CASE REPORT

Idiopathic intracranial hypertension and transverse sinus stenoses

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SUMMARY

An 18-year-old woman was diagnosed with idiopathic intracranial hypertension (IIH) and bilateral transverse sinus stenoses (TSS), after presenting with papilledema and decreased visual acuity. Lumbar puncture revealed an opening pressure of >60 cm H₂O. MRI showed bilateral TSS believed to be associated with the IIH. Initial treatment consisted of symptom relief by a temporary lumbar drain for cerebrospinal fluid (CSF) diversion, while the pros and cons of a more permanent solution by insertion of a ventriculoperitoneal shunt (VPS) or bilateral transverse sinus stent was discussed. A VPS was inserted since the patient had improved with CSF diversion. MRI verified reopening of the venous sinuses after shunt placement, and the patient remains asymptomatic with no signs of relapse after 3 years of follow-up.

BACKGROUND

Idiopathic intracranial hypertension (IIH) has a rather high and increasing incidence, with more than 1 in 100 000 persons affected.¹ The pathophysiology of IIH remains unclear; however, its association with bilateral transverse sinus stenoses (TSS) is reported in up to 90% of the IIH patients.² Opinions differ as to what should be regarded as the optimal treatment modality when invasive treatment is necessary, with both ventriculoperitoneal shunt (VPS) and endovascular stenting being evaluated.³ Considering the differences of these types of treatments, as well as our current lack of understanding of the basic pathophysiology of IIH, it is interesting that both treatment options have the potential for achieving similar results. Under any circumstance, it is important to be alert both with regard to suspecting the condition, and by actively treating it so that visual sequelae may be avoided. We present a case of successfully treated IIH with reversion of the concomitant TSS by VPS, and discuss the condition and its possible treatment options.

CASE PRESENTATION

An 18-year-old woman of East African descent with a body mass index (BMI) of 26; was admitted to the emergency department with a 4-week history of a slowly progressing occipital headache and neck pain. There was no history of trauma, and apart from a brief episode of fever 5 days earlier; no signs of infection were present. There was a slight nuchal rigidity, but the patient did not have any photophobia, focal neurological deficits, diplopia, blurred vision or visual obscurations.

INVESTIGATIONS

On admission, routine laboratory studies and a CT scan of the head were both normal. No CSF pleocytosis or biochemical deviations were present. With the patient assuming the right lateral decubitus position, a lumbar puncture was performed, and the patient stretched body and legs to avoid an artificially high pressure; opening pressure was >60 cm H₂O. Neuroophthalmological examination demonstrated bilateral Frisén grade 3 papilledema and *chorioretinal folds*. Corrected visual acuity was 0.5 left and 0.8 right. Perimetry showed central depression of the visual fields, and enlargement of the blind spots. Anterior slit lamp examination and extraocular motility were both normal. Consequently, IIH was suspected and a supplementary MRI, including venographic (MR venography) and the less flow artefact-sensitive T2 sequences, revealed bilateral stenoses of the transverse sinuses (figure 1).

DIFFERENTIAL DIAGNOSIS

Tension headache had been suspected by the patient's general practitioner, who had treated her with analgesics without success. Owing to the history of fever and nuchal rigidity, the patient was referred to the emergency department, where viral meningitis and neuroborreliosis were suspected. These considerations were disproved when normal blood and liquor analyses were obtained. The decreased visual acuity in this case was deemed to be caused by a combination of longstanding papilledema and spreading of oedema to the macular area due to IIH.

TREATMENT

Pharmacological therapy with acetazolamide was not considered as the primary treatment option in this case, since the patient had papilledema, decreased visual acuity, highly elevated lumbar cerebrospinal fluid (CSF) pressure and TSS. For immediate relief, a lumbar drain diverting 8–10 mL of CSF per hour was inserted. Meanwhile, endovascular stenting of the stenoses and/or permanent CSF diversion were discussed as treatment options. However, after 4 days of CSF diversion her headache, papilledema, *chorioretinal folding* and vision had improved. At this point, a new MRI (figure 2) confirmed resolved morphology of the venous sinuses, and a VPS with an adjustable pressure control valve (Strata II Regular, Medtronic Inc, Minneapolis) was inserted as a more permanent solution.

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Figure 1 MRI with venography at presentation. Axial (A) and sagittal (B) sequences, as well as coronal (C) T2 sequences showing bilateral stenoses of the transverse sinuses, as marked with arrows.

OUTCOME AND FOLLOW-UP

The remaining postoperative course was uneventful, and the patient was discharged a couple of days after surgery. After 3 years of follow-up, the patient continued to be asymptomatic, without papilledema, and with improved visual acuity 0.8 left and 1.0 right, respectively.

DISCUSSION

IIH is defined as raised intracranial pressure in the absence of hydrocephalus or mass, structural or vascular lesions and with normal CSF composition. Symptoms and signs may reflect those of generalised intracranial hypertension, and the pathophysiology remains unclear.^{3–4} Bilateral TSS is a common finding in patients with IIH, and it has been argued that TSS might be the actual cause of, or at least an exacerbating factor in, IIH by induction of increased venous pressure as well as secondary reduction of passive CSF absorption.⁵ Consequently, endovascular stenting of TSS has been described as a successful treatment of IIH.^{3–6–7} However, venous sinus pressures are reduced by CSF withdrawal, and resolution of TSS has been reported after CSF drainage suggesting that TSS in IIH is a phenomenon secondary to increased intracranial pressure rather than the cause of it.^{8–11} A study by Bono *et al* might, however, prove this hypothesis to be too simplistic. In patients treated pharmacologically with acetazolamide, the TSS persisted, even after normalisation of the CSF pressure.¹² This could suggest that depletion of intracranial CSF volume—rather than lowering of the intracranial pressure—is the cause of resolved TSS in IIH treated with CSF withdrawal.

There is a controversy whether stenting or shunting is the optimal treatment for TSS in IIH. In our opinion, it is

reasonable to initiate treatment with CSF diversion. Patients responding with resolved stenoses after CSF diversion can be considered suitable candidates for VPS insertion. Conversely, stenting might be appropriate for non-responders to CSF diversion. Additional research into the pathophysiology of IIH, as well as controlled clinical trials of the available treatment modalities are required to optimise treatment strategies in the future.

Learning points

- ▶ Headache and impaired visual acuity as a result of idiopathic intracranial hypertension (IIH) with transverse sinus stenoses can be reversed by cerebrospinal fluid (CSF) diversion.
- ▶ At present, both CSF diversion by ventriculoperitoneal shunt and endovascular stenting of the venous sinuses are feasible alternatives, but we suggest temporary CSF diversion as the first step when deciding between shunting and stenting.
- ▶ Treatment of IIH can be very challenging. In our opinion, this patient benefited from collaboration between neurologists, neurosurgeons, neuroradiologists and ophthalmologists. We recommend a multidisciplinary approach in similar cases.

Contributors AF and JB contributed to the conception and design. SS acquired the data, and drafted the article. MA, AF and JB performed analysis and interpretation of the data, and revised the manuscript for important intellectual content. All authors approved the final manuscript.

Figure 2 MRI with venography after 4 days of cerebrospinal fluid diversion. Axial (A) and sagittal (B) sequences, as well as coronal (C) T2 sequences showing reopened transverse sinuses, as marked with arrows.

Competing interests None.

Patient consent Obtained.

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