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Case Report

A case of hydrocephalus confounded by suprasellar arachnoid cyst and concomitant reversible cerebral vasoconstriction syndrome

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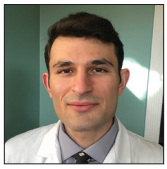

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ABSTRACT

Background: Obstructive hydrocephalus is a neurologic condition that has varied clinical and imaging presentations, as well as a multitude of congenital etiologies including aqueductal stenosis and less commonly arachnoid cysts. Aqueductal stenosis is a physical limitation to cerebrospinal fluid flow along the course of the aqueduct, which results in enlargement of the third and lateral ventricles. Arachnoid cysts are thin walled and fluid filled central nervous system lesions that can result in mass effect on adjacent structures. While arachnoid cysts are mostly asymptomatic, they may present with neurological symptoms that vary depending on the location of the lesion. Suprasellar cysts in particular may cause obstructive hydrocephalus as well as endocrine dysfunction. Reversible cerebral vasoconstriction syndrome (RCVS) is an unusual condition caused by cerebral arterial vasoconstriction that often presents initially with a thunderclap headache. Frequently, there is some environmental trigger associated with this condition. RCVS more commonly affects women and can induce stroke.

Case Description: A 57-year-old female presented to the emergency department with progressive headache and visual changes. Initial workup suggested the patient's symptoms where related to RCVS but subsequent surgical management of what was presumed to be long standing, compensated hydrocephalus resulted in resolution of the patient's symptoms.

Conclusion: We report, to the best of our knowledge, the first case of aquedutal stenosis and suprasellar arachnoid cyst with concomitant RCVS. The presence of multiple pathologies found on radiologic imaging illustrates the challenges presented by incidental findings and subsequent anchoring bias in medical diagnosis.

Keywords: Aqueductal stenosis, Endoscopic third ventriculostomy, Reversible cerebral vasoconstriction syndrome, Suprasellar arachnoid cyst

INTRODUCTION

Obstructive hydrocephalus, also known as non-communicating hydrocephalus, is caused by the impedance of cerebrospinal fluid (CSF) flow within the ventricles. Patients with obstructive hydrocephalus may suffer from headache, nausea, vision abnormalities, and altered gait. Obstructive hydrocephalus most commonly results following infection and hemorrhage, but there exist a multitude of congenital etiologies including aqueductal stenosis and arachnoid cysts. Aqueductal stenosis most frequently affects the proximal portion of the aqueduct, but also can

occur distally, with a web or membrane of tissue obstructing flow into the fourth ventricle.[6] Stenosis may arise either congenitally or due to postinflammatory aqueductal gliosis. Patients may present at various stages of life, with up to 10% of adult hydrocephalus cases arising from aqueductal stenosis. Primary congenital aqueductal stenosis is mostly idiopathic but has been found to be X-linked, autosomal recessive, or autosomal dominant in a minority of cases.[18] Infection with toxoplasmosis, Epstein-Barr virus, and cytomegalovirus has also been linked to congenital stenosis. Researchers have postulated that adult onset primary aqueductal stenosis asymptomatically arises in childhood following birth trauma or perinatal infection. These patients are thought to develop decompensated hydrocephalus later in life. Headache is one of the most frequently reported symptoms in aqueductal stenosis. Approximately 80% of adults improve following endoscopic third ventriculostomy (ETV), though symptoms and hydrocephalus may recur.

Primary arachnoid cysts are thin-walled congenital lesions derived from the arachnoid mater. These cysts are comprised of meningothelial cells that secrete fluid similar to CSF. However, these cysts do not in fact communicate with either the subarachnoid space or ventricles.[10,13] Secondary arachnoid cysts are the result of trauma, intracranial hemorrhage, and other intracranial insults. Arachnoid cysts make up approximately 1% of intracranial masses. Most patients are asymptomatic, but those who are symptomatic often present during childhood. Symptomatic patients may suffer from headache, focal neurologic deficits, and seizures.[7] The size of these cysts and their location determines the extent of mass effect and its associated symptoms. Approximately 9% of arachnoid cysts are suprasellar cysts, which originate from an abnormality in the membrane of Liliequist or the interpeduncular cistern and progressively expand from the prepontine space.[8,12] Consequently, the floor of the third ventricle is displaced. This can occlude the ventricles, distort, and obstruct the aqueduct, and thus result in obstructive hydrocephalus. Nearly 90% of patients with suprasellar cysts suffer from obstructive hydrocephalus and are symptomatic. Due to compression of the optic chiasm, patients may also suffer from poor visual acuity or visual field deficits, as was the case in more than half of patients according to one study.[15] Furthermore, compression of the pituitary infundibulum may result in endocrine abnormalities in children, including delayed menarche as well as reduced production of growth hormone.[11] Patients with symptomatic suprasellar arachnoid cysts may require surgical interventions such as ETV, in which a neurosurgeon fenestrates the floor of the third ventricle to allow CSF egress to the prepontine cisterns.

During the work-up of patients with nonspecific neurologic presentations such as headache and visual changes, confusion as to the underlying causative etiology can be encountered when diagnostic testing yields findings that have overlapping symptomatology. Careful diagnostic decision-making with awareness of the pitfalls of anchoring bias is necessary to ultimately achieve the correct diagnosis and treatment. In the workup for headache, finding evidence of subarachnoid hemorrhage (SAH) results in well-established differential pathways that require the exclusion of specific dangerous etiologies such as aneurysm and vascular malformations. Once excluded from the study, less common etiologies such as reversible cerebral vasoconstriction syndrome (RCVS) can be considered.[19] RCVS is notable for the self-limiting segmental vasoconstriction and dilatation of the cerebral vasculature, which generally resolves within 12 weeks.[3] Serious complications, however, may occur and include ischemic and hemorrhagic stroke.[4,5]

Herein, we present, to the best of our knowledge, the first reported case of aqueductal stenosis confounded by a suprasellar arachnoid cyst and concomitant RCVS.

CASE PRESENTATION

A 57-year-old female with a medical history of hypertension, hyperlipidemia, depression, and anxiety presented to the emergency department (ED) with progressive frontal headache 2–3 weeks in duration. Her headache was associated with transient blurry vision. The patient denied nausea, photophobia, neck pain, or altered gait. Head computed tomography (CT) showed enlargement of the lateral and third ventricles, empty sella turcica without transependymal resorption of CSF, or acute intracranial hemorrhage. Findings were consistent with chronic hydrocephalus [Figure 1].

Magnetic resonance imaging (MRI) of the brain demonstrated chronic cortical SAH (cSAH)/superficial siderosis of the parasagittal frontoparietal sulci and left anterior frontal sulci [Figure 2]. Prominent flow-related signal voids adjacent to a distal anterior cerebral artery branch was detected on T2-weighted imaging, requiring CT angiogram for further evaluation to rule out an aneurysm or vascular malformation. Bilateral multifocal stenoses of multiple intracranial arterial circulations were subsequently found on CT angiogram. Cerebral angiography confirmed these results [Figures 3a-c]. Vasculitis, vasospasm, and atherosclerosis as well as RCVS were listed as possible causes of the patient's cerebrovascular findings.

An MRI of the brain with CSF flow study was subsequently performed, revealing partial obstruction of the cerebral aqueduct [Figure 4]. CSF obstruction was found at the junction of the cerebral aqueduct and the superior aspect of the fourth ventricle.

ETV with the right frontal brain biopsy was performed. Intraoperative endoscopy located a large cyst within the third ventricle obscuring the ventricular floor [Figure 5]. The cyst

was cauterized and fenestrated. Ventricle and brain biopsy results found normal leptomeninges and cortical gray matter. The patient endorsed significant improvement in her headaches and vision and was neurologically intact following surgery.

CSF studies were negative for malignancy and bacterial, viral, or fungal infection. Her CSF total protein level was found to be low, with a concentration of <10 mg per dL. Her CSF was negative for white blood cells (WBCs), but contained 600 red blood cells (RBCs) per μΛ. She was also found to be antinuclear antibody negative. Postoperative head CT demonstrated a significant decrease in hydrocephalus [Figure 1b].

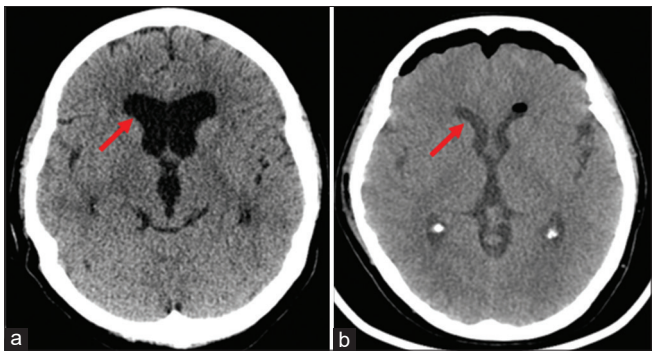

Figure 1: (a) Preoperative axial computed tomography (CT) demonstrating ventriculomegaly. (b) Postoperative Axial CT demonstrating decrease in ventricle size following endoscopic third ventriculostomy. Pneumocephalus is evident on scan.

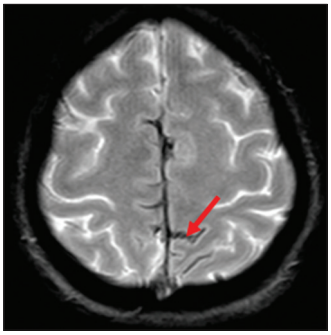

Figure 2: Axial T2 sequence magnetic resonance imaging (MRI) illustrating cortical subarachnoid hemorrhage (SAH)/siderosis from chronic SAH.

DISCUSSION

The subject of this case study was found to have three concomitant conditions, aqueductal stenosis, suprasellar arachnoid cyst, and RCVS following diagnostic workup and subsequent surgical management with ETV. The initial workup of this patient on ED presentation resulted in a brain MRI demonstrating cortical SAH and a CTA showing multifocal intracranial arterial stenoses. These findings were initially assumed to explain the patient's clinical syndrome and a diagnosis of RCVS was presumed as other etiologies of cSAH and multifocal arterial stenosis were excluded from the study. The finding of compensated hydrocephalus as well as mass effect within the suprasellar cistern was given less weight in the diagnostic decision making, ultimately resulting in a delayed diagnosis and treatment. In this way, the findings of cSAH and vascular stenoses were used to anchor the diagnostic workup and thinking about this patient's condition until other evidence dislodged these initial assumptions of causality.

RCVS is thought to originate from abnormally elevated cerebral arterial tone due to excessive sympathetic output and neuronal stimulus.[3] Medium and small distal intracranial arteries are thought to be affected earlier on in the course of the disease, with vasoconstriction moving proximally to affect the larger more central arteries. Severe headaches may result from stimulation of the trigeminal afferents located inside of the leptomeninges. Patients may suffer from moderate headaches between severe episodes.

Reperfusion injury of the leptomeningeal arteries may result in cSAHs, which have been found in 22% of RCVS cases.[1,3] CT is not always able to visualize cSAH, with only

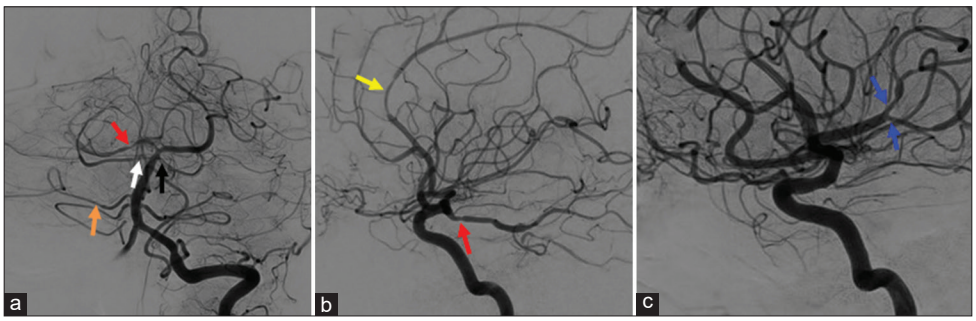

Figure 3: Arrows point to multifocal stenosis of medium to small caliper cerebral arteries demonstrated on angiography. (a) Red arrow: R. posterior cerebral artery; White arrow: R. superior cerebellar artery; Orange arrow: R. anterior inferior cerebellar artery; Black arrow: L. superior cerebellar. (b) Yellow arrow: R. anterior cerebral artery; Red arrow: R. posterior cerebral artery. (c) Blue arrows: L. middle cerebral artery.

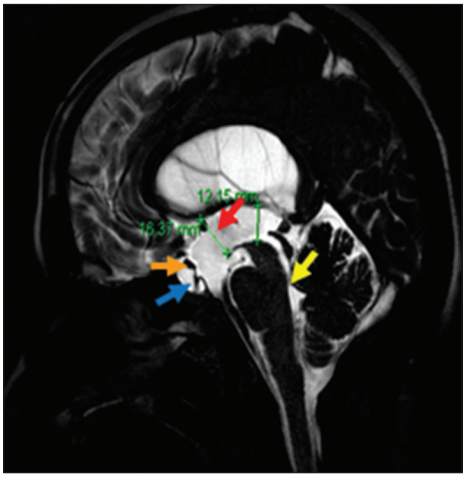

Figure 4: MRI/cerebrospinal fluid: cerebrospinal fluid flow study. Red arrow: arachnoid cyst membrane; Orange arrow: splayed optic tracks; Blue arrow: flattened infundibular recess; Yellow arrow: stenotic cerebral aqueduct.

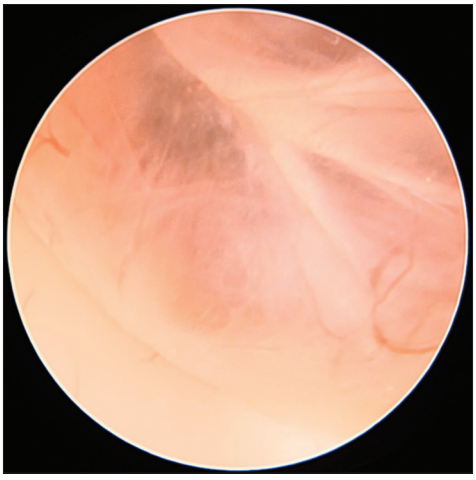

Figure 5: Intraventricular arachnoid cyst: Endoscopic third ventriculostomy screenshot.

52% of such bleeds detected by CT. MRI is a more sensitive modality for the detection of cSAH, which generally appears as iso to hypointense on T2-weighted imaging [Figure 2] and hyperintense on FLAIR. The cortical siderosis found on imaging results from hemosiderin deposition from chronic SAH. Evaluation of the etiology of the patients cSAH as well as concern over possible aneurysm seen on T2-weighted imaging led physicians to acquire a CT angiogram, which first demonstrated multifocal stenosis possibly due to RCVS. It is important to note that the flow-related loss of signal on MRI secondary to turbulent flow has been shown to mimic aneurysms.[9]

RCVS must be differentiated from primary angiitis of the central nervous system (PACNS), an idiopathic vasculitis limited to the medium-sized and small arteries of the central and peripheral nervous systems. PACNS causes headache, paresis, and other nonspecific symptoms, requiring glucocorticoids with or without cyclophosphamide as treatment.[2] The CSF of patients with RCVS is normal or near-normal with protein concentrations of <100 mg/dL and WBC counts of <15 WBCs/μΛ.[11] By comparison, PACNS patients' CSF WBC counts or total protein levels are elevated in more than 90% of cases.[4,17] Critical to the patient's diagnosis with RCVS was her CSF laboratory results, which included the absence of WBCs as well as a low total protein level.

The patient did not report having an extremely painful (thunderclap) episode of headache, a finding more typically associated with RCVS than PACNS. However, the patient's CSF did not exhibit pleocytosis, thereby bolstering the case for RCVS. In addition, a significant number of RBCs were found in the CSF, which is an expected finding in RCVS in the event of cSAH.[3] With a presumed diagnosis of RCVS as an anchor, the patient's hydrocephalus was initially thought to be long standing and compassed associated with prior exposure to SAH. It was not until additional questions about the etiology of the suprasellar mass effect and the relevance of aqueductal stenosis prompted a delayed repeat brain MRI with CSF flow study that the relevance of the patient hydrocephalus became apparent.

Arachnoid cysts are mostly asymptomatic lesions but may present with neurological symptoms such as headache, nausea, and vision abnormalities. Suprasellar cysts may cause obstructive hydrocephalus through their compression of the third ventricle.[7] Associated compression of optic chiasm may also induce visual impairment. However, the thin walls of the cyst make it difficult to visualize on imaging. In this case, the cyst wall was only visible on the heavily T2-weighted sagittal sequence of the CSF flow study [Figure 4]. This sequence demonstrated a thin-walled intraventricular cyst within the suprasellar cistern and its associated mass effect on the infundibular recess and optic tracks. In addition, the CSF flow study demonstrated evidence of diminished CSF flow at the level of cerebral aqueduct with focal stenosis at the junction of the aqueduct and fourth ventricle.

Together these findings presented anatomic lesions that both potentially obstructed the flow of CSF and could explain the patient's hydrocephalus. This hydrocephalus could result in symptoms of intermittent headache as well as visual changes especially in the setting of suprasellar mass effect. With these findings apparent on the repeat MRI and uncertainty as to the causative etiology of the patient's symptoms, the decision was made to perform an ETV as treatment for the patient's hydrocephalus. This procedure addressed both of the patient's anatomic lesions by creating an alternative pathway for CSF flow. Following the surgery, the patient's symptomatology resolved, and the patient was successfully discharged home.

Alternatively, a neurological surgeon may elect to insert a shunt to treat an arachnoid cyst. This treatment, however, may result in overdrainage of CSF caused by excessive shunting leading to intracranial hypotension and symptoms that include headache, nausea, dizziness, and seizures.[14] Rates of shunt overdrainage have varied widely from 1% to over 50% of shunted patients. Usage of low-pressure opening valves has been shown to predispose patients to overdrainage. However, programmable valves allow clinicians to increase the valve's opening pressure and subsequently reduce CSF drainage restoring a balanced flow.[16]

CONCLUSION

A 57-year-old female who presented with progressive, severe headache, and visual changes was found to have obstructive hydrocephalus, aqueductal stenosis, RCVS, and a suprasellar arachnoid cyst following diagnostic work-up and ETV. Both RCVS and hydrocephalus can produce overlapping clinical syndromes. Initial assumptions regarding the importance of cSAH and multifocal vasospasm as causative etiologies for the patient's symptoms biased the clinical decision-making away from focusing on hydrocephalus. This bias ultimately delayed the patient's accurate diagnosis and treatment until additional testing challenged these initial assumptions of causality. This case report illustrates the importance of cognitive biases such as anchoring especially when dealing with complex medical diagnosis in the setting of multiple overlapping clinical syndromes. Clinicians must be prepared to reject their initial assumptions and change diagnostic course when such biases are suspected.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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