Case Report

Posterior Fossa Syndrome After Surgical Removal of a Pineal Gland Tumor

Dana L. Ellis BSca,*, Julie Kanter MDb, John W. Walsh MDc, Stacy S. Drury MD, PhDd

aMedical Program, Tulane University School of Medicine, New Orleans, Louisiana
bDivision of Hematology/Oncology, Department of Pediatrics, Tulane University School of Medicine, New Orleans, Louisiana
cDivision of Child and Adolescent Psychiatry, Department of Psychiatry, Tulane University School of Medicine, New Orleans, Louisiana
dDivision of Pediatric Neurosurgery, Department of Neurosurgery, Tulane University School of Medicine, New Orleans, Louisiana

ARTICLE INFORMATION

Article history:
Received 6 July 2011
Accepted 28 September 2011

ABSTRACT

Posterior fossa syndrome, characterized by oromotor or oculomotor apraxia, emotional lability, and mutism, occurs in some children after infratentorial tumor resection, and is thought to involve injury to the dentatothalamocortical tract. Previous cases of posterior fossa syndrome involved pediatric patients with cerebellar and other posterior fossa tumors. To heighten awareness that posterior fossa syndrome can occur after resections of tumors in other neuroanatomic locations, we present a 16-year-old boy who developed this syndrome after surgical removal of a supratentorial pinealoma, and we include a discussion of his self-reported signs.

Introduction

The cerebellum has long been associated with complex neural networks involved in the coordination of movement, working memory, and linguistic processing. More recently, insights were achieved regarding the more specific roles of the cerebellum in cognition and behavior, suggesting its importance in producing coordinated motor, cognitive, and affective behaviors [1,2].

Cerebellar mutism or posterior fossa syndrome was first reported in 1979 by Hirsh after the resection of a tumor in the posterior fossa [2]. Posterior fossa syndrome is characterized by postoperative mutism, and is frequently associated with other neurobehavioral manifestations, including cognitive impairment, ataxia, emotional lability, and behavioral disturbances [3]. The syndrome usually manifests 1-2 days after surgery, and can persist for several months [4].

The majority of previous reports on posterior fossa syndrome involved pediatric patients with cerebellar and other posterior fossa tumors. Over 30% of patients who develop this syndrome have undergone the resection of midline medulloblastomas, using an infratentorial approach [5]. To highlight the intricate nature of the dentatothalamocortical tract and heighten the awareness that posterior fossa syndrome can occur after resections of tumors in other neuroanatomic locations, we a 16-year-old boy who developed this syndrome after surgical removal of a supratentorial pinealoma via a supracerebellar and infratentorial approach. We also include a discussion of his self-reported signs, because previous reports did not include self-reports in their descriptions.

Case Report

The patient is a 16-year-old boy with no significant previous medical history. He presented at the emergency room with a 2-week history of headache and a 1-week history of emesis and diplopia. An initial review of his systems, a physical examination, and laboratory data produced unremarkable results, except for an elevated blood pressure of 142/76 mm Hg. A computed tomography scan of his head was performed in the emergency room and revealed obstructive hydrocephalus secondary to an intraventricular mass, beginning in the region of the pineal gland and occupying the third and left lateral ventricles. Results of magnetic resonance imaging of his head and spine also demonstrated obstructive hydrocephalus, attributable to the mass effect of the intraventricular pineal tumor (Fig 1), along with evidence of spinal metastasis (Fig 2).

An elective craniotomy with mass biopsy and subtotal resection plus ventriculostomy was performed the next morning. The lesion was reached through a supracerebellar and infratentorial approach, and an examination of the pathology revealed that the tumor was a mature teratoma. Cerebrospinal fluid studies demonstrated elevated levels of α-fetoprotein and β-human chorionic gonadotropin, and the tumor was designated a mixed teratoma and germ-cell tumor in the region of the pineal gland, with spinal metastasis. On postoperative day 7, the patient was transferred from the Pediatric Intensive Care Unit to the floor, where he received his first round of chemotherapy with etoposide and ifosfamide.

On postoperative day 11, the pediatric psychiatry service was consulted to assess the patient’s continued flat affect, poor coping, and concerns about avoidance of his diagnosis. Upon initial evaluation, the pediatric psychiatry service found that the patient manifested significant neurocognitive deficiencies, including slowed processing speed, deficits in long-term memory, and a decreased ability to name objects. A psychiatric examination was also significant for indicating flat affect, a paucity of speech, and limited spontaneous speech or interaction. Although his tumor was not located in the posterior fossa, the constellation of signs and the use of a supracerebellar surgical approach were consistent with posterior fossa syndrome. At this...
point, we could not discern whether the patient manifested any signs of ataxia or motor coordination difficulties because he had not yet attempted to ambulate. On postoperative day 19, the patient was able to get out of bed and into a chair, and physical therapy was initiated for postsurgical deconditioning and his unsteady gait. During interviews on postoperative days 20-25, he continued to answer only in monosyllables, and exhibited minimal eye contact, poor short-term memory, and slow processing speed. These signs were consistent with a diagnosis of posterior fossa syndrome, and further represented a significant difference from his premorbid personality. The patient was discharged on postoperative day 25 with improved, but still impaired, ambulation, mild diplopia, and continued flat affect. No focal neurologic deficits were evident. The patient presented every 2 weeks thereafter for chemotherapy. During the first follow-up visit he continued to exhibit flat affect. An ophthalmologic examination revealed resolving papilledema and paresis of left cranial nerves 5 and 6. Four weeks after discharge (2 months after surgery), he presented for chemotherapy. A complete resolution of his neurocognitive slowing, memory problems, and flat affect were then observed. The results of magnetic resonance imaging and cerebrospinal fluid studies were consistent with significant improvement, involving decreased tumor size and decreased cerebrospinal fluid levels of a-fetoprotein and b-human chorionic gonadotropin.

On subsequent follow-up visits, the patient reported significant difficulty during his first month after surgery in regard to memory (e.g., forgetting the names of his nurses and favorite basketball teams). The patient also recalled a substantial level of ataxia and vertigo for weeks after his tumor resection. He explained that he experienced the feeling of spinning in his chair while seated, and a great amount of unsteadiness while attempting to ambulate for approximately 3 weeks after surgery. He also recalled a certain degree of anhedonia because he did not want to do anything, but he did not think the anhedonia was related to overly high levels of concern about his tumor or diagnosis. He acknowledged that his interpersonal interactions, mood, and affect in the month after surgery were inconsistent with his normal outgoing and interactive personality, and he could not identify why he had acted differently. He was confident that his altered behavior was not the result of anxiety related to his cancer or his chemotherapy.

Discussion

Postoperative posterior fossa syndrome in children with brain tumors is one of the better-described neurobehavioral syndromes after infratentorial injury. Mutism, oromotor or oculomotor apraxia, emotional lability, axial hypotonia, and cerebellar or pseudobulbar dysfunction comprise the principal signs involved in this syndrome, with an onset of signs 1-2 days into the postoperative period, and variable times to recovery [4,6]. Mutism and emotional lability are generally transient, but significant long-term cognitive, behavioral, and motor deficits are frequently evident [4].

Perturbation of the cerebello-cerebral network during or after surgical tumor resection is presumed to underlie many of the signs of posterior fossa syndrome [7]. When a tumor of the pineal region is small and/or biopsy is the primary reason for surgery, as in the present case, the infratentorial supracerebellar approach is often used. Although successful, this approach involves some disadvantages. The surgical field through a posterior fossa craniotomy is often restricted, and although the surgeon can depress the cerebellum to expand the surgical opening, this maneuver has the potential to result in postoperative cerebellar swelling [7,8]. Moreover, removing tumors extending into the lateral and anterior third ventricles is often difficult, as in the present case, because they may require a long, narrow course to the foramen of Monro [7]. Retraction of the cerebellum, together with sectioning of the superior vermic veins, may result in postoperative cerebellar swelling [7]. This swelling has the potential to cause transient injury or functional disturbances of the cerebellum and the proximal dentatothalamocortical pathway, leading to findings of posterior fossa syndrome [9]. Given its role as primary outflow tract for the cerebellum and the vulnerability of its more proximal...
aspects to surgical injury, the dentatothalamocortical pathway has garnered increased attention with respect to posterior fossa syndrome [9].

The dentatothalamocortical pathway originates within the dentate nuclei and projects through the ipsilateral superior cerebellar peduncle. After decussation in the midbrain tegmentum, these projections synapse within the contralateral ventrolateral nucleus of the thalamus. From the thalamus, second-order axons terminate in the primary motor cortex and in secondary and tertiary association areas within the frontal and parietal lobes [9]. Therefore, in addition to its influence on the coordination of planned motor activity, the cerebellum, via the dentatothalamocortical tract, exerts significant influence on cognition and behavior [1,2].

Our report demonstrates that individuals with supratentorial tumors, depending on the surgical approach to the resection, may be at increased risk for posterior fossa syndrome. To the best of our knowledge, only one case of posterior fossa syndrome after resection of a pineal region tumor was reported (in an adult), and the signs in that patient included severe ataxia and dysarthria, which resolved after 3.5 months [10].

Conclusion

Posterior fossa syndrome has been increasingly reported after a range of neurosurgical procedures. No surgical approach or pattern of postoperative injury has been universally accepted as a predictor of this syndrome [9]. The disrupted physiology underlying the neurobehavioral deficits remains largely unknown, and unfortunately, a deficient understanding of posterior fossa syndrome has limited the implementation of preventive measures. We have demonstrated that this entity can occur after the resection of a supratentorial tumor, and that this outcome is likely attributable to injury to the efferent pathways from the dentate gyrus. Early recognition of this syndrome could facilitate preventive and restorative patient care, prevent subsequent complications, decrease the length of hospital stays, and promote patient and family understanding of and coping with the syndrome.

Dana Muhlfelder, BSc, initiated the literature search for the initial draft of the manuscript, and helped interview the patient after his recovery from posterior fossa syndrome.

References