

Endoscopic Third Ventriculostomy for Hydrocephalus Due to Unruptured Pineal AVM: Case Report and Review of the Literature

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Abstract

Introduction Arteriovenous malformations (AVM) are considered to be static congenital lesions; however, a subset may possess dynamic pathophysiological capabilities of growth, regression or other morphological changes with time. We report on an adult patient harboring a pineal AVM who presented with progressive symptoms of obstructive hydrocephalus and was successfully treated by endoscopic third ventriculostomy (ETV).

Clinical presentation This 63-year-old man was incidentally diagnosed 8 years previously with an asymptomatic unruptured pineal AVM and followed conservatively until he developed a progressive impairment in consciousness with gait apraxia and incontinence over a period of 2 months. Magnetic resonance imaging (MRI) revealed obstructive hydrocephalus due to nidal compression at the level of the aqueduct. Treatment by ETV resulted in resolution of neurological deficits and ventriculomegaly without evidence of complications at 18-months follow-up.

Conclusion Treatment by ETV in adults with obstructive hydrocephalus due to deep pineal AVMs is a reasonable option in selected cases. A discussion of the pathological mechanisms and therapeutic options for this rare entity is presented.

Keywords

- ▶ endoscopy
- ▶ third ventriculostomy
- ▶ vascular malformation
- ▶ hydrocephalus.

Introduction

Arteriovenous malformations (AVM) are not entirely static and recent reviews have shown that a subset of AVMs can possess clinically dynamic capabilities of growth, remodeling, regression, and even de novo reappearance after complete resection or radiosurgery.^{1–4} Although AVMs typically present in young adults with hemorrhage, symptomatic obstructive hydrocephalus in adults is relatively rare and is usually due to AVM rupture with subarachnoid hemorrhage and mechanical blockage of the cisterns or arachnoid villi.^{5–7} We describe a case of an unruptured pineal AVM in an adult

who presented with progressive hydrocephalus and was successfully treated by endoscopic third ventriculostomy (ETV).

Case Presentation

History and examination

This 63-year-old right-handed man was originally incidentally diagnosed with an asymptomatic pineal AVM 8 years prior at another institution and managed conservatively. Follow-up radiographic studies 1 year prior to presentation revealed mild ventriculomegaly, which was reported to have remained

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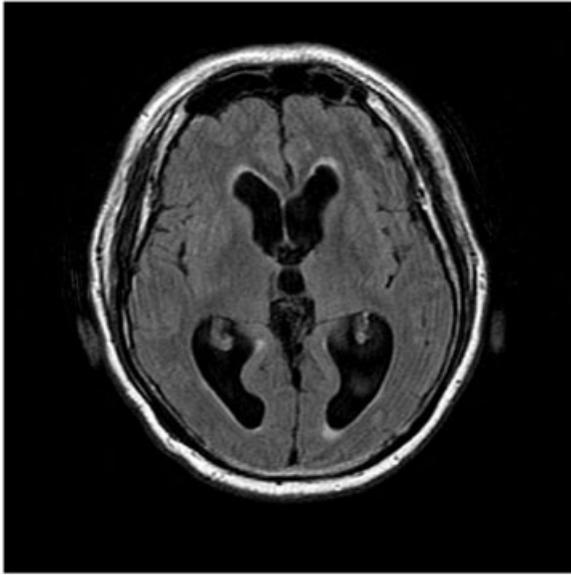


Fig. 1 Axial FLAIR magnetic resonance image obtained from another institution 1 year prior to presentation showing an irregular pineal lesion with relatively moderate ventriculomegaly.

stable over time (►Fig. 1). Past medical history was significant for hypertension, angina pectoris, and cervical spondylosis with left upper extremity proximal weakness. Two months prior to admission, the patient presented to our emergency room with a complaint of a sudden onset of dizziness. The neurological exam was unremarkable. Head computerized tomography (CT) demonstrated a 21 × 11 mm pineal body mass with dilated third and lateral ventricles and a fourth ventricle within the normal limits (►Fig. 2A). The patient was released on the same day with the intention of follow-up magnetic resonance imaging (MRI) but failed to present for the next exam. One month later, the patient was apprehended by the police for illicit behavior and brought to our outpatient clinic for inspection. Neurological exam was significant for a mild disturbance of consciousness with no complaints of headache, nausea, or vomiting. He refused treatment and was discharged the same day. Two weeks later,

the patient was found confused, unable to walk, and incontinent in his home by public officials and was transferred to our institution for treatment.

On admission, the patient was afebrile with normal vital signs and no headache, nausea, or vomiting. The initial neurological exam was significant for a mild disturbance of consciousness with deficits in higher cognitive function characterized by short-term memory loss (Hasegawa Dementia Scale-Revised [HDS-R] 13/30, Mini-Mental State Examination [MMSE] 17/30), acalculia, and difficulties in self-care, ataxic gait, and bowel and bladder incontinence. Pupils were 2.5 mm isocoric and symmetric, but sluggish to light with normal convergence. Left grip weakness (0 kg) was unchanged from prior exams. MRI and MR angiography as well as head CT angiogram demonstrated a pineal AVM with a small nidus fed by the right posterior choroidal and pericallosal arteries and dilated veins draining into the deep galenic venous system (►Fig. 2B, C). No intraparenchymal or intraventricular hemorrhage was observed. Preoperative considerations for a cause of obstruction at this location included direct compression by the nidus or a dilated draining vein. Because of the patient's progressive course yet relatively stable clinical condition with moderate symptoms of obstructive hydrocephalus, elective ETV was scheduled 1 week after admission.

Treatment

A standard ETV via a right frontal trajectory using a flexible fiberscope (N-4L, Machida Co., Japan) was performed. The right lateral ventricle was cannulated using a peel-away sheath. Intraoperative findings demonstrated a nidal mass bulging from the posterior rostral portion of third ventricle wall forming a slit-like occluded orifice at the mouth of the aqueduct (►Fig. 3A). Fenestration was performed in the floor of the third ventricle at the center of the tuber cinereum (►Fig. 3B). After catheter puncture and balloon dilation using a balloon dilator (Fuji Systems Co., Japan), fenestration was achieved as evidenced by visualization of good to-and-fro cerebrospinal fluid (CSF) movement with floor pulsations. Finally, the basilar artery and prepontine structures were identified.

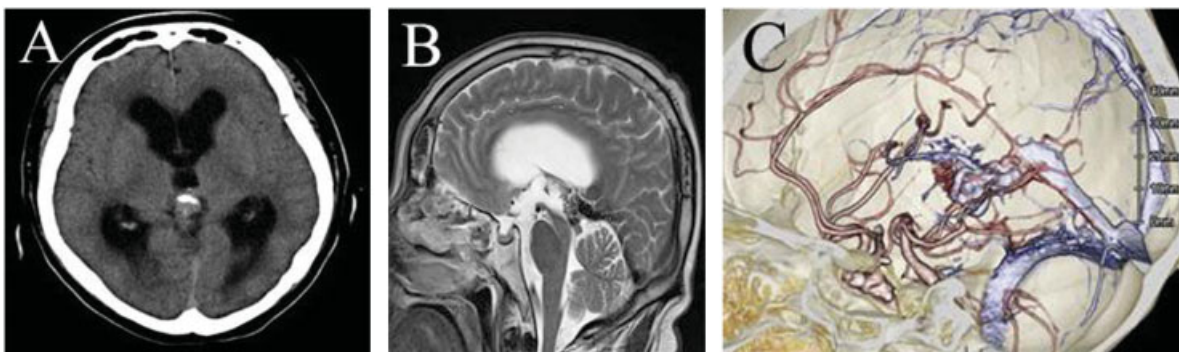


Fig. 2 (A) Head computerized tomography (CT) 2 months prior to admission showing a 21 × 11 mm pineal body mass with dilated third and lateral ventricles and a fourth ventricle within the normal limits. (B) Sagittal T2-weighted magnetic resonance image demonstrates a flow void within the pineal body region with aqueductal stenosis. (C) Head CT angiogram demonstrates a pineal AVM with a small nidus fed by the right posterior choroidal and pericallosal arteries and dilated veins draining into the deep galenic venous system.

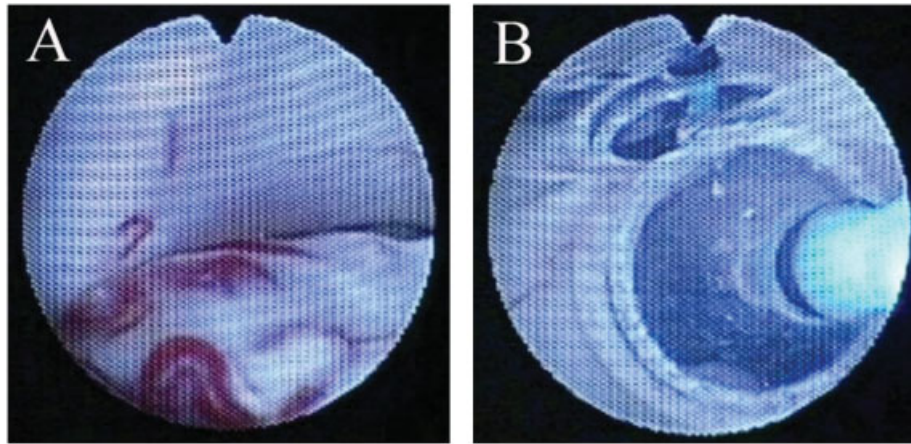


Fig. 3 (A) Intraoperative findings demonstrated a nidus-like vascularized mass in the posterior rostral third ventricle wall with evidence of occlusion of the aqueduct. (B) Fenestration via balloon dilation with a balloon dilator was made in the floor of the third ventricle at the center of the tuber cinereum.

Posttreatment course

Postoperative head CT showed no evidence of intraventricular hemorrhage or other bleeding. The patient's gait disorder and consciousness level improved on discharge with independence in daily activities and an improvement in the HDS-R and MMSE-R measures (13/30 to 17/30 and 17/30 to 22/30, respectively). Follow-up exam at 6 months demonstrated almost normal neurological exam and resolution of the ventriculomegaly on head CT (►**Fig. 4**). The patient has remained in stable condition throughout 18 months of follow-up.

Discussion

AVMs have been described as congenital high-flow shunts between arteries lacking a muscularis layer and veins with a

central tangled nidus devoid of capillaries.⁸ Although the underlying pathogenesis remains unclear, AVMs have been originally considered to be due to persistence of primordial vascular connections by Dandy in 1928, and later Yasargil presented a theory relating to an abnormality of capillary formation.^{9,10} Pathological examination has shown that over time, feeding arteries accumulate fibromuscular tissue and the draining veins become dilated and thickened.¹¹ This morphological change over time has been postulated to be due to molecular mechanisms involving growth factors that may cause AVM formation and remodeling, whereas mechanical blood flow stresses from shunting leading to hemorrhage, arterial hypotension, and venous hypertension have been suggested to cause angiogenesis. Moreover, some have postulated that AVMs may be slow-growing vascular tumors or the result of aberrant adult vasculogenesis related to acquired somatic mutations.¹

Unruptured cerebral AVMs are rarely associated with obstructive hydrocephalus in adults, whereas unruptured nongalenic pial cerebral AVMs associated with obstructive hydrocephalus in neonatal patients are relatively well known.^{6,12} These congenital lesions typically cause ventriculomegaly via mechanical obstruction from dilated deep draining veins or by the nidus itself.¹³ Unusual cases of hydrocephalus due to CSF overproduction by choroidal AVMs have also been reported.¹⁴ To date, there have been 10 cases of adult patients with AVMs presenting with hydrocephalus due to direct obstruction in the area of the foramen of Monro or the aqueduct either by draining veins (such as a dominant thalamostriate-internal cerebral vein complex or infratentorial AVMs with a dilated vein of Galen) or obstruction by the nidus itself.^{6,7,13,15–17} Gibrasert and colleagues have also found one adult patient with an unruptured AVM who presented with hydrocephalus without evidence of mechanical outflow restrictions, presumably due to distant venous outflow obstruction leading to venous congestion in posterior fossa pial veins with accompanying alterations in the hydrodynamic relationship between the intraventricular spaces and the medullary veins.¹³ They further postulated



Fig. 4 Follow-up head computerized tomography at 6 months demonstrated resolution of the ventriculomegaly.

that venous outflow obstruction may have indirectly lead to increased pressure within the posterior fossa veins and deep venous system, which in turn caused decreased CSF reabsorption and resultant hydrocephalus.¹³ This form of mal-resorptive hydrocephalus is in contrast to reports of adult cases of pial high-flow AVMs with intracranial hypertension not associated with hydrocephalus postulated to be due to venous outflow obstruction.¹⁸ Although pathological specimens were not taken in our case, we postulate that the AVM in our case underwent changes in morphology leading to mechanical aqueductal stenosis and progressive symptomatic hydrocephalus. Although the mechanism is unknown, chronic venous hypertension could have led to arterial tissue hypoxia with resultant angiogenesis and nidus size increases and resulting obstructive hydrocephalus.

Treatment of hydrocephalus has traditionally been performed by ventriculoperitoneal (VP) shunt placement. In the most recent series report of eight patients with hydrocephalus due to unruptured AVMs, all patients were treated with VP shunt placement.¹³ However, results showed a high rate of malfunction due to shunt obstruction and a high rate of symptomatic overdrainage (subdural hematoma [SDH], hygroma, and slit ventricles), possibly due to the effects of excessive decreases in ventricular pressure and increased dilation in the draining veins. In patients with vein of Galen AVMs and hydrocephalus treated by shunt insertion, complications of SDH, hygroma, or dilation of draining veins have been described to be due to an altered hydrodynamic equilibrium whereby venous outflow restrictions cause less reabsorption of CSF and ventricular dilation. In another report, a patient with a posterior fossa AVM developed hydrocephalus and was treated by shunting. However, postoperatively, a consciousness deficit and Parinaud syndrome were observed, presumably due to overdrainage with upward herniation and distortion of the mesencephalon.⁸ Not surprisingly, all symptoms resolved after shunt ligation. In our case, despite the close proximity of the AVM to the tectal region, there was no evidence of altered eye movements or Parinaud syndrome, and no obvious radiographic evidence of brain stem mass effects. However, to explain the signs of impaired consciousness that occurred in our case, it is plausible that the AVM could have been exerting sufficient mass effects that were not radiologically visible. More likely, hemodynamic changes within the AVM may have caused obstructive hydrocephalus and subsequent impaired consciousness with associated gait disturbance and incontinence.

Although definitive treatment of hydrocephalus has been accomplished by direct obliteration of the AVM (via surgical or endovascular or endoscopic assisted techniques),^{13,19,20} it is frequently difficult to treat deep-seated AVMs by direct surgery, and radiosurgical considerations would be ineffective for treating acute symptomatic hydrocephalus because of the insufficiently slow volume reduction time. In our case, based on the relatively small nidus size, one possible option would have been radiotherapy, especially prior to the onset of symptoms or after ETV; however, the patient had refused such treatment in the past and chose post-ETV conservative management.

Another concern in treating AVMs associated with hydrocephalus is regarding lesions involving either the foramen of Monro or the floor of the third ventricle, which would not be applicable to ETV treatment, excepting the possibility for endoscopic fenestration (septostomy) for cases of unilateral foraminal obstruction. Although VP shunts with gravitational devices are one possible solution to avoid the potential complications encountered with treatment by standard shunting, when technically possible, we believe that ETV is a more appropriate solution, especially for cases of adult-onset obstructive hydrocephalus due to unruptured AVMs involving aqueductal stenosis because these lesions typically demonstrate a slow process of obstruction at the aqueduct, either by direct mechanical obstruction or by distant venous congestion leading to mal-resorptive hydrocephalus. Although the follow-up period was short in our case, ETV may prove to provide long-term efficacy in cases of both acute or progressive aqueductal obstructive hydrocephalus associated with mass lesions in the pineal region. The mechanism for effective outcome of ETV in these cases may be due to a gradual resolution in altered venous-CSF ventricular hydrodynamics with relatively slower shrinkage in the size of the draining veins or nidus. Based on this reasoning, ETV may provide a more physiologic hemodynamic environment capable of lessening the more abrupt effects and complications found with VP shunting. However, further studies and scientific proof are needed to substantiate this hypothesis.

Conclusions

This case highlights the dynamic nature of AVMs and the potential for development of progressive hydrocephalus due to pineal AVMs. Although more long-term follow-up studies and further investigation of the pathophysiological mechanisms of hydrocephalus in AVMs are needed, ETV can be a safe and effective treatment option in selected patients. Sufficient access to the third ventricle floor is a prerequisite for endoscopic management.

Conflict of Interest

None

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