Teaching NeurolImage:

Tremor in aqueductal stenosis and response to endoscopic third ventriculostomy

Marc W. Halterman,
MD, PhD
G. Edward Vates, MD,
PhD
Garrett Riggs, MD,
PhD

VIDEO

Address correspondence and reprint requests to Dr. Marc W. Halterman, Department of Neurology, University of Rochester School of Medicine and Dentistry, 601 Elmwood Avenue, Box 673, Rochester, NY 14642
Marc_Halterman@urmc.rochester.edu

Idiopathic aqueductal stenosis (AS) may account for up to 59% of cases presenting with triventricular noncommunicating hydrocephalus.¹ The clinical presentations associated with triventricular hydrocephalus differ depending on age at onset and the acuity of obstruction. Acute syndromes include Parinaud's syndrome (vertical gaze restriction, lid retraction, and pupillary abnormalities), the rostral mid-brain syndrome (upward gaze palsy, retraction nystagmus, pyramidal and extrapyramidal signs), and deficits in arousal. In the very young, chronic diencephalic compression can produce the bobble-head doll syndrome (high frequency head movements, limb ataxia, tremor, and cognitive deficits). Resolution of transtentorial pressure gradients by CSF diversion typically produces rapid improvement.²

We describe an adult patient with upper extremity tremor due to decompensated hydrocephalus from AS, who demonstrated improvement following endoscopic third ventriculostomy.

CASE REPORT An 18-year-old right-handed African American man presented with chronic, progressive tremor of the right hand. He was the product of an uncomplicated pregnancy and satisfied normative criteria for both growth and developmental milestones including head circumference. There was no history of head injury or encephalitis. At age 9 he developed action tremor in his dominant hand, which began in the distal upper extremity and progressed over 6 months to include the proximal arm and right leg. These symptoms were aggravated by stress, and interfered with activities of daily living. While he was considered less agile than his peers, he had no functional impairment or problems with falls. Rest tremor was not appreciated. By age 13, he was diagnosed with essential tremor, and trials of primidone and propranolol were unsuccessful.

At presentation the tremor was unchanged, but his academic performance had declined and he had frequent headaches. He denied nausea, visual disturbances, weakness, or problems with bladder control. On examination, head circumference measured

62 cm and mild frontal bossing and hypomimia were present. Recall at 5 minutes was impaired (one of three objects), he could not complete serial sevens, and he was unable to provide the date. Language function was intact. Funduscopy was benign, pupils were 3 mm and reactive, and versions were preserved without nystagmus. Strength was full throughout with a spastic catch elicited on elbow extension bilaterally. Neither postural nor rest tremor was elicited, but finger-to-nose testing elicited a bilateral, 4 Hz action tremor, which was worse on the right (see video 1 on the *Neurology* Web site at www.neurology.org). A postural tremor was present in the right leg with standing. Stride length, arm swing, tandem gait, and turns were preserved.

MRI of the brain demonstrated expanded lateral ventricles, midbrain compression, low lying cerebellar tonsils, and transependymal interstitial edema (figure 1). The cortical mantle was thinned and the fourth ventricle was preserved. While CSF flow studies demonstrated flow across the foramen magnum along both anterior and posterior aspects of the cranio-cervical junction (figure 2A), cross sectional phase contrast MRI documented absent flow in the aqueductal canal, supported by the appearance on sagittal high resolution gradient echo imaging. There were no areas of abnormal gadolinium enhancement. These features were consistent with the diagnosis of AS.³

Endoscopic third ventriculostomy was performed to arrest progression of the hydrocephalus.⁴ Ventriculographic guidance provided visualization of the distended architecture of the lateral ventricle and vascular structures (figure 1D). Six months postoperatively, the patient could hold a drinking glass without spilling, and his headaches had resolved. Follow-up MRI studies demonstrated an open ventriculostomy site (figure 2B), and decompression of the lateral ventricles with expansion of the cortical mantle (figure 2, C and D). On examination, the patient's free-recall and dysmetria (video 2) had improved.

Preoperative axial FLAIR (A, B) and sagittal (panel C) T2-weighted MR images. Note the prominent ventriculomegaly involving the lateral and third ventricles with transependymal edema and preserved architecture of both the fourth ventricle and cerebellum. (D) Endoscopic appearance of dilated right ventricle with distended vascular and connective tissue elements.

DISCUSSION Tremor is an uncommon presenting symptom of occult hydrocephalus. Essential tremor (ET) is the most common cause of tremor in childhood and presents most commonly in the second and sixth decades.⁵ While ET has a predilection for hand and arm involvement, the lack of family history (elicited in greater than 75% of cases) or response to medication trials, along with the associated symptoms of headache and cognitive decline, were features prompting consideration of an alternate diagnosis.

For all causes of obstructive hydrocephalus, age at the time of diagnosis also exhibits a bimodal distribution with an early peak between months 1 and 4 (23% of cases) with a broader distribution spanning ages 8 to 17. A retrospective analysis of 213

patients (age 1 month to 18 years) with obstructive hydrocephalus found headache, vomiting, psychomotor retardation, and gait disturbances to be the most common presenting symptoms.¹ Patients under the age of 30 with chronic AS (present for greater than 6 months) most often complain of headache, while patients over age 63 exhibit dementia, gait disturbance, and urinary incontinence.⁶ Of note, tremor was present in only 12.5% (3/25) of cases and was seen later in the disease course. Also, patients initially diagnosed and treated for triventricular hydrocephalus can present with parkinsonism without tremor, which responds to dopaminergic therapy and shunt revision.⁷

Surgical options for AS include shunting, choroid plexectomy, third ventriculostomy, and aqueductoplasty. Intervention may be warranted if a pattern of gradual clinical decline and transependymal edema on imaging are present. Endoscopic third ventriculostomy (ETV) creates a communication between the third ventricle and the subarachnoid space, and is an option in patients older than 6 months of age.⁸ In a retrospective analysis with 42 months of follow-up, investigators demonstrated that ETV provided durable patency in 90% of patients with AS, excluding cases due to cysts, tumors, or post-hemorrhagic ventriculitis.⁹ Third ventriculostomy can be performed with success rates between 80 and 100%.¹⁰ In cases of late onset AS (n = 31) treated with ETV, improvement occurred in 84% of patients after 26 months.⁶

While stereotactic approaches have significantly reduced the procedure-related mortality associated with open third ventriculostomy (5% vs 27%), current techniques are associated with rare but fatal vascular injuries.^{4,11,12} While decompression of the third ventricle, upward deviation of the brainstem, and reduction in the size of the lateral ventricles are predictable changes postoperatively,¹³ clinical improvement can precede demonstrable radiographic improvement. Improved posture, gait, and cognition were achieved in greater than 50% of patients (32/58) within 2 weeks postoperatively when imaging data were largely unchanged.¹⁴

Our experience suggests that late onset AS should be considered in the differential diagnosis for new onset tremor in adolescence. Furthermore, this case supports a role for endoscopic third ventriculostomy in the treatment of AS.

REFERENCES

1. Cinalli G, Sainte-Rose C, Chumas P, et al. Failure of third ventriculostomy in the treatment of aqueductal stenosis in children. *J Neurosurg* 1999;90:448–454.

Pre- and postoperative cardiac-gated cine-MRI images (A vs B, respectively) demonstrate the absence of CSF flow across the aqueduct of Sylvius and turbulent flow in the third ventricle following ventriculostomy. Axial FLAIR-weighted images obtained 5 months postoperatively demonstrate expansion of the cortical mantle and partial reversal of transependymal edema (C, D); the fluid collection adjacent to the right frontal craniectomy site can be seen.

2. Russman BS, Tucker SH, Schut L. Slow tremor and macrocephaly: expanded version of the bobble-head doll syndrome. *J Pediatr* 1975;87:63–66.
3. Lev S, Bhadelia RA, Estin D, Heilman CB, Wolpert SM. Functional analysis of third ventriculostomy patency with phase-contrast MRI velocity measurements. *Neuroradiology* 1997;39:175–179.
4. Hopf NJ, Grunert P, Fries G, Resch KD, Perneczky A. Endoscopic third ventriculostomy: outcome analysis of 100 consecutive procedures. *Neurosurgery* 1999;44:795–804.
5. Bain PG, Findley LJ, Thompson PD, et al. A study of hereditary essential tremor. *Brain* 1994;117(Pt 4):805–824.
6. Fukuhara T, Luciano MG. Clinical features of late-onset idiopathic aqueductal stenosis. *Surg Neurol* 2001;55:132–136.
7. Zeidler M, Dorman PJ, Ferguson IT, Bateman DE. Parkinsonism associated with obstructive hydrocephalus due to idiopathic aqueductal stenosis. *J Neurol Neurosurg Psychiatry* 1998;64:657–659.
8. Kadrian D, van Gelder J, Florida D, et al. Long-term reliability of endoscopic third ventriculostomy. *Neurosurgery* 2005;56:1271–1278.
9. Feng H, Huang G, Liao X, et al. Endoscopic third ventriculostomy in the management of obstructive hydrocephalus: an outcome analysis. *J Neurosurg* 2004;100:626–633.
10. Kelly PJ. Stereotactic third ventriculostomy in patients with nontumoral adolescent/adult onset aqueductal stenosis and symptomatic hydrocephalus. *J Neurosurg* 1991;75:865–873.
11. McLaughlin MR, Wahlig JB, Kaufmann AM, Albright AL. Traumatic basilar aneurysm after endoscopic third ventriculostomy: case report. *Neurosurgery* 1997;41:1400–1403.
12. Jones RF, Kwok BC, Stening WA, Vonau M. The current status of endoscopic third ventriculostomy in the management of non-communicating hydrocephalus. *Minim Invasive Neurosurg* 1994;37:28–36.
13. Oka K, Go Y, Kin Y, Utsunomiya H, Tomonaga M. The radiographic restoration of the ventricular system after third ventriculostomy. *Minim Invasive Neurosurg* 1995;38:158–162.
14. Tisell M, Tullberg M, Hellstrom P, Blomsterwall E, Wikkelsö C. Neurological symptoms and signs in adult aqueductal stenosis. *Acta Neurol Scand* 2003;107:311–317.

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