

Neurological sequelae from brachiocephalic vein stenosis

Report of 2 cases

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Stenosis of central veins (brachiocephalic vein [BCV] and superior vena cava) occurs in 30% of hemodialysis patients, rarely producing intracranial pathology. The authors present the first cases of BCV stenosis causing perimesencephalic subarachnoid hemorrhage and myoclonic epilepsy.

In the first case, a 73-year-old man on hemodialysis presented with headache and blurry vision, and was admitted with presumed idiopathic intracranial hypertension after negative CT studies and confirmatory lumbar puncture. The patient mildly improved until hospital Day 3, when he experienced a seizure; emergency CT scans showed perimesencephalic subarachnoid hemorrhage. Cerebral angiography failed to find any vascular abnormality, but demonstrated venous congestion. A fistulogram found left BCV occlusion with jugular reflux. The occlusion could not be reopened percutaneously and required open fistula ligation. Postoperatively, symptoms resolved and the patient remained intact at 7-month follow-up.

In the second case, a 67-year-old woman on hemodialysis presented with right arm weakness and myoclonic jerks. Admission MRI revealed subcortical edema and a possible dural arteriovenous fistula. Cerebral angiography showed venous engorgement, but no vascular malformation. A fistulogram found left BCV stenosis with jugular reflux, which was immediately reversed with angioplasty and stent placement. Postprocedure the patient was seizure free, and her strength improved. Seven months later the patient presented in myoclonic status epilepticus, and a fistulogram revealed stent occlusion. Angioplasty successfully reopened the stent and she returned to baseline; she was seizure free at 4-month follow-up.

Central venous stenosis is common with hemodialysis, but rarely presents with neurological findings. Prompt recognition and endovascular intervention can restore normal venous drainage and resolve symptoms. (<http://thejns.org/doi/abs/10.3171/2013.1.JNS121529>)

KEY WORDS • central venous stenosis • jugular venous reflux • idiopathic perimesencephalic subarachnoid hemorrhage • myoclonic epilepsy • vascular disorders

NEUROLOGICAL sequelae from abnormal cerebral venous drainage are predominantly caused by intracranial venous thrombosis or a vascular malformation. Competency of the IJV valve prevents an alteration in the extracranial venous system from affecting the cerebral circulation.^{4,14} Anatomical studies have found that chronic venous hypertension will make the IJV valve dysfunctional.^{4,15} When the IJV valve is incompetent, the resulting jugular venous reflux can lead to intracranial pathological conditions.^{2,3,5,6,12,20}

Venous hypertension is a significant problem in patients with chronic hemodialysis. Approximately 30% of

patients receiving hemodialysis will develop stenosis of the central veins (BCV or SVC) attributable to proximal venous catheterization.^{1,10,18} Of this subset, half will develop symptomatic venous hypertension from venous engorgement, most commonly presenting as ipsilateral chest and upper-extremity edema.¹

We present 2 cases of BCV stenosis producing intracranial venous hypertension and different neurological findings. These are the first reports of central venous stenosis associated with idiopathic PSAH and recurrent myoclonic epilepsy.

Case Reports

Case 1

History and Examination. This 73-year-old hyperten-

Abbreviations used in this paper: AVF = arteriovenous fistula; BCV = brachiocephalic vein; IJV = internal jugular vein; PSAH = perimesencephalic subarachnoid hemorrhage; SVC = superior vena cava.

Neurological sequelae from brachiocephalic vein stenosis

sive African-American man on hemodialysis, who had a history of a pacemaker placement and idiopathic intracranial hypertension, presented with 2 days of disorientation, perseveration, headache, and blurry vision. On admission, a CT/CT angiography study of the head showed no acute findings or vascular abnormalities, and evaluation by an ophthalmologist was remarkable for papilledema. A lumbar puncture recorded an opening pressure of 35 cm H₂O with 15 ml of CSF drained off. The patient's mental status mildly improved following the lumbar puncture and he was started on acetazolamide. His symptoms were attributed to idiopathic intracranial hypertension and side effects of topiramate started the day prior to admission for headaches.

Operation. On hospital Day 3, the patient complained of a severe headache prior to having a witnessed grand mal seizure. Emergency head CT revealed SAH in the basilar cisterns (Fig. 1). Cerebral angiography failed to find an aneurysm or vascular malformation, but demonstrated extremely slow cerebral circulation time with marked cortical venous dilation. A fistulogram of the left upper-extremity AVF showed left BCV occlusion with left jugular venous reflux (Fig. 2). An attempt to recanalize the left BCV was unsuccessful and the patient was taken to the operating room for emergency fistula ligation.

Postoperative Course. Postoperatively, all of his symptoms quickly improved. Repeat cerebral angiogram on postoperative Day 1 showed normal arterial-venous transit time, resolution of cortical venous distention, and antegrade jugular flow. The patient returned to his baseline neurological status within 2 days, was discharged home, and remained neurologically intact at 6-month follow-up.

Case 2

History and Examination. This 67-year-old hypertensive African-American woman on hemodialysis presented with 2 weeks of disorientation, progressively worsening right arm weakness, and "jerking" movements. In the emergency room, a CT head scan demonstrated left frontal lobe edema and an MRI study of the brain confirmed bilateral (left > right) subcortical white matter edema (Fig. 3). Findings on an MR angiography study were of concern for early venous visualization and possible dural AVF. The neurological evaluation showed that her seizures were consistent with myoclonic epilepsy, and levetiracetam was started.

Operation. Cerebral angiography demonstrated delayed drainage and venous engorgement, but did not show any underlying vascular malformations (Fig. 4). A fistulogram showed high-grade stenosis of the left BCV with left jugular venous reflux (Fig. 5). The left BCV was successfully treated with angioplasty and stent insertion, leading to immediate antegrade flow through the left IJV (Fig. 6).

Postoperative Course. Postprocedure the patient was seizure free, her right arm strength improved greatly, and



Fig. 1. Case 1. Axial head CT scan showing SAH in the basilar cisterns.

she was discharged home. Seven months after discharge, she developed problems with her hemodialysis access and was scheduled for a fistulogram the following day. The next morning, the patient was found by her family in status epilepticus, the first seizure since her original admission. An MRI study obtained on presentation noted new bilateral thalamic edema and improved bilateral subcortical white matter edema when compared with the prior admission. A fistulogram showed complete occlusion of the left BCV stent with retrograde flow in the left IJV. Angioplasty was again successful in reopening the left BCV, with immediate resolution of jugular venous reflux. The patient improved quickly to her neurological baseline and remained seizure free at 4-month follow-up.

Discussion

Central Venous Stenosis

Central venous stenosis is prevalent in 16%–50% of all patients with central venous catheters; higher rates are associated with multiple catheter placements, indwelling catheters, and pacemaker wires,^{1,10,18} and it is usually asymptomatic in patients not receiving hemodialysis. Other less common causes of central venous stenosis include radiation to the neck and chest, tumor compression (SVC syndrome), and deep venous thrombosis of the upper extremity. With hemodialysis, central venous stenosis is thought to be caused by high-pressure turbulent flow stimulating intimal hyperplasia. The combination of venous stenosis and increased blood flow with hemodialysis results in venous engorgement and produces clinical manifestations.²

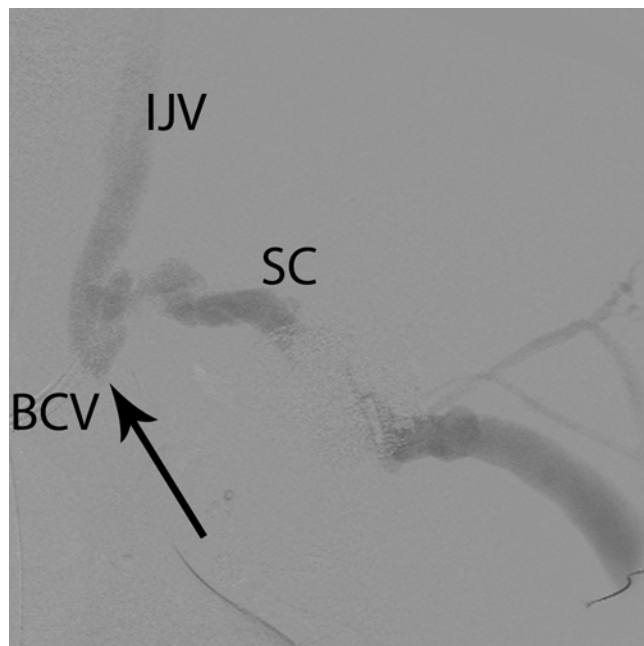


FIG. 2. Case 1. Left upper-extremity fistulogram showing BCV occlusion (arrow) with left IJV reflux. SC = subclavian vein.

Diagnosis of central venous stenosis can be elusive, and the senior authors (W.O.B. and R.A.A.) advise that a thorough evaluation must include a fistulogram (if present). The National Kidney Foundation guidelines recommend percutaneous transluminal angioplasty, with or without stent placement, as the preferred approach to central venous stenosis.¹⁸ If the stenosis proves to be refractory to endovascular methods, surgical options include revision or ligation of the AVF and bypass.

The IJV Valve and Functionality

The IJV valve was first described in the 16th century and long thought to have no physiological significance.¹⁵ However, growing evidence from more recent investigations demonstrates that these valves are indeed functional and serve to avert backflow into the IJVs.^{14,15,20} Competency of the IJV valve prevents alterations to the intracranial circulation, which may be why reports of neurological changes from jugular venous reflux are rare.

With long-standing venous hypertension, as with central venous stenosis, Fisher et al.⁴ found that the IJV valve became dysfunctional in 10 (67%) of 15 patients. This finding is corroborated by the fact that jugular venous reflux was present in our report and in all previous central venous case reports.^{3,5,6,12}

Venous Hypertension and PSAH

Since van Gijn et al.¹⁷ first described PSAH in 1985, the exact cause remains unresolved. Most authors suggest a venous source given its relatively benign clinical course and low complication rate when compared with aneurysmal SAH.

Putative risk factors predisposing patients to PSAH

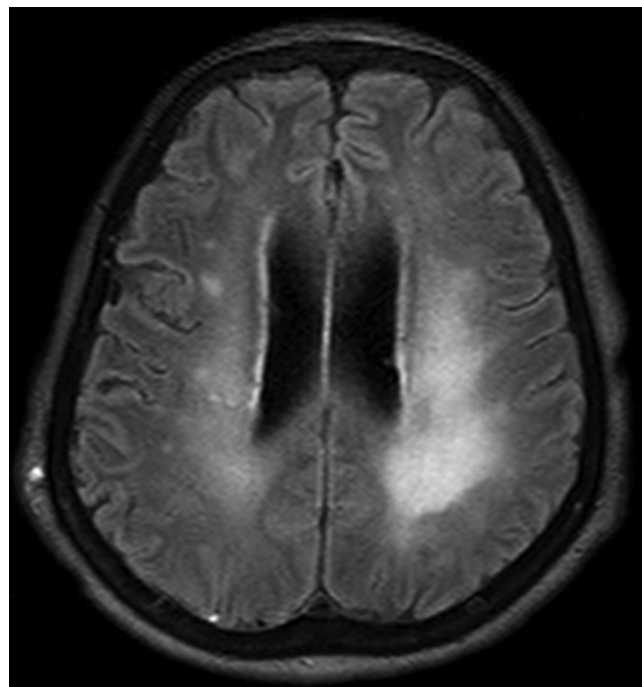


FIG. 3. Case 2. Bilateral subcortical white matter edema on FLAIR imaging.

are also a source of debate. One proposal is that increased venous pressure can lead to vessel engorgement and potentially rupture. This hypothesis is supported in the literature with reports on PSAH precipitated by straining and physical exertion^{11,21} as well as publications highlighting aberrant venous drainage patterns^{7,16,19,21} and underlying venous pathologies.^{8,9,13} Our first case also corroborates

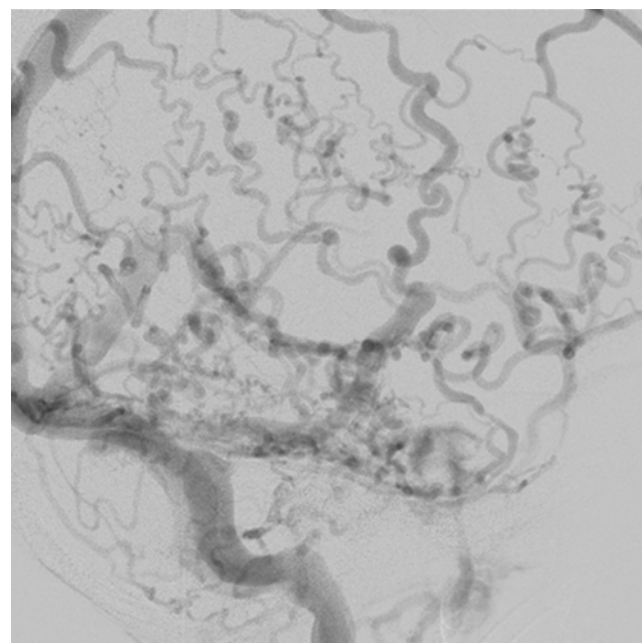


FIG. 4. Case 2. Cerebral angiogram, venous phase, showing delayed drainage and venous engorgement.

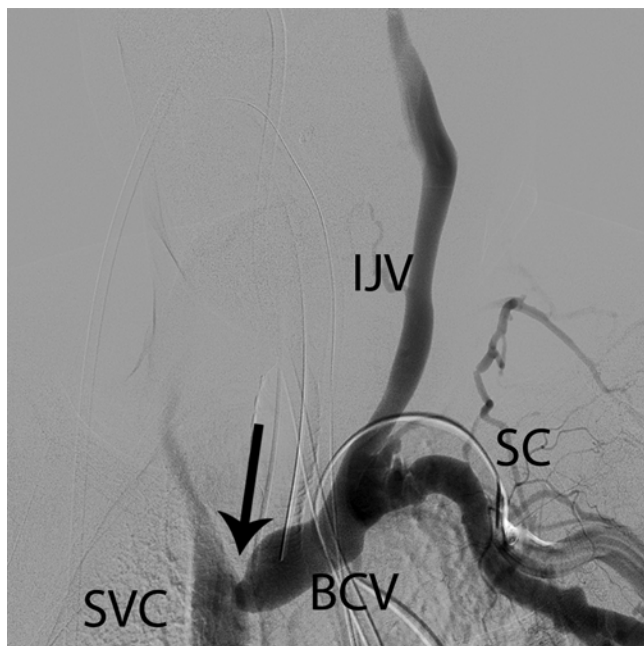


FIG. 5. Case 2. Left upper-extremity fistulogram demonstrating high-grade stenosis (arrow) of the BCV with left IJV reflux.

the idea that increased venous pressure can lead to PSAH, and to our knowledge this the first report on central venous hypertension being associated with PSAH.

Conclusions

Central venous stenosis is a common finding in patients receiving hemodialysis that rarely presents with

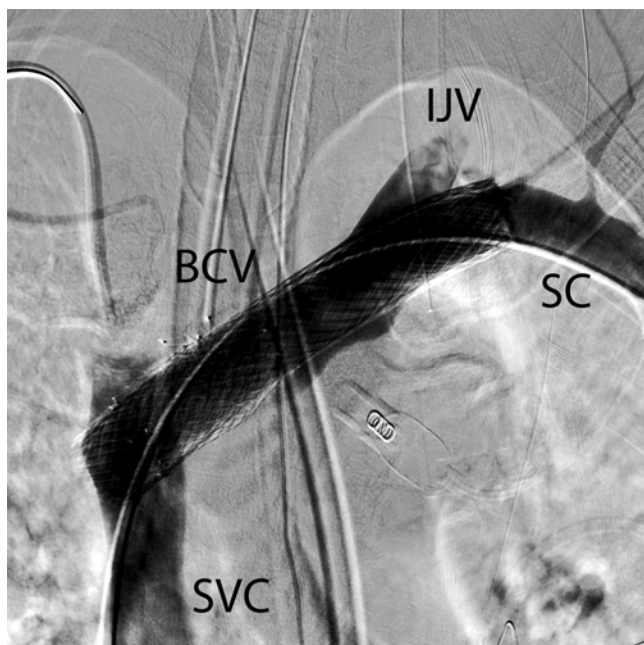


FIG. 6. Case 2. Angiogram obtained after angioplasty and stent placement, with antegrade flow through the left IJV.

neurological findings. We share 2 cases of venous hypertension from BCV stenosis transmitted to the intracranial circulation and resulting in different neurological sequelae. Prompt recognition and intervention can lead to restoration of normal venous drainage and resolution of symptoms.

Disclosure

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