CASE REPORT

Delayed relapse in pseudotumor cerebri due to new stenosis after transverse sinus stenting

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SUMMARY

A patient presented with recurrent severe pseudotumor cerebri (PTC). Transverse sinus stenting is a very effective treatment option, however stenosis and intracranial hypertension can recur. In our patient, stenting initially resulted in resolution of papilloedema. However, after 5 years, a new stenosis developed which required further stenting. This case highlights the fact that, in patients with PTC who undergo transverse sinus stenting, a small proportion require repeat treatment due to formation of a new stenosis, usually adjacent to the existing stent. Patients with severe disease, such as ours, may be at higher risk of recurrence. Regardless of the severity, all patients who undergo stenting should have regular ocular follow-up.

BACKGROUND

Pseudotumor cerebri (PTC) is a syndrome of intracranial hypertension without hydrocephalus or intracranial mass lesion, characterized by some or all of the following: headache, pulsatile tinnitus, transient visual obscurations, and papilloedema with progressive loss of visual fields leading eventually to blindness. It has also been called 'benign' or 'idiopathic' intracranial hypertension but, since it is not necessarily either benign or idiopathic, PTC—the original name—seems more appropriate.¹

PTC is often caused by or at least perpetuated by a stenosis with a pressure gradient in the dominant transverse sinus producing raised sagittal sinus pressure. Transverse sinus stenosis can be both a cause and an effect of intracranial hypertension through a positive feedback loop: high cerebrospinal fluid (CSF) pressure compresses and collapses the transverse sinus which then aggravates transverse sinus stenosis, which then further raises venous pressure and then further raises CSF pressure.²

Stenting of the transverse sinus stenosis is effective treatment for PTC² and, in the long term, cheaper than CSF shunting.³ Although there have so far been no direct comparisons in a clinical trial, transverse sinus stenting reduces intracranial pressure and resolves the papilloedema.^{2,4}

We present a patient with severe PTC syndrome and intracranial hypertension successfully managed with a single transverse sinus stent and included in our previous publication.² The patient then presented 5 years after the first stent, again with severe PTC syndrome due to a new stenosis just distal and external to the stent, and was again successfully treated by further stenting.

CASE PRESENTATION

An obese 38-year-old woman (body mass index >45 kg/m²) presented in November 2008 with recent severe headache, nausea, vomiting, pulsatile tinnitus, and transient visual obscurations with deteriorating vision. At the time she had severe papilloedema causing peripheral constriction of the visual fields, but her visual acuity was 6/9 and 6/12 and she had mild abducens nerve paresis bilaterally.

An ATECO (auto-triggered elliptic-centric-ordered three-dimensional gadolinium-enhanced MR venography) study showed a filling defect anteriorly within the dominant right transverse sinus. Cerebral venography demonstrated a filling defect typical of an arachnoid granulation causing a >75% stenosis of the right transverse sinus and a superior sagittal sinus pressure of 94 mm Hg with a 36 mm Hg gradient across the stenosis (figure 1A). A 9×40 mm Complete Iliac Self Expanding stent (Medtronic, Minneapolis, USA) was deployed after pretreatment with antiplatelet agents (figure 1B). Repeat pressures demonstrated immediate resolution of severe intracranial venous hypertension and resolution of the pressure gradient with pressures of 27 and 24 mm Hg above and below both stents. The papilloedema and visual loss improved, the headaches, pulsatile tinnitus and diplopia resolved, and her ophthalmologist discharged her for regular review.

Exactly 5 years after stenting, she again developed visual obscurations, diplopia, and pulsatile tinnitus. Her ophthalmologist again noted gross papilloedema and right perioptic nerve and retinal hemorrhages (figure 2).

INVESTIGATIONS

Repeat MRI demonstrated features of PTC syndrome (figure 3).

Cerebral venography and manometry showed a stenosis in the right transverse sinus, just above the proximal end of the stent (figure 4A). The stenosis was adjacent to and not within the stent.

The pressure in the superior sagittal sinus measured 91 mm Hg, 88 mm Hg at the torcula, 14 mm Hg in the right mid-transverse sinus below the stenosis with a gradient of 77 mm Hg—almost the same pressures as before the first stent was deployed (figure 4B).

DIFFERENTIAL DIAGNOSIS

Cerebral venous sinus thrombosis can present similarly. This was excluded by MRI.

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Figure 1 (A) Lateral projection of right transverse venous sinus venography demonstrating intrinsic stenosis (arrow) in the anterior segment of the sinus. (B) Lateral projection post-stenting demonstrating full expansion of the stent where stenosis was previously present.

TREATMENT

It was decided the most appropriate step at this point was to stent the new adjacent stenosis. After pretreatment with antiplatelet agents, an 8×40 mm Complete Iliac Self Expanding stent was placed across the stenosis ending in the previously placed stent. A venogram following stent placement showed complete stent expansion and good flow through the stent, with the sagittal sinus pressure now 22 mm Hg and the pressure gradient 4 mm Hg (figure 4C).

OUTCOME AND FOLLOW-UP

Again the papilloedema and visual abnormalities resolved and months after stent deployment she remains symptom-free.

DISCUSSION

In our series of 52 stented patients, six required repeat stenting due to recurrence of the papilloedema and transverse sinus stenosis (11.5%). The average sagittal sinus pressure and the pressure gradient across the stenosis were larger than in those patients who did not require restenting.²

Furthermore, a systematic review of transverse sinus stenting showed a pooled retreatment rate of 10.6% (22/207 patients),

Figure 2 Typical features suggestive of pseudotumor cerebri showing dilated optic nerve sheaths (arrows), flattened posterior margins of the globes, and raised optic discs.

17 of whom underwent repeat stenting of a new stenosis, often adjacent to the stent but also occurring in the contralateral transverse sinus.⁵ To our knowledge, in-stent stenosis is uncommon but it does occur and is often asymptomatic.⁶

This case emphasizes the need for indefinite follow-up in a patient stented for PTC. Despite initial success, new stenoses can develop even years later. Non-specific symptoms such as headache are unreliable measures of treatment response and, in some patients, headaches may still continue despite normalization of pressures² and may occur due to other headache syndromes such

Figure 3 Prior to treatment, bilateral funduscopy (right) shows loss of optic nerve margin and pale appearance indicating a swollen optic nerve head and associated with retinal hemorrhages.

Figure 4 (A) Lateral projection of right transverse venous sinus venography demonstrating stenosis (arrow) posterior to the previous stent. (B) Pressure tracing recorded on pullback of microcatheter through the venous sinuses. Note the large amplitude pressure waves pre-stenosis indicating low compliance (A) and post-stenosis with high compliance (B). The difference in pressures each side of the stenosis (ie, pressure gradient of 77 mm Hg) is considered to be severe. Lower frequency waves are intermixed and are related to the respiratory cycle. (C) Lateral projection after stenting demonstrating improvement in stenosis (arrow).

as migraine or tension. Since optic nerve injury and blindness can result from papilloedema in PTC,⁷ regular surveillance by an ophthalmologist or a neurologist who can perform an adequate ocular assessment (funduscopy and quantitative visual field testing) is suggested. If papilloedema recurs, patients will then need reinvestigation with CT or MR venography followed perhaps by catheter cerebral venography/manometry.

Learning points

- ▶ Transverse sinus stenting can be an effective treatment option in patients with moderate to severe pseudotumor cerebri.
- ▶ Despite being effective, stenosis recurrence can occur in a proportion of patients (11–12%).
- ▶ Stenosis recurrence usually manifests as adjacent to the existing stent and in-stent stenosis or restenosis is uncommon.
- ▶ Stented patients require ongoing ocular or radiological follow-up to detect stenosis recurrence.

Contributors HSW: primary authorship. GMH: managed clinical care of the patient from a neurological perspective and contributed to clinical/technical aspects of the publication. GP: performed stenting procedures for the patient and contributed to procedural/technical aspects of the publication. AM: compiled ophthalmological information including funduscopy images. TA: managed patients' ophthalmological care prior to and after admission.

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