Article published online: 2025-01-20

Late Presentation of Congenital Aqueduct Stenosis in Adulthood with High-Riding Basilar Artery: Case Report

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J Neurol Surg Rep 2025;86:e4–e7.

Abstract

Background A rare variant of congenital aqueductal stenosis (CAS) is known as adult-onset CAS, characterized by the emergence of symptoms during adulthood.

Case Description A 35-year-old man presented complaining of acute-onset headache and vomiting. Magnetic resonance imaging of the brain revealed an acute hydrocephalus due to an aqueductal web. The patient was treated with a ventriculoperitoneal shunt and doing well. His condition was complicated by subdural hematoma as a result of overshunting, which was treated by frontoparietal craniotomy and eventually, he underwent valve revision to a programmable valve.

Conclusion Additional research is needed to gain a deeper understanding of the hypothesis related to adult aqueductal stenosis. This will help shed light on the asymptomatic childhood phase and potentially reduce complications by identifying late CAS early.

Keywords
- aqueductal
- stenosis
- congenital
- high-riding basilar
- subdural
- hematoma

Introduction

Congenital aqueductal stenosis (CAS) is the most common form of noncommunicating hydrocephalus. A complete or partial obstruction of cerebrospinal fluid (CSF) happens at the aqueduct of Sylvia's during fetal life, which results in the dilation of the lateral and third ventricles with normal-sized fourth ventricle. Three quarters of CAS patients had unknown etiology, though there were many etiologies of this disorder including infection, hemorrhage, intoxication, deficiencies, genetic factors, central nervous system malformation, and tumors.[1] Some patients with CAS might not show any symptoms until adulthood, or there might be mild symptoms that might pass unnoticed or undocumented. Usually in a small percentage of acute onset in adults, symptoms may be present for 1 to 4 weeks. Symptoms would include headache, nausea, vomiting, visual disturbances, seizures, changes in mental state, and coma. More often the onset is subacute where the symptoms from less than 6 months or chronic for more than 5 months, the symptoms would be characterized by intracranial hypertension syndrome progressively taking hold.[2] Given the diverse etiologies and variable presentation of CAS ranging from asymptomatic cases in early life to acute or subacute symptom onset in adulthood, the management and long-term follow-up of such cases present significant clinical challenges. Against this backdrop of complexity, this study describes a case of adult-onset CAS treated with shunt surgery, with an 11-month follow-up.

received
March 6, 2024
accepted after revision
June 5, 2024

DOI https://doi.org/10.1055/s-0044-1788037.
ISSN 2193-6358.

Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany

Case Presentation

In 2023, a 35-year-old man known case of allergic sinusitis presented to the emergency department complaining of severe headache for 2 days and vomiting several times for 1 day. Clinical examination revealed a normal neurological examination. Computed tomography (CT) of the brain without contrast showed lateral and third ventriculomegaly associated with enlargement of the proximal aqueduct of Sylvius and normal-sized fourth ventricle.

While the endoscopic third ventriculostomy (ETV) procedure was initially considered due to its high rate of success, the decision was ultimately made to insert an emergency external ventricular drain (EVD) instead. This was due to the patient presentation with a high-riding basilar artery, as showed in ►**Fig. 1**, which increased the risk of vascular injury from the ETV—a risk that carries potentially fatal consequences. The patient improved clinically afterward. CSF analysis was unremarkable. Postoperative magnetic resonance imaging (MRI) three-dimensional (3D) fast imaging employing steady-state acquisition of the brain showed marginal reduction in the lateral and third ventriculomegaly and a thin tissue membrane separating the dilated aqueduct from the normal-sized fourth ventricle, in keeping with the aqueductal web (►**Fig. 1**).

Afterward, EVD was removed and a ventriculoperitoneal shunt medium pressure valve was inserted. On follow-up, after 13 days, the patient complained of blurring of vision. Accordingly, optical coherence tomography (OCT) was done, which showed mild bilateral papilledema.

After 3 months postoperatively, he was readmitted with a history of multiple motor vehicle accidents, complaining of difficulty concentrating and dysgraphia. His clinical examination was intact except for an equivocal Babinski sign. He underwent an urgent CT scan of the brain that showed a large left subdural isodense hematoma. He underwent emergent frontoparietal craniotomy with hematoma evacuation and opening of septations. Postoperative CT of the brain showed significant radiological regression.

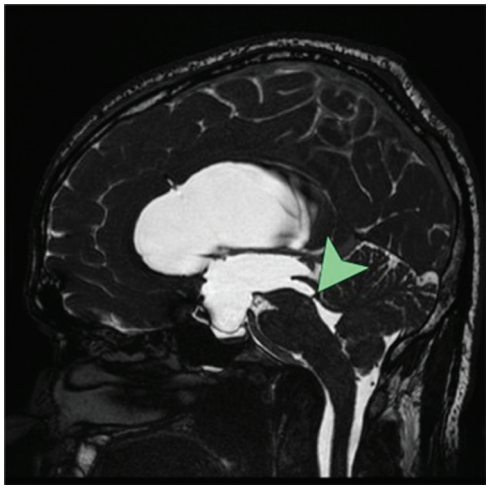

Fig. 1 Postoperative MRI 3D FIESTA of the brain indicates aqueductal stenosis. Sagittal 3D FIESTA sequence of the brain shows marginal reduction in the lateral and third ventriculomegaly accompanied by the normal-sized fourth ventricle. There is a thin tissue membrane (green arrowhead) separating the dilated aqueduct from the normal-sized fourth ventricle, in keeping with the aqueductal web. FIESTA, fast imaging employing steady-state acquisition; MRI, magnetic resonance imaging; 3D, three-dimensional.

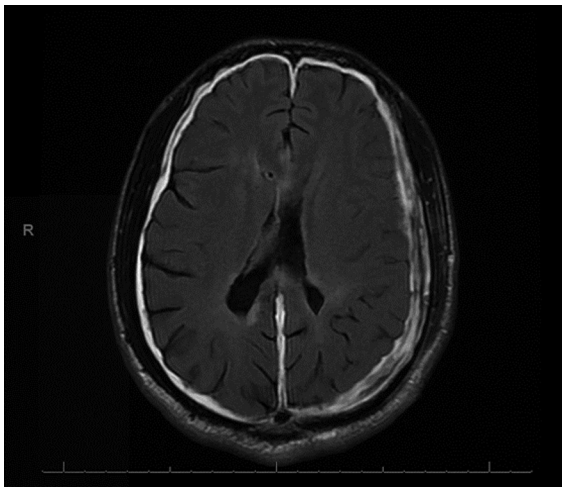

Fig. 2 MRI of the brain reveals a left recollection of subdural hematoma. Axial T2-MRI of the brain showed a left holohemispheric subdural hypointense hematoma measuring about 1.9 cm in maximum depth with midline shift measuring about 0.3 cm. MRI, magnetic resonance imaging.

After 2 months of follow-up, an MRI of the brain revealed a recollection of left side hypointense chronic subdural hematoma (CSDH) (►**Fig. 2**). Accordingly, the medium pressure valve has been replaced with a programmable valve, which the patient has become stable and clinically improved after.

Discussion

CAS is inherited primarily in an X-linked fashion and shows male predominance.[3] CAS is 0.5 to 1 per 1,000 births in the pediatric group.[4] CAS became a well-documented condition within the young population. However, late presentation is a rare phenomenon where the incidence of CAS in adults has not been documented yet, despite this there are few case reports and case series regarding late CAS.[4,5]

Patients with aqueductal stenosis (AS) present with headaches, seizures, vomiting, visual problems, postural disturbances, incontinence, impairment in wakefulness, cognitive function, and endocrine dysfunction.[6–9] Uncommon symptoms include CSF rhinorrhea, trigeminal neuralgia, hearing loss, and vertigo.[7,8,10] Older children, adolescents, and adults could present in acute (symptoms for 1–4 weeks), subacute (symptoms for less than 6 weeks), or chronic (symptoms for more than 6 weeks) manner.[1]While the clinical picture is similar, the literature shows variability according to age. Due to the elevation of intracranial pressure, headaches are the most encountered sign and the most notable in the young population.[1,10] The older populations' symptoms tend to mimic those of normal pressure hydrocephalus, such as urinary and fecal incontinence and impairment in gait, posture, memory, and wakefulness.[2,8,9,11,12] However, one

study stated the occurrence of these symptoms in most patients regardless of age but are milder in the younger population and are overshadowed by headaches and other signs of increase intracranial pressure.[9] The significance of symptom duration and severity regarding the prognosis was noted in one study in which abrupt and severe onset of symptoms resulted in poorer outcomes.[5] In contrast, another study indicated the irrelevance of symptom duration regarding age or severity.[9]

Few hypotheses were proposed regarding the relative asymptomatic childhood and symptomatic adulthood. One hypothesis suggests that the decompensation of a prior compensated hydrocephalus causes the late onset of symptoms.[8] Another hypothesis explains a two-hit process in which the increased resistance of CSF flow due to deep white matter ischemia can participate in manifesting the symptoms during adulthood.[12,13] The subject of our case presented signs of increased intracranial pressure consisting of a severe headache spanning over the course of 2 days and several vomiting episodes with no impairment in visuals, gait, balance, wakefulness, defecation, or voiding. Rodis et al recently proposed a classification of AS into four clinical subtypes, I to IV (based on patient age). In our case, the patient met type III AS (acute type), which is more frequent in adolescence and early adulthood. Also, headaches and signs of raised intracranial pressure were predominant symptoms of this type.[1]

Generally, advanced MRI techniques are needed in order to diagnose and determine the etiology of the stenosis. Phase-contrast MRI aids in evaluating aqueductal patency.[14] Recent studies have shown that 3D (sampling perfection with application optimized contrast using different flip angle evolutions) with the variant flip angle mode technique is beneficial in demonstrating luminal morphology and alone is usually sufficient for the diagnosis of AS.[15,16] The typical morphology of AS includes funneling of the aqueduct above the site of stenosis, lateral and third ventriculomegaly, and an upward bowing of the corpus callosum.[1,17] In our case, CT of the head revealed enlargement of the lateral and third ventricles, a proximal dilation of the aqueduct of Sylvius preceding the stenosis, and a normal-sized fourth ventricle.

Currently, three surgical treatment options are available: ETV, shunt surgery, and endoscopic aqueductoplasty. ETV operation, which re-establishes the physiological CSF route, is characterized by fewer complications, and the need for a revision is low, making ETV the preferred treatment option. Shunt surgery is highly effective; it can be used as a first-line or second-line treatment option in the cases of recurrence after ETV or if ill-suited anatomical conditions are present, for instance in ►**Fig. 1**, a high-riding basilar artery is located anterior to the pons and closely adjacent to the floor of the third ventricle (tuber cinereum), making ETV unfavorable. However, in shunt surgery, there is a high complication rate, and revisions are common.[17] Endoscopic aqueductoplasty is considered to be a substitute for ETV in a very selected patient group, such as those with short AS and membranous occlusion. The reobstruction and complication rate are high, given the critical nature of the mesencephalon; structures such as the tegmentum and the tectum can be affected, resulting in oculomotor, trochlear nerve palsies, and Perinaud's syndrome.[1] In this case, an anatomical variation was present, the patient had a narrow prepontine cistern and a high-riding basilar artery. This played a role in the treatment option of CAS where in most individuals, the basilar artery bifurcates into the two posterior cerebral arteries at the upper border of pons. In the case of a high-riding basilar artery, the aforementioned bifurcation happens directly under the floor of the third ventricle, making the ETV operation which fenestrated the floor of the third ventricle a high-risk operation. Therefore, shunt surgery was preferred in our case. Other surgical approaches mentioned in the literature concerning the presence of a high-riding basilar artery or an option in the case of the presence of a high-riding basilar artery include ETV using a Penumbra Artemis Neuro Evacuation Device and extra-axial ETV.[18,19]

A follow-up took place 13 days postoperative. The report subject complained of having blurred vision; therefore, an OCT was performed, showing mild bilateral papilledema. As hydrocephalus progresses, a set of complications arise. Neuro-ophthalmic complications include papilledema resulting in decreased central visual acuity due to the enlargement of the concentric physiological blind spot in the acute presentation; however, in the chronic presentation, preservation of central vision is seen until the late stages due to the ischemic process affecting the arcuate nerve fiber bundle first before the papillomacular bundle that surrounds the macula which is responsible for the high-resolution and colored central vision. Transient visual obscuration can also be seen; it can be triggered by movement, the patient might experience sudden blurring of the vision or a complete grayout, but the restoration of vision is always swift and complete.[20]

Subdural hematomas can be another complication arising from shunt surgery in the case of overdraining. The pathogenesis includes the rise in negative pressure caused by overshunting, resulting in bridging veins becoming vulnerable due to the increased strain caused by the separation of the brain from the dura. Osmotic mechanisms also participate, facilitating the growth of the subdural collection.[21] In our case, after approximately 2 and a half months of placing the V/P shunt, the patient was complicated by CSDH due to overshunting.

This case underscores the importance of individualized treatment approaches, especially in patients with unique anatomical variations such as a high-riding basilar artery. However, the reliance on findings from a single case poses limitations, particularly regarding the generalizability and applicability of the treatment strategies to a broader patient population. Consequently, there is a critical need for further research, including larger cohort studies or controlled trials, to validate these findings and treatment recommendations. Such studies would contribute to establishing more definitive surgical options for CAS, ensuring safer and more effective management strategies for these patients, especially those presenting with or at risk of complex complications.

Conflict of Interest
None declared.

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