Unruptured arteriovenous malformation in a patient presenting with obstructive hydrocephalus

Case report and review of the literature

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✓The authors report on a patient harboring an unruptured cortical arteriovenous malformation (AVM), who had presented with obstructive hydrocephalus due to compression of the cerebral aqueduct by a large venous varix. Although patients with ruptured AVMs are known to either present with or later suffer from obstructive hydrocephalus, those with unruptured AVMs who present in this manner are quite rare. Moreover, hydrocephalus caused by a venous varix draining an AVM, to our knowledge, has never been previously reported in the literature. This report serves to illustrate two primary points, namely, that tortuous venous varices draining AVMs can result in obstructive hydrocephalus and that this unusual circumstance can be fostered in the setting of venous outflow obstruction.

KEY WORDS • obstructive hydrocephalus • arteriovenous malformation • varix • shunt

ARTERIOVENOUS MALFORMATIONS represent the most common intracranial vascular malformation as classified by McCormick in 1966.⁶ As a whole, vascular malformations are rarely associated with hydrocephalus.^{2,7–10} In patients with AVMs, hydrocephalus occurs almost exclusively in the setting of subarachnoid or intraventricular hemorrhage; the notable exception is an AVM of the vein of Galen, which is seen in the pediatric population.

Obstructive hydrocephalus due to a venous varix in an unruptured AVM has not been previously reported in the literature. The patient described herein suffered this rare phenomenon and serves to emphasize that varices, besides being critical from a hemodynamic standpoint, can impede by mass effect the normal pathway of CSF circulation, and their presence usually suggests stenosis in the venous outflow of the AVM.

CASE REPORT

History and Examination. This 55-year-old man with a 6-month history of mild, nonspecific headaches suffered a near-drowning accident while on vacation. He was found face down off the ocean shore and received cardiopul-

monary resuscitation before being transported to an area hospital. On arrival, a CT scan of the brain revealed a left-sided parietooccipital AVM causing obstructive hydrocephalus. An endotracheal tube was inserted in the patient for presumed aspiration and airway protection, and he was sedated and airlifted to our institution.

On admission, he had a low-grade fever but otherwise normal vital signs. His Glasgow Coma Scale score was 5T, and an initial neurological examination revealed pupils that were sluggish but symmetric to light, a weak corneal reflex, a strong gag reflex, and minimal withdrawal to central pain in all four extremities. A CT angiogram demonstrated a left parietooccipital AVM with a 3-cm nidus (Fig. 1). A complex draining pattern was noted with extensive venous varices. One varix was encroaching on the quadrigeminal cistern, and thus the sylvian aqueduct was compressed. No intraparenchymal or intraventricular hemorrhage was noted, however, and the entire AVM was thought to lie supratentorially.

Treatment. A right frontal external ventricular drainage catheter was inserted, and the patient's neurological function improved markedly to an extent that he was spontaneously opening his eyes and moving his extremities to commands. Despite a 2-week course of external drainage, his ventricles did not decrease in size and he ultimately underwent a shunt insertion procedure.

Abbreviations used in this paper: AVM = arteriovenous malformation; CSF = cerebrospinal fluid; CT = computed tomography.

FIG. 1. Axial (a), sagittal (b), and coronal (c) CT angiograms demonstrating a varix originating from the AVM nidus, undulating, and becoming most dilated at the left atrium. It then drains into the vein of Galen, where venous stenosis and subsequent outflow obstruction can be seen.

Deep venous thromboses treated with inferior vena cava filter placement and aspiration pneumonia that lead to a prolonged period of intubation complicated his hospitalization. Three weeks after his admission, he had recovered his neurological baseline functions, and he was transferred to a rehabilitation hospital shortly thereafter.

Posttreatment Course. The patient returned to our clinic for further counseling and was recovering quite well. He reported that he was ambulating and functioning well with only mild short-term memory problems and complete resolution of the chronic headaches he had experienced before his hospitalization. In the interim, he also underwent cerebral angiography (Fig. 2) to further delineate the features of the AVM as well as to assist us with a thorough discussion of the various treatment options. The angiogram revealed the presence of multiple large venous varices draining into the deep venous system and outflow obstruction at the vein of Galen, which likely represented the cause of the varices. In addition, there were several flow-related aneurysms on the left distal and middle cerebral arteries, left distal anterior cerebral artery, and the left

posterior cerebral artery. After discussing the natural history of the lesion, the various treatment options, and the aggressive anatomical features of the AVM, the patient ultimately elected to undergo stereotactic radiosurgery for treatment of the AVM.

DISCUSSION

Unruptured AVMs cause obstructive hydrocephalus only rarely, and the reported cases of this phenomenon represent AVMs that were either dural-based or had intraventricular extension by way of location (that is, posterior fossa lesions).^{2,7–10} We presented a case in which a large venous varix produced obstructive hydrocephalus in a patient with an unruptured AVM, and to our knowledge, this phenomenon has not been previously reported in the literature. Outside the setting of an AVM, venous varices have been noted to cause obstructive hydrocephalus.¹⁰

One can perhaps explain why obstructive hydrocephalus occurs with such rarity in AVMs from an embryological basis. It is generally accepted that AVMs have

FIG. 2. Anteroposterior (a) and lateral (b) cerebral angiograms showing the dimensions of the varix in addition to its draining pattern. Flow-related aneurysms are also apparent (b), as is the size and extent of the varix near the cerebral aqueduct.

feeding arteries originating from the pial surface and developing in a conical configuration with the apex residing in the deepest portion of the parenchyma. The draining veins commonly lead to dural sinuses or sylvian veins, and because these venous outlets lie peripherally with respect to the CSF pathway, they fail to encroach on this pathway with any real frequency. Thus, resultant obstructive hydrocephalus is rare.

In addition, even in circumstances in which a draining varix or vein is located in proximity to the CSF pathway, the varix must grow to a sufficient size to encroach on this pathway. Outside of recognizing that varices are associated with high-flow lesions, little has been reported on what induces varix formation and growth. Arteriovenous malformations that develop varices tend to have a paucity of draining veins, and in some instances the drainage of a high-flow AVM can be limited to one draining vein, which through persistent hemodynamic challenge can dilate and develop into a varix. In the patient in the present case, the high flow of this lesion, as evidenced by the flow-related aneurysms that developed, was also coupled with areas of venous outflow obstruction as manifested by areas of stenosis within the varix. Thus, the venous outflow obstruction resulted in venous hypertension and subsequent progressive enlargement of the varix. Furthermore, the varix also enlarged unabated because the quadrigeminal cistern lacked any dural structure that could exert a buttressing force and limit its expansion. Factoring in these hemodynamic issues with histopathological data demonstrating that varices are devoid of a medial layer and that they possess only the intimal and adventitial layers of the vessel wall, we hypothesized that a venopathy developed given

the stresses that these attenuated venous walls accrued over time.

What remains unclear, however, is the exact pathophysiology behind how this patient presented clinically. We propose two possible mechanisms. First, the patient could have suffered progressive worsening of the hydrocephalus (as evidenced by the progressive headaches preceding the drowning event) due to snorkeling and breath-holding, activities he was performing just before the near-drowning event. These activities could have led to decreased venous outflow (by way of increased thoracic pressure) and increased CO_2 , resulting in raised intracranial pressure leading to a loss of consciousness and near drowning. As such, the chronic hydrocephalus and associated AVM could have been diagnosed, in a sense, incidentally through the accident. Second, he could simply have experienced a seizure while snorkeling, prompting the near-drowning event. Resuscitation efforts could have led to increased venous and intracranial pressure expanding the varix and leading to the acute hydrocephalus seen on his admission. We know that the patient had no history of seizure, and there was no evidence of seizure as assessed by electroencephalography during his initial hospital evaluation. Moreover, he has not had any seizures during the follow-up period and has never taken antiepilepsy medications.

Several comments with regard to treatment of the hydrocephalus are warranted. We elected to treat it with a ventriculoperitoneal shunt primarily because surgical intervention and definite resection of the AVM was not feasible given the patient's tenuous medical condition. If we had resected the AVM, the varix would no longer have obstructed the sylvian aqueduct, and perhaps a CSF diversion strategy would have been obviated. However, given

the chronic nature of the hydrocephalus and the fact that the patient's ventricles did not decrease in size despite a 2-week trial of external drainage, we believed that shunting was the best treatment strategy in this case. Note, however, that particular caution should be exercised when treating posterior fossa AVMs together with obstructive hydrocephalus. Esparza and colleagues⁵ reported on such a lesion involving the torcular herophili in a patient who had obstructive hydrocephalus and was treated with a shunting procedure. Postoperatively, the patient unexpectedly became stuporous and Parinaud syndrome developed. The authors believed that upward herniation and distortion of the mesencephalon was the most likely cause for these complications, and the patient's symptoms were in fact completely relieved after ligation of the shunt. Finally, hydrocephalus caused by aqueductal stenosis can also be treated with interventriculostomy (aqueductal cannulation) and third ventriculostomy, and these treatment options merit consideration.^{1,3,4}

References

1. Avman N, Dincer C: Venous malformation of the aqueduct of Sylvius treated by interventriculostomy: 15 years follow-up. **Acta Neurochir (Wien)** **52**:219–224, 1980
2. DeFeo DR, Kusske JA, Rush JL, Pribram H: Aqueductal occlusion by midline arteriovenous malformation. **Surg Neurol** **5**: 59–62, 1976
3. Drake JM: Ventriculostomy for treatment of hydrocephalus. **Neurosurg Clin N Am** **4**:657–666, 1993
4. Elvidge AR: Treatment of obstructive lesions of the aqueduct of Sylvius and the fourth ventricle by interventriculostomy. **J Neurosurg** **24**:11–23, 1966
5. Esparza J, Lobato RD, Munoz MJ, Chillon D, Portillo JM, Lamas E: Giant cerebral arteriovenous malformation producing a noncommunicating hydrocephalus. **Surg Neurol** **15**:76–80, 1981
6. McCormick WF: The pathology of vascular ("arteriovenous") malformations. **J Neurosurg** **24**:807–816, 1966
7. McKissock W, Paterson JH: A clinical survey of intracranial angiomas with special reference to their mode of progression and surgical treatment: a report of 110 cases. **Brain** **79**: 233–266, 1956
8. Paine KW, McKissock W: Aqueduct stenosis; clinical aspects, and results of treatment by ventriculocisternostomy (Torkildsen's operation). **J Neurosurg** **12**:127–145, 1955
9. Pribil S, Boone SC, Waley R: Obstructive hydrocephalus at the anterior third ventricle caused by dilated veins from an arteriovenous malformation. **Surg Neurol** **20**:487–492, 1983
10. U HS, Kerber C: Ventricular obstruction secondary to vascular malformations. **Neurosurgery** **12**:572–575, 1983

Manuscript submitted January 26, 2007.

Accepted March 8, 2007.

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