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Surgical management of arrested hydrocephalus: Case report, literature review, and 18-month follow-up

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ABSTRACT

Objective: Arrested hydrocephalus is stable ventriculomegaly without evidence of neurologic deterioration or symptoms. Management of arrested hydrocephalus in asymptomatic adults is controversial, with little clinical data. This case highlights the potential for decompensation in adults with arrested hydrocephalus and reviews the literature regarding pathophysiology and management of this clinical entity.

Patients and methods: A 39 year-old gentleman with arrested hydrocephalus incidentally found during work-up for new-onset seizure and managed conservatively for ten years presented with increasing headache, memory loss, gait instability and urinary and fecal incontinence. Stable massive triventriculomegaly was documented on serial brain imaging, and ophthalmologic exam revealed no papilledema.

Results: The patient underwent endoscopic third ventriculostomy with immediate post-operative improvement of headache, resolution of incontinence, and cessation of seizures. At 15 months after surgery, neuropsychiatric testing demonstrated improvement in visuomotor skills, problem solving, verbal fluency and cognitive flexibility compared to his pre-operative baseline. At 18 months after surgery he remained seizure free with full continence and significant improvement in headaches.

Conclusion: Early recognition of arrested hydrocephalus and its potential for decompensation may prompt surgical treatment and prevent neurologic deterioration.

1. Introduction

Adult hydrocephalus is a heterogeneous clinical entity that likely encompasses multiple syndromes with varied etiologies. This is reflected in the literature, where various groups have referred to the condition as "arrested hydrocephalus," "asymptomatic hydrocephalus," "occult hydrocephalus," "compensated hydrocephalus," and more recently, "long-standing overt ventriculomegaly of adulthood (LOVA)," or "late-onset idiopathic aqueductal stenosis (LIAS)" [1–5]. These diagnoses share a radiographic finding of stable, enlarged ventricles and a clinical history of normal development and neurologic function. Patients rarely present with symptoms or stigmata of elevated intracranial pressure, and diagnosis of hydrocephalus is often incidental and unexpected.

The chronicity and natural history of adult hydrocephalus is unclear. Historical case series indicate that children who are hydrocephalic can spontaneously "arrest" and as many as half achieve both normal head circumference and cognitive ability into adulthood [1,6,7]. Idiopathic occlusion of the sylvian aqueduct is also thought to cause adult hydrocephalus, although the timing and pathophysiology of this event is not understood [8]. Regardless of the underlying cause, arrested hydrocephalus is frequently viewed as a stable condition and some clinicians have advocated a conservative approach with serial imaging and watchful waiting [6,9–11].

On the other hand, clinicians have also reported that patients with arrested hydrocephalus experience subacute neurologic decline and progressive ventricular enlargement even after lengthy periods of stability [9,12]. These patients are said to have "decompensated," and are considered candidates for surgical treatment. More recently, neuropsychiatric methods have suggested that long-standing ventriculomegaly without clinical deterioration can result

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in subtle neurologic sequelae [3,13]. Chronic hydrocephalus has also been linked to sub-clinical cellular and molecular damage [14,15].

Optimal management of adult patients with arrested hydrocephalus is widely debated. Here, we describe a case of arrested hydrocephalus with delayed decompensation and review the literature to date on this controversial clinical entity.

2. Patient and methods

2.1. History and examination

A 30 year-old gentleman presented to the neurology department with new-onset generalized tonic-clonic seizures. As part of a routine work-up for epilepsy, a brain magnetic resonance imaging (MRI) was obtained. This revealed ventriculomegaly of the lateral and third ventricles and possible aqueductal stenosis (Fig. 1). A spot electroencephalogram (EEG) showed global slowing but no focal epileptic discharges. His neurologic exam was non-focal and fundoscopic exam revealed no papilledema. Antiepileptic medication was initiated, and he was referred for neurosurgical consultation. Decision was made to proceed with expectant management due to his benign clinical history.

Over the next five years, the patient began to complain of problems at work as a custodian. Because of a perceived cognitive decline, neuropsychological testing was conducted which revealed borderline Full Scale IQ (78) and low-average General Ability Index (81). He had weakness in processing speed, executive functioning, and verbal fluency. Memory tests demonstrated a range from impaired to below average. He did not report any symptoms concerning for increased intra-cranial pressure, and his neurologic decline was not felt to be related to his stable ventriculomegaly.

Nine years after his initial presentation, he began to complain of significant headache, memory decline, urinary and fecal incontinence, and difficulty with ambulation. He was asked to return to neurosurgery clinic for further evaluation. On physical exam, he had a narrow, unsteady gait and he was unable to tandem walk. His memory was severely impaired. Ophthalmologic examination at this time revealed no optic nerve swelling or pallor. At this point, the patient was felt to have had a significant decompensation secondary to hydrocephalus and was offered surgical treatment.

2.2. Imaging

Several images were obtained during course of his nine-year observation period in the context of emergency department visits for seizure and headache. Repeat head CT did not show change in ventricular size (Fig. 2). No transependymal flow was seen. Spinal MRI revealed no intraspinal pathology.

2.3. Management

With evidence of clinical decompensation, ventriculoperitoneal shunt and endoscopic third ventriculostomy were discussed as surgical options. Endoscopic third ventriculostomy was primarily favored for two reasons. In the setting of long-standing ventriculomegaly, we felt that shunt placement carried significant risk of over-drainage and subdural hematoma formation. In addition, endoscopic third ventriculostomy avoids the risks associated with implanting a foreign body, such as infection and malfunction. The possibility of shunt placement in the event of failed third ventriculostomy was explicitly discussed.

2.4. Operation

We used a standard surgical approach for endoscopic third ventriculostomy. In brief, we made a small curvilinear incision just above coronal suture. Burr hole was placed on the coronal suture approximately twenty-five millimeters lateral to midline. Upon initial ventricular cannulation we obtained an opening pressure that was greater than 30 cm of water. The septum pellucidum appeared fenestrated like a “tattered sail.” Standard third ventriculostomy was performed on the floor of third ventricle using endoscopic spreading forceps.

2.5. Post-operative course

One-month after surgery our patient reported complete resolution of incontinence and headaches with noticeable improvement in his gait. He reported having no seizures since his surgery. His family reported that cognition and memory remained at his pre-operative baseline. Post-operative MRI demonstrated flow through the third ventriculostomy (Fig. 3).

3. Results

At fifteen months after surgery formal neuropsychiatric testing was repeated. This patient’s full scale IQ modestly increased to 80 from 78. Specifically, he had improved performance on visuomotor skills, problem solving, verbal fluency and cognitive flexibility compared to his pre-operative baseline. At eighteen months after surgery he remained seizure free and fully continent. His headaches significantly improved, and he was weaned off of topiramate.

In retrospect, we were overly reassured by this patient’s stable imaging and relatively preserved mental status. We believe that

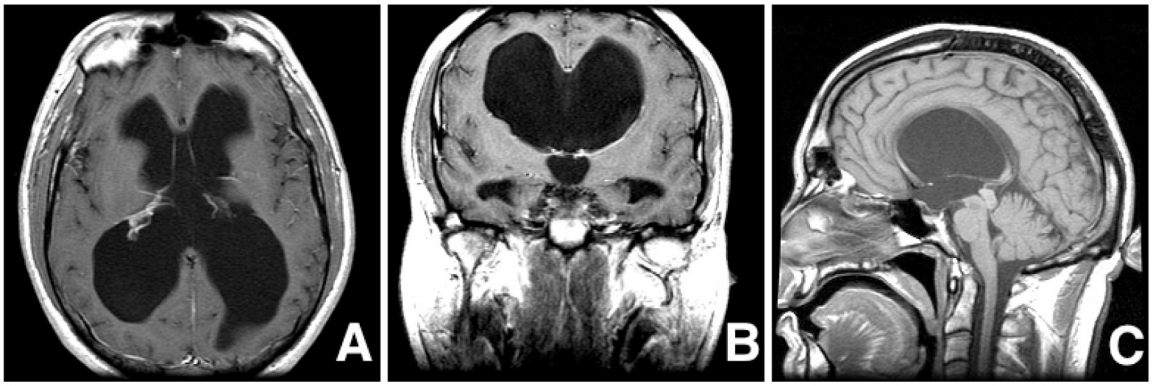

Fig. 1. Presenting MRI of the brain obtained during work-up of new seizures. Axial (panel A), coronal (panel B) and sagittal (panel C) slices demonstrate hydrocephalus involving the lateral and third ventricles.