Innominate vein stenosis causing raised intracranial pressure and blindness

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ABSTRACT

Central venous stenosis causing elevated intracranial pressure is a rare and potentially reversible cause of blindness in patients undergoing hemodialysis. We present the case of a 69-year-old man with progressive vision loss and raised intracranial pressure that was successfully treated with an innominate vein stent. (J Vasc Surg Cases and Innovative Techniques 2020;6:282-4.)

Keywords: Pseudotumor cerebri; Central venous stenosis; End-stage renal disease; Innominate vein stent; AV fistula

CASE REPORT

A 69-year-old African American man presented with a 2-month history of bilateral progressive vision loss. He had a past medical history of type 2 diabetes mellitus without retinopathy and end-stage renal disease. His vision loss impaired his ability to perform daily activities and to recognize facial features. Furthermore, he complained of a distinct "drumlike" noise in his right ear but denied headaches or other neurologic symptoms.

The patient had a right upper extremity brachial artery to axillary vein prosthetic hemodialysis access. At the time of presentation, the access was functioning normally. Six months earlier, a right subclavian vein angioplasty was performed for access maintenance. He had a history of previous internal jugular permcaths. He denied symptoms of central venous stenosis, including right arm or facial swelling.

The patient was referred to a neuro-ophthalmologist. His visual acuity examination demonstrated preserved central visual fields; the right eye had 20/40 vision, and the left eye had 20/100 vision. A dilated fundus examination demonstrated bilateral papilledema. A lumbar puncture was performed, confirming elevated intracranial pressure with an opening pressure of 50 cm of water. The examination findings of the cerebrospinal fluid were otherwise normal.

Intracranial imaging by magnetic resonance angiography and venography failed to demonstrate an arteriovenous (AV) fistula, venous sinus thrombosis, or other intracranial abnormalities. Computed tomography venography (Fig 1) of the neck and chest revealed a high-grade right innominate vein stenosis. The vascular surgery service obtained a shuntogram that confirmed a high-grade right innominate vein outflow stenosis (Fig 2) with minimal emptying into the superior vena cava and heart. The primary outflow for the AV graft was through an intracranial pathway. Flow was directed retrograde into the right jugular vein, continuing into the sigmoid sinus across the transverse sinus, communicating then through the cavernous sinus to the left transverse sinus and antegrade into the left internal jugular (Fig 3).

The right innominate vein was treated with a 14 by 60-mm self-expanding stent and postdilated with a 12-mm balloon (Fig 4). The stent restored a brisk flow through the innominate vein once again and while doing so reversed the retrograde flow into the cranial circulation. After intervention, the patient reported partial resolution of the blindness, and repeated lumbar examination demonstrated a return to a normal intracranial pressure. The patient's right AV graft was ligated to reduce the risk of recurrent intracranial hypertension secondary to in-stent stenosis of the innominate vein. Currently, a thigh graft is used as the hemodialysis access site. The patient is now 16 months post procedure. The patient's vision has not fully recovered, but he has not had any further deterioration and is able to live independently.

Consent was obtained from the patient for the publication of this case report.

DISCUSSION

Pseudotumor cerebri is a rare but known complication of central venous stenosis or occlusion in the setting of a functional ipsilateral AV access. It is important to recognize and to acknowledge the findings of progressive vision loss and headaches in dialysis access patients because it is a possible reversible cause of blindness. To date, there are only 13 reported cases of AV access-induced intracranial hypertension in the English literature.[1]

Central venous stenosis is a common complication seen in end-stage renal disease patients maintained on hemodialysis. Central venous stenosis is usually secondary to previous central venous catheters and the turbulent flow from the AV access. This predisposes

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Fig 1. Computed tomography venography of the neck and chest revealed a high-grade right innominate vein stenosis, emphasized by *arrows*.

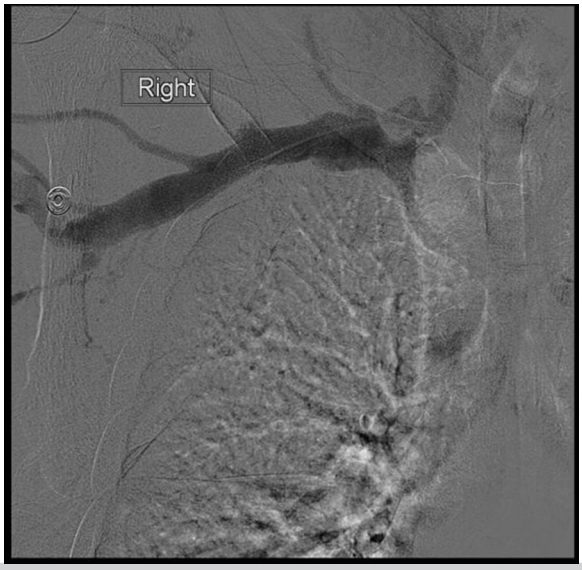

Fig 2. The shuntogram confirmed the findings of computed tomography venography.

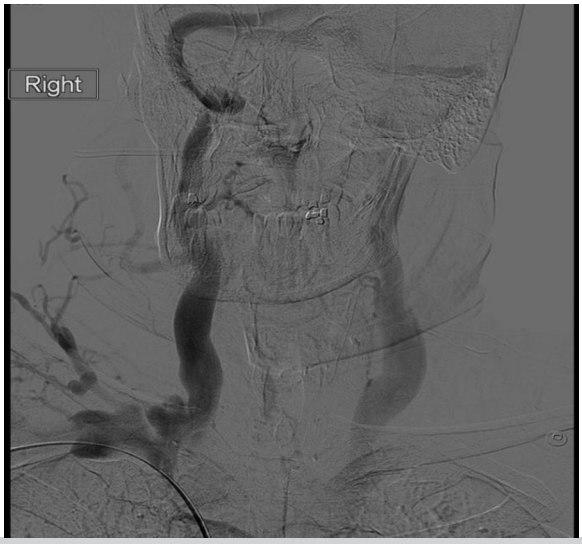

Fig 3. The shuntogram demonstrated that the primary outflow for the arteriovenous (AV) graft was through a retrograde intracranial pathway.

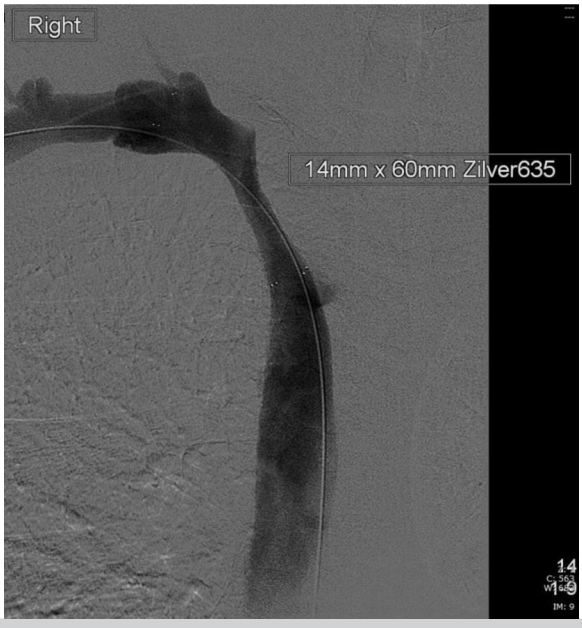

Fig 4. The right innominate vein was treated with Zilver stent, reversing the retrograde flow.

patients to endothelial damage to the vein wall and subsequent intimal hyperplasia and stenosis.[2] Most patients who develop central venous outflow obstruction present with ipsilateral arm swelling, high venous pressures on dialysis, chest wall collaterals, and prolonged bleeding from the needle puncture site after cannulation.

The shunting of blood from the venous outflow tract intracranially is the precipitating mechanism for development of pseudotumor cerebri in these patients. The intracranial venous circulation does not have venous valves, which allows pressurization of the entire intracranial venous network. The high venous pressure arising from the shunted blood impairs the reabsorption of cerebrospinal fluid across the arachnoid villi, leading to high cerebrospinal fluid pressure.[1] The anatomic requirements for this to occur are as follows. First, the location of the outflow stenosis or occlusion must affect the brachiocephalic vein.[1] The ipsilateral internal jugular vein must be patent. Finally, in patients with central venous stenosis, the majority of patients will develop a robust network of collaterals through the axillary and subclavian veins. Blood is routed to chest wall and neck collaterals and drains back into the central venous circulation through the azygos system. In the absence of these collaterals, the majority of blood from the hemodialysis access passes intracranially through the internal jugular vein.

With the high percentage of patients on hemodialysis who have initiated dialysis with a central venous catheter and one-third of hemodialysis patients having some form of central stenosis,[3] it is surprising that we do not see this complication more frequently. The rate of papilledema in patients with a functioning access is estimated to range from 0% to 8%, but it is rarely clinically manifested.[4]

Despite the immediate reduction in intracranial pressure, the patient did not completely resolve the underlying loss of visual acuity. It is likely that he experienced long-standing elevated intracranial pressure that damaged the optic nerve. Reported cases of reversible vision loss have often been associated with a short duration of symptoms or mild vision loss.[1,5] This emphasizes the importance of early recognition and treatment of this rare and potentially devastating problem.

REFERENCES

1. Mackay DD, Biousse V. Hemodialysis graft-induced intracranial hypertension. Neurol Clin Pract 2015;5:494-7.
2. Quencer KB, Oklu R. Hemodialysis access thrombosis. Cardiovasc Diagn Ther 2017;7:S299-308.
3. U.S. Renal Data System. USRDS 2012 annual data report: atlas of chronic kidney disease and end-stage renal disease in the United States. Bethesda, Md: National Institutes of Health, National Institute of Diabetes and Digestive and Kidney Diseases; 2012. Available at: http://www.usrds.org/adr.aspx. Accessed December 2017.
4. Taban M, Taban M, Lee MS, Smith SD, Heyka R, Kosmorsky GS. Prevalence of optic nerve edema in patients on peripheral hemodialysis. Ophthalmology 2007;114:1580-3.
5. Change S, Masaryk T, Lee M. Optic nerve edema: complication of peripheral hemodialysis shunt. Semin Ophthalmol 2004;19:88-90.

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