Stenting as a treatment for exercise-induced intracranial hypertension from bilateral jugular vein obstruction

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Practical Implications

Consider pathophysiology-based treatment by jugular stenting for intermittent exercise-induced intracranial hypertension due to jugular vein obstruction.

Secondary pseudotumor cerebri syndrome (SPCS) is defined as intracranial hypertension caused by factors other than an underlying infectious, malignant, or macroscopic structural brain lesion.¹ We report a case of exercise-induced SPCS due to decompensation of borderline venous return following bilateral jugular vein (JV) obstruction treated with jugular stenting based on pathophysiological reasoning.

Case report

An 18-year-old non-obese healthy man experienced moderate head trauma, a left brachial plexus avulsion, and asymptomatic left vertebral artery dissection following a motorcycle accident. He had a prolonged intensive care unit (ICU) stay with central venous monitoring.

Eight years after his accident, he started experiencing early morning pulsatile unilateral frontotemporal headaches associated with photophobia, visual blurring, tinnitus, and occasional vomiting. These episodes were precipitated by strenuous physical activity and became more frequent and prolonged in the subsequent 4 years. Neurologic examination including fundoscopy, laboratory tests, and noncontrast cerebral CT was unremarkable. Migraine without aura was diagnosed and treated appropriately, although there was no improvement. Some episodes lasted up to 6 days, so the diagnosis was revised to recurrent status migrainosus.²

A lumbar puncture (LP) performed when the patient was symptomatic revealed an opening pressure (OP) of >50 cm H₂O with normal CSF findings. Cerebral MRI showed distension of bilateral perioptic subarachnoid spaces with flattening of the posterior sclera of the left eye (figure 1A) and a partially empty sella (figure 1B), suggesting the diagnosis of idiopathic intracranial hypertension,³ as no underlying etiology was identified.

The patient noticed immediate relief after LP, with normalization of OP on follow-up. Oral acetazolamide did not prevent headache recurrences, and only frequent therapeutic LPs relieved symptoms. A typical headache could be triggered by exercise on a stationary bicycle under medical observation. A placebo effect of LP was ruled out by performing a sham LP, which was ineffective. During an asymptomatic phase, a test of CSF compartment compliance demonstrated normal pressure increase.

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Figure 1 Imaging

(A) T2-weighted fat-saturated image of orbits showing distension of bilateral perioptic subarachnoid spaces with flattening of the posterior sclera of the left eye (2005). (B) Sagittal T1-weighted image shows partially empty sella (2005). (C) Magnetic resonance venogram done during headache prior to lumbar puncture (LP) showing right transverse sinus stenosis (October 11, 2005). (D) Magnetic resonance venogram done 24 hours after therapeutic LP and headache relief, showing improved flow in right transverse sinus (November 11, 2005). (E) 3D-reconstructed cervical venous magnetic resonance angiogram (February 2007) with gadolinium-enhanced magnetic contrast (Vasovist, gadofosveset trisodium, Epix Pharmaceuticals, Lexington, MA) showing chronic subocclusion of the left internal jugular vein (JV) with stenosis of the right internal JV (red line indicating arteries and blue lines indicating veins). (F) Sagittal T1-weighted image obtained 5 years after stenting (2012) shows reexpansion of previously compressed pituitary gland.

Cerebral venous magnetic resonance angiography (MRA) during a severe headache episode revealed narrowing of the dominant right transverse sinus (figure 1C), with normalization after therapeutic LP (figure 1D). Cervical venous MRA showed stenosis of the right internal JV extending up to the confluence of the internal and external JV, chronic subocclusion of the left internal JV (figure 1E), and well-developed collaterals (figure e-1A at Neurology.org/cp). Venous digital subtraction angiography (DSA) between attacks showed apparent occlusion of the left JV, a normal-appearing right JV, and no pressure gradient along the right JV or the right transverse sinus on manometry. An extensive thrombophilia profile was negative.

During a prolonged headache episode, the patient developed aphasia, right faciobrachial weakness, and bradycardia. We documented compression of the transverse sinuses on venous DSA. A therapeutic LP reversed all deficits. After multidisciplinary discussion and with the informed consent of the patient, right JV stenting was proposed. A long segment stenosis of the right JV was confirmed during therapeutic DSA. After stenting, retromandibular vein collaterals immediately collapsed, improving venous flow in the right JV (figure e-1, B and C).

After 4 weeks, severe headaches recurred, leading to suicidal ideation. Venous CT angiography showed right JV stent thrombosis, despite simultaneous treatment with aspirin and vitamin K antagonists (VKAs). The thrombus initially resolved with IV heparin only to recur after switching to VKAs. He was then started on a triple therapy of aspirin, subcutaneous

Supplemental Data

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therapeutic nadroparin (discontinued after 4 months), and clopidogrel (discontinued after 2 years), preventing further thromboses (table e-1).

Currently, at age 43 and 7 years after the last in-stent thrombosis, the patient has occasional minor tension-type headache, usually triggered by physical exertion and promptly responding to paracetamol. He is now working part-time and has resumed his social and sporting activities.

DISCUSSION

Chronic obstruction of both JVs, posttraumatic on the left and possibly iatrogenic on the right, following JV catheter insertion (for ICU monitoring) caused intermittent exercise-induced SPCS. Treatment with CSF drainage was initially therapeutic, but a severe episode with transient focal neurologic symptoms and Cushing reflex prompted us to perform unilateral JV stenting, which, after initial in-stent thromboses, resolved symptoms.

Possible causes of SPCS include disturbances in intracranial (cerebral venous sinus thrombosis) or extracranial (JVs) venous outflow. A self-limiting venous collapse feedback loop, reversible with LP or sinus venous stenting, has been proposed.^{4,5} Traumatic JV thrombosis may cause intracranial hypertension and has been treated with endovascular stenting and balloon angioplasty.^{6,7} Although the reasons for the delay between the trauma and the onset of headaches remain unclear, the triggering of headaches by exercise could best be explained by pressure elevation in the vena cava, decompensating a preexisting borderline venous return. The ensuing SPCS may then be further aggravated by a vicious cycle including narrowing of intracranial transverse venous sinuses, as documented in our patient (figure e-2).

In accordance with this proposed pathophysiologic mechanism, JV stenting should be considered in selected cases.

REFERENCES

1. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology* 2013;81:1159–1165.
2. Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition (beta version). *Cephalalgia* 2013;33:629–808.
3. Ball AK, Clarke CE. Idiopathic intracranial hypertension. *Lancet Neurol* 2006;5:433–442.
4. De Simone R, Ranieri A, Montella S, Bilo L, Cautiero F. The role of dural sinus stenosis in idiopathic intracranial hypertension pathogenesis: the self-limiting venous collapse feedback-loop model. *Panminerva Med* 2014;56:201–209.
5. De Simone R, Ranieri A, Montella S, et al. Intracranial pressure in unresponsive chronic migraine. *J Neurol* 2014;261:1365–1373.
6. Duke BJ, Ryu RK, Brega KE, Coldwell DM. Traumatic bilateral jugular vein thrombosis: case report and review of the literature. *Neurosurgery* 1997;41:680–683.
7. Bishop FS, Finn MA, Samuelson M, Schmidt RH. Endovascular balloon angioplasty for treatment of posttraumatic venous sinus thrombosis: case report. *J Neurosurg* 2009;111:17–21.

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AUTHOR CONTRIBUTIONS

P. Michel: conception, data acquisition and analysis, critical revision of the manuscript. A. Angelillo-Scherrer: analysis, interpretation, critical revision of the manuscript. P. Maeder: data analysis, interpretation, critical revision of the manuscript. S. Binaghi: data acquisition, analysis, critical revision of the manuscript. I.A. Meyer: data analysis, critical revision of the manuscript. P. Correia: data acquisition, analysis, interpretation, critical revision of the manuscript.

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