidence in lesion and stimulation studies in humans and monkeys seems to confirm a periaqueductal gray matter lesion. Possible risk factors would include a large midline posterior fossa tumor, the presence of associated hydrocephalus and dilation of the aqueduct of Sylvius, an inferior transventricular, transvermian, or telovelar surgical approach, and aggressive resection. Awareness and recognition of this complication lead to more useful preoperative counseling of the family and reassurance that the downgaze palsy is self-limited and will resolve within a few days of surgery.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


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