ELSEVIER

Visual Disturbance Associated With Postoperative Cerebellar Mutism

Stacie R. Daniels, MD*,
Leon E. Moores, MD†, and
Marc P. DiFazio, MD*

Cerebellar mutism is an uncommon complication of posterior fossa surgery. Manifestations include disturbances of articulation, prosody, and pitch, and, if severe, complete mutism. Symptoms are independent of recognizable cortical or brainstem injury, and recovery is variable, with permanent deficits frequently observed. Cerebellar dysfunction is commonly invoked as an etiology, although controversy remains concerning the mechanism. Visual impairment has been reported only once before in the setting of this disorder. We report a confirmatory case of sudden, severe visual loss in association with cerebellar mutism after resection of a midline medulloblastoma in a 7-year-old.

Daniels SR, Moores LE, DiFazio MP. Visual disturbance associated with postoperative cerebellar mutism. Pediatr Neurol 2005;32:127-130.

Introduction

Cerebellar mutism is a well-recognized complication of posterior fossa surgery, most often occurring in children after resection of midline cerebellar masses. Symptoms consist of a dense expressive aphasia, often with complete inability to phonate, associated with a variable degree of cognitive and behavioral impairment. Taken together, these manifestations have been more recently characterized as the posterior fossa syndrome.

To date, only one report has been made of visual impairment in association with this syndrome [1]. We report a confirmatory case of transient blindness in the setting of posterior fossa syndrome after resection of a medulloblastoma in a 7-year-old male in association with a constellation of behaviors reminiscent of autistic spectrum disorders.

Case Report

Our patient is a 7-year-old male who presented to his pediatrician with a chief complaint of progressive headache and gait disturbance. He was previously well, with normal growth and development, as well as social functioning for age. Initial examination demonstrated pronounced gait ataxia but was otherwise normal. Notably, ophthalmologic examination was without abnormalities. Neuroimaging revealed a large midline cerebellar mass with mild obstructive hydrocephalus (Fig 1). Gross total resection was completed via a midline vermian approach, and was uncomplicated. Pathologic examination demonstrated medulloblastoma.

In the immediate postoperative period, the patient was mute, irritable, and indifferent to his surroundings, including family members. He demonstrated profound visual inattention, and did not blink to confrontation or fix on or follow objects presented to him. Neuro-ophthalmologic examination was noteworthy for absent optokinetic nystagmus and intact pupillary reflexes. No disc edema or retinal abnormalities were observed. The patient's behavior was markedly abnormal, with alternating periods of agitation and quiet placidity, unrelated to medication administration or his surroundings. Vocalizations were limited to screams and grunts, and he did not respond to his parent's presence, touch, or reassurances. The patient underwent urgent magnetic resonance imaging with diffusion weighting, which did not reveal abnormalities beyond the residual tumor bed edema (Fig 2a, b). Specifically, there were no cortical abnormalities or brainstem injury evident. Electroencephalogram demonstrated mild diffuse background slowing, with areas of posterior focal slowing, left greater than right.

Over several weeks, the patient gradually began to briefly consider faces and other visual stimuli. Profound impairment of expressive language continued. Within 4 months of surgery, examination of the patient's visual functioning was normal. Repeat magnetic resonance imaging again demonstrated normal cortical appearance. Expressive and receptive language functioning also began to improve, and within 6 months of surgery was close to normal, although there appeared to be a continued decrease in speech rapidity, and appropriate prosody was diminished with occasional "sing-song" expressions. Behavior was approximate to his presurgical baseline, although the parents continued to report a lowered threshold for angry outbursts, and a decreased ability to tolerate frustration. However, he was able to interact with peers and family, and had no other demonstrable social impairment.

From the *Departments of Neurology and †Neurosurgery, Walter Reed Army Medical Center, Washington, DC.
Presented in poster form at the 2001 American Association of Neurological Surgeons/Congress of Neurological Surgeons section on Pediatric Neurological Surgery Annual Meeting, New York, New York

Communications should be addressed to:
Dr. DiFazio; Department of Neurology; Walter Reed Army Medical Center; Washington, DC 20307.
Received March 9, 2004; accepted July 19, 2004.

doi:10.1016/j.pediatrneurol.2004.07.014 • 0887-8994/05/$—see front matter

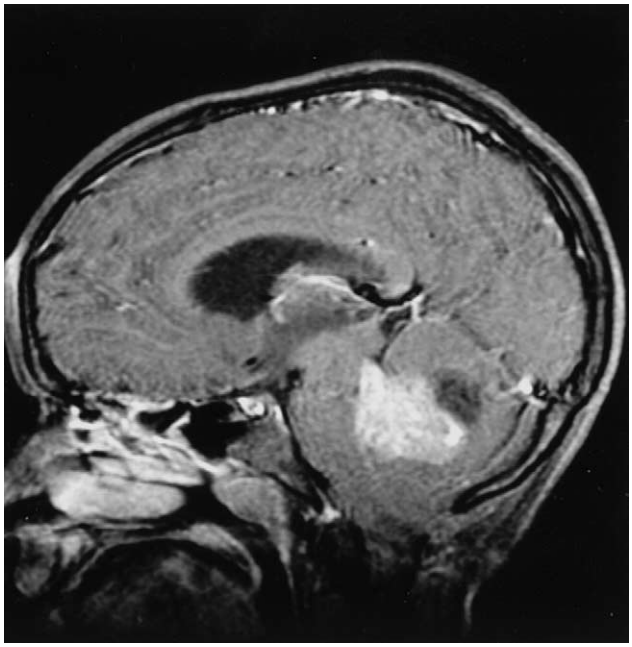

Figure 1. Sagittal T_1-weighted magnetic resonance imaging with contrast (TR 467 ms, TE 14 ms). Large enhancing mass is observed in the midline cerebellum, with resultant hydrocephalus and brainstem deformation.

Discussion

Posterior fossa syndrome is a well-known sequela of posterior fossa surgery in children, particularly in those who have undergone resection of midline vermian masses. Although the initial reported case was published relatively recently in 1985, it is recognized to occur in at least 15-20% of patients undergoing this type of procedure [2,3].

The mechanism of speech impairment is still unknown, although it had been previously characterized as "mutism" and was attributed to profound dysarthria associated with cerebellar motor outflow impairment [2,3]. More recently, manifestations have been expanded greatly beyond speech impairment, and attentional, language, and behavioral disturbances are well-recognized components of the disorder [4,5]. The widely expanded clinical manifestations, without evidence of cerebral cortical injury, provide further support for an important role for the cerebellum in cognitive functioning.

Extensive evidence, originating from work published in the early 20th century, has substantiated a modulatory effect of the cerebellum on human and animal behavior and emotion [6]. Wide-ranging direct and indirect anatomic pathways and interaction between the cerebellum and limbic, parietal, and frontal cortices were postulated and confirmed in multiple human and animal studies [6-8]. Early investigation demonstrated a modulatory effect of cerebellar stimulation on animal behavior, with changes in response to so-called "sham rage" induced by hypothalamic stimulation [6]. These studies ultimately localized these effects to the vermis and fastigial nucleus [6-8]. Human studies of cerebellar lesions demonstrate impairment in a variety of cognitive domains, including attentional, language, and behavioral disturbances [5,9]. One study, utilizing vermian stimulation in severely emotionally disturbed individuals, resulted in improvement in the majority of treated patients [10].

More recently, impairment in time-based performance has also been reported in patients with cerebellar injury. Such impairments have been attributed to injury to parallel-array "microprocessors" present within the underlying crystalline structure of the cerebellum [11,12]. This array is postulated to work in concert as a powerful learning and processing network [11,12].

Analysis of past reports of cerebellar lesions in children has led to a theorized higher degree of complexity and organization of cerebellar cortex than was previously understood. Postulated anatomic characteristics include a vermian equivalent to the limbic system, with emotional and behavioral disturbances occurring after removal of midline medulloblastomas [5,9,13]. The cerebellar hemispheres may serve to modulate higher cortical functions such as thought, executive functioning, and language. A distinction between right and left anatomic regions as observed in the topographic organization of the cerebral hemispheres may exist as well [13].

Visual impairment has recently been reported as a component of the posterior fossa syndrome in several children after removal of midline cerebellar tumors [1]. This report indicated a lack of other ocular or visual pathway dysfunction and normalization of visual functioning concurrent with reacquisition of speech functioning. Although other mechanisms were considered (ischemia, optic nerve injury caused by relief of obstructive hydrocephalus, shunt placement trauma), the proposed mechanism was a deficit in visual attention, brought on by an interruption of cerebellar centers dedicated to this function. This finding occurred in individuals who also manifested autistic behavior; that is, patients were inattentive to family members and their surroundings, in association with a severe disturbance in their ability to communicate [14]. These behavioral manifestations improved, although long-term follow-up was not reported.

Our patient demonstrated similar manifestations. As is postulated in the original report, we believe that his symptoms likely represent a previously unrecognized syndrome associated with posterior fossa/cerebellar injury, and not a new manifestation, given the extreme impairment that some patients manifest, and the attendant difficulties in examining such patients. Our report supports the postulate that this syndrome is not associated with primary visual impairment (i.e., optic nerve or tract injury), and did not occur as a result of ischemia affecting the optic tracts or cortex. Investigation with functional studies (diffusion-weighted magnetic resonance imaging and electroencephalography) allowed us to exclude an epileptic or ischemic etiology for the visual impairment, consistent with this patient's complete resolution of visual function-

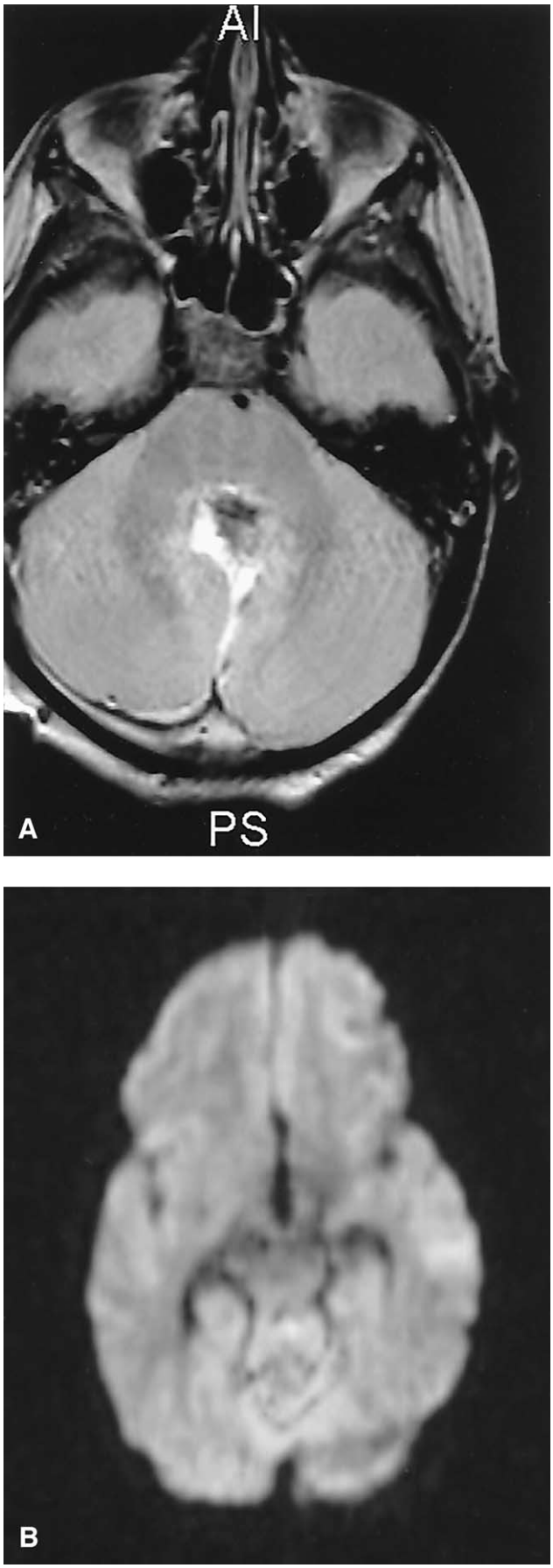

Figure 2. (A) Axial T_2-weighted magnetic resonance imaging (TR 2600 ms, TE 90 ms). Postoperative changes are observed in the midline vermian region. (B) Axial diffusion-weighted magnetic resonance imaging (TR 10,000 ms, TE 97 ms). No evidence of ischemic injury. AI = anterior inferior, PS = posterior superior.

ing months after surgery. Interestingly, a visual projection from the tectum to the cerebellum was supported in animal studies from the early part of the last century, indicating that a visual representation is likely present in the cerebellum [15]. Our patient's behavioral decompensation was also clearly related to the surgical excision itself, as its onset was immediate, and departed dramatically from presurgical functioning. In this case, the deterioration cannot be attributed to other therapies that have been postulated in the past, including radiotherapy or chemotherapy, and given that the onset was so abrupt, it is unlikely to be a primary psychiatric response to his surroundings and medical condition. In fact, our patient's current neuropsychiatric condition continues to mimic that of patients with autism, albeit of a less severely affected nature than previously. His parents report continued difficulties with transitions between situations, and frequent tantrums and a low tolerance for frustration, all attributes commonly observed within the autistic spectrum. Although it is possible that these manifestations are attributable to subsequent medical and pharmacologic therapies, they began with dramatic manifestations of social, communication, and language impairments and we believe that they represent residua from his initial injury.

Although a consistent cerebellar lesion has not been found in individuals with autism, a variety of reports indicate that a subset of such patients may have a primary cerebellar substrate for their symptoms [11,12,16]. Given that the manifestations of autism are extremely heterogeneous and the proposed etiologies vary widely, it is possible that some individuals represent an interruption of primary cerebellar centers that allow for modulation of visual attention, as well as other more complex areas of cognition, disturbing appropriate social and cognitive functioning. More detailed studies are warranted to determine the cerebellar role in cognition, vision, and attention, with special attention to those individuals affected by autism.

Conclusion

We report a case of acute visual disturbance in association with postoperative cerebellar mutism. This case confirms the findings of Liu et al. [1], and expands the clinical manifestations that may be observed in this syndrome. Additional investigation into the contribution of the cerebellum to behavioral, visual, and emotional functioning is crucial, and may aid in the further understanding of congenital disturbances of behavior, such as the autistic spectrum disorders.

The views and opinions contained herein are the private ones of the authors and are not to be construed as representing the views of the Department of Defense or Department of Army.

References

[1] **Liu** G, Phillips C, Malloy P. Visual impairment associated with mutism after posterior fossa surgery in children. Neurosurg 1998;42: 253-7.

[2] **Van Calenbergh** F, Van de Laar A, Plets C. Transient cerebellar mutism after posterior fossa surgery in children. Neurosurg 1995;37: 894-8.

[3] **Gelabert-Gonzalez** M, Fernandez-Villa. Mutism after posterior fossa surgery: Review of the literature. Clin Neurol Neurosurg 2001;103: 111-4.

[4] **Pollack** IF. Posterior fossa syndrome. Int Rev Neurobiol 1997; 41:411-32.

[5] **Vandeinse** D, Homyak J. Linguistic and cognitive deficits associated with cerebellar mutism. Ped Rehabil 1997;1:41-4.

[6] **Anand** B, Malhotra C, Singh B, Dua S. Cerebellar projections to the limbic system. J Neurophysiol 1959;22:451-8.

[7] **Heath** R, Cox A, Lustick L. Brain activity during emotional states. Am J Psychiat 1974;131:858-62.

[8] **Heath** R, Dempesy C, Fontana C, Myers W. Cerebellar stimulation: Effects on septal region, hippocampus, and amygdala of cats and rats. Biol Psychiat 1978;13:501-29.

[9] **Levisohn** L, Cronin-Golomb A, Schmahamann J. Neuropsychological consequences of cerebellar tumor resection in children: Cerebellar cognitive affective syndrome in a pediatric population. Brain 2000;123:1041-50.

[10] **Heath** RG. Modulation of emotion with a brain pacemaker. J Nerv Ment Dis 1977;165:300-17.

[11] **Pierce** K, Courchesne E. Evidence for a cerebellar role in reduced exploration and stereotyped behavior in autism. Biol Psychiatry 2001;49:655-64.

[12] **Allin** M, Matsumoto H, Santhouse M, et al. Cognitive and motor function and the size of the cerebellum in adolescents born very pre-term. Brain 2001;124:60-6.

[13] **Riva** D, Giorgi C. The cerebellum contributes to higher functions during development: Evidence from a series of children surgically treated for posterior fossa tumors. Brain 2000;123:1051-61.

[14] **Rapin** I. Introduction and overview. In: Bauman ML, Kemper TL, eds. The neurobiology of autism. Baltimore: Johns Hopkins University Press, 1994:1-17.

[15] **Snider** RS. Electro-anatomical studies on a tectocerebellar pathway. Anat Rec 1945;91:299.

[16] **Courchesne** E, Karns C, Davis H, et al. Unusual brain growth patterns in early life in patients with autistic disorder. Neurology 2001;57:245-54.