Acute surgical management in idiopathic intracranial hypertension

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SUMMARY

Idiopathic intracranial hypertension is a headache syndrome with progressive symptoms of raised intracranial pressure. Most commonly, it is a slow process where surveillance and medical management are the main treatment modalities. We describe herein an acute presentation with bilateral sixth nerve palsies, papilloedema and visual deterioration, where acute surgical intervention was a vision-saving operation.

BACKGROUND

Idiopathic intracranial hypertension (IIH) is a headache syndrome with signs and symptoms of raised intracranial pressure without causative findings on CT or MRI.^{1 2} Usually, symptoms are progressive, typically presenting with worsening headache and over-time evidence of positive neurological findings such as papilloedema and lateral rectus paresis. However, acute onset is very uncommon, as in our case. Because of this rarity, it is important to recognise the need for urgent surgical intervention where appropriate, without prior medical management, particularly in the setting of acute visual loss.

CASE PRESENTATION

A 34-year-old Caucasian woman presented to an ophthalmology clinic with acute onset headache, bilateral sixth nerve palsies and marked papilloedema. Her headache had worsened over the preceding 5 days and she described it as 'heaviness' in her head. Her vision had become blurred and her temporal fields were obscured. She had no nausea, meningism or migrainous symptoms. She reported no jaw claudication, recent weight loss, temporal pain or other associated signs of giant cell arteritis. She was a non-smoker and had a body mass index of 38. She had no other significant medical history or hereditary genetic conditions in her family. She was not taking regular medication, and had no recent foreign travel, long haul flight or trauma. On cranial nerve examination, there was evidence of bilateral sixth nerve palsies, double vision and decreased sensation in the first division of the trigeminal nerve bilaterally. Her visual acuity was 6/9 bilaterally, and there was obvious papilloedema on funduscopy. Her neurological examination was otherwise normal.

INVESTIGATIONS

Her vital signs were normal, blood investigations including full blood count, renal profile, coagulation factors, erythrocyte sedimentation rate, C reactive protein (CRP) and endocrine function test were within normal limits. A CT brain revealed normal brain parenchyma with no evidence of

ventricular enlargement. MRI and MR venography revealed no evidence of venous sinus thrombosis.

DIFFERENTIAL DIAGNOSIS

- ▶ Venous sinus thrombosis
- ▶ Subarachnoid haemorrhage
- ▶ Meningitis
- ▶ Ophthalmoplegic migraine (for review, see ref. 3)

TREATMENT

She was transferred to our department for emergency ventriculoperitoneal (VP) shunt insertion. A volumetric CT brain performed prior to surgery for the purposes of neuronavigation demonstrated increased tortuosity of the optic nerves with indentation of the globes in the region of the optic nerve heads (figure 1). These are radiological features of raised intracranial pressure.

OUTCOME AND FOLLOW-UP

Postoperatively, her headache gradually improved but her cranial nerve signs persisted. Fluorescein angiography showed no diffuse disc leak. A repeat MR venogram (figure 2) and MRI brain (figure 3) with contrast was performed, as about one-fifth of patients with venous sinus thrombosis may have intracranial hypertension only, without signs of cortical vein thrombosis.⁴ The repeat imaging was negative. On formal ophthalmological assessment, her visual acuity was 6/6 on the right and 6/9 on the left. She had persistent bilateral sixth cranial nerve palsies and papilloedema. Octopus visual field assessment showed constriction of both eye fields (figure 4). She was discharged home, 6 days later, with routine outpatient follow-up.

Figure 1 (A,B) Non-contrast axial view of CT brain demonstrates increased tortuosity of the optic nerves (full arrow) with indentation of the globes in the region of the optic nerve heads (dashed arrow).

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Figure 2 (A–D) MR venogram with contrast demonstrates normal flow within the major cerebral venous sinuses and deep cerebral venous system without evidence of thrombosis.

DISCUSSION

Sixth cranial nerve palsy causes binocular horizontal diplopia due to a paretic lateral rectus muscle, particularly in the direction of the involved muscle. The two most common aetiologies are vasculopathic and unknown, followed more rarely by trauma and metastatic disease.^{5 6} Over the age of 50, vasculopathic (diabetes and hypertension) aetiology is most common.

Bilateral dural arteriovenous malformation and ophthalmoplegic migraine have also been reported in literature.^{7 8} The aetiology is of prognostic value, with universal recovery of pressure palsies and rare improvement with tumour involvement.⁴ Sixth cranial nerve palsy has a well-described association with IIH and happens in about 20–33% of patients. It is frequently bilateral, especially in the paediatric population. This is thought to be attributable to traction of the sixth cranial nerve resulting from intracranial hypertension.^{9–12}

High-pressure headache, papilloedema and lateral rectus paresis are part of the modified Dandy criteria for diagnosis of IIH.¹³ However, benign intracranial hypertension-like syndrome is also the most frequent symptom in patients with cerebral vein thrombosis, present in about 80% of cases.¹⁴ This may confuse the clinician in differentiating these two separate diagnoses. In practice, the diagnosis of IIH is made when venous obstruction and other causes of raised intracranial pressure have been ruled out on conventional imaging.¹⁵ The clinician should also perform lumbar puncture to measure the opening pressure (>250 mm H₂O is significant), cerebrospinal fluid manometry and analysis.¹³

Our patient's main risk factor appears to be obesity,^{12 16 17} and this correlates to an incidence of approximately 1/100 000/year rising to 13/100 000/year in women between 20 and 44 years who are 10% above ideal body weight, and 19/100 000/year in those 20% above ideal body weight.¹ Increased

Figure 3 (A) T2-weighted brain MRI demonstrates no secondary signs of raised intracranial pressure with grossly normal globes and optic nerves (full arrow); (B) the right parietal ventricular catheter is in position and terminates in the right lateral ventricle (dashed arrow).

Figure 4 Octopus visual field assessment demonstrates constriction of both fields.

intra-abdominal and intrathoracic pressures are the proposed mechanism causing raised central venous pressures, but this does not explain the fact that although obesity is common, IIH is rare.

The ophthalmic hallmark is papilloedema which may be bilateral, asymmetrical or even unilateral.^{1 10 18} Of note, papilloedema may be absent in about 6% of patients with proven IIH.¹⁰ Patients usually complain of visual field changes with associated double vision. Central scotomas, inferior nasal defects and peripheral constriction are the next most common field defects.^{17 19} Optic nerve drusen or an anomalously raised disc, in the presence of headaches, can mimic papilloedema leading to a mistaken diagnosis of IIH. This is the reason our patient underwent fluorescein angiography.¹⁸

IIH is a well-described as chronic condition, with signs and symptoms that gradually worsen over time.²⁰ Cases such as asymptomatic papilloedema with progressive visual loss have been reported months to years after the initial episode of increased intracranial pressure, thus emphasising the lack of a direct relation between papilloedema, symptoms of headache, visual disturbance, lateral rectus paresis and increased intracranial pressure.^{1 15 18} Owing to this variability, repeat ophthalmological assessment should be performed to determine the progression of the condition.²¹ It is, therefore, very uncommon to have sudden onset of all symptoms together within less than 2 weeks.

In current practice, diagnosis is made by thorough history and clinical examination, followed by radiological investigations such as CT and MRI. MR venography is the procedure of choice for the diagnosis of dural venous sinus thrombosis in BIH. Pathognomonic features of IIH include undilated ventricles in the presence of intracranial hypertension.²² Initial management is always medical, using a combination of carbonic anhydrase inhibitors, furosemide, steroids and managed weight reduction. Steroid treatment is safe where investigations for venous sinus thrombosis have proved negative. Withdrawal of steroids may lead to a rebound increase in ICP and side effects

of their use can be problematic in IIH patients.^{17 23} Single diagnostic and therapeutic lumbar punctures have also been used in cases of benign self-limiting conditions, and repeated procedures are employed in the setting of rapidly declining vision to temporarily lower the CSF pressure while planning a more aggressive treatment.^{1 12 17} CSF diversion surgery, for example, optic nerve sheath fenestration, lumboperitoneal shunt and VP shunt is reserved in refractory cases.^{1 17 18 22}

We inserted a VP shunt acutely to stabilise vision.¹ The literature emphasises the need for early diagnosis, followed by surveillance or medical management. However, treatment should be tailored depending on the speed of progression of symptoms. We chose VP shunt over lumboperitoneal shunt owing to evidence of superiority of long-term patency rates, presumably because of the superior stability of the proximal end of the shunt system.²⁴

In summary, we present a middle-aged woman with acute onset of bilateral sixth nerve palsies and papilloedema. By exclusion, we conclude that our patient suffered from IIH and underwent emergency VP shunt insertion.

Learning points

- ▶ Idiopathic intracranial hypertension (IIH) is a headache syndrome with signs and symptoms of increased intracranial pressure without causative findings on CT or MRI.
- ▶ The treatment is tailored to the clinical signs and symptoms and usually patients proceed to either surveillance or medical management.
- ▶ It is crucial to treat patients urgently by means of a surgical approach in acute presentations suggestive of raised intracranial pressure such as worsening headache, positive cranial nerve findings, for example, sixth cranial nerve palsy (unilateral or bilateral) and papilloedema.

Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Acheson JF. Idiopathic intracranial hypertension and visual function. *Br Med Bull* 2006;79:233–44.
- 2 Lessell S. Pediatric pseudotumour cerebri (idiopathic intracranial hypertension). *Surv Ophthalmol* 1992;37:155–66.
- 3 Steele H, Aram J, Wilhelm T, et al. Not a microvascular sixth nerve palsy. *Pract Neurol* 2007;7:400–4.
- 4 Ferro JM, Canhao P, Bousser MG, et al. Prognosis of cerebral vein & dural sinus thrombosis: results of the International Study on Cerebral Vein & Dural Sinus Thrombosis (ISCVT). *Stroke* 2004;35:664–70.
- 5 Keane JR. Bilateral sixth nerve palsy, analysis of 125 cases. *Arch Neurol* 1976;33:681–3.
- 6 Bioussé V, Mendicino ME, Simon D, et al. The ophthalmology and intracranial vascular abnormalities. *Am J Ophthalmol* 1998;125:527–44.
- 7 Tan FU, Sener HO, Bilgic S, et al. Bilateral sixth-nerve palsy associated with dural arteriovenous malformation. *Acta Neurol Belg* 2001;101:110–12.
- 8 Verhagen WIM, Prick MJ J, Van Dijk Azn R. Onset of ophthalmoplegic migraine with abducens palsy at middle age? *Headache J Head Face Pain* 2001;43:798–800.
- 9 Rush JA. Pseudotumour tumour cerebri: clinical profile and visual outcome in 63 patients. *Mayo Clin Proc* 1980;55:541–6.
- 10 Radhakrishnan K, Ahlskog JE, Garitty JA, et al. Idiopathic intracranial hypertension. *Mayo Clin Proc* 1994;69:169–80.
- 11 Patton N, Beatty S, Lloyd IC. Bilateral sixth and fourth cranial nerve palsies in idiopathic intracranial hypertension. *J R Soc Med* 2000;93:80–1.
- 12 Pakosova E, Timarova G, Krivosik M, et al. Idiopathic intracranial hypertension: a retrospective clinical study. *Bratisl Lek Listy* 2011;112:691–4.
- 13 Digre KB, Corbett JJ. Idiopathic intracranial hypertension (pseudotumour cerebri): a reappraisal. *Neurologist* 2001;7:2–67.
- 14 Bousser MG, Barnett HJM. Cerebral venous thrombosis. In: Mohr JP, Choi DW, Grotta JC, Weir B, Wolf PA. *Stroke-pathophysiology diagnosis and management*. 4th edn, Philadelphia: Churchill Livingstone, 2004:301–25.
- 15 Bioussé V, Bousser MG. Benign intracranial hypertension. *Rev Neurol* 2001;157:21–34.
- 16 Walker RW. Idiopathic intracranial hypertension: any light on the mechanism of the raised pressure? *J Neurol Neurosurg Psychiatry* 2001;71:1–7.
- 17 Ambika S, Arjundas D, Noronha V, et al. Clinical profile, evaluation, management and visual outcome of idiopathic intracranial hypertension in a neuro-ophthalmology clinic of a tertiary referral ophthalmic center in India. *Ann Indian Acad Neurol* 2010;13:37–41.
- 18 Soler D, Cox T, Bullock P, et al. Diagnosis and management of benign intracranial hypertension. *Arch Dis Child* 1998;78:89–94.
- 19 Wall M, Hart WM, Burde RM. Visual field defects in idiopathic intracranial hypertension (pseudotumour cerebri). *Am J Ophthalmol* 1983;96:654–69.
- 20 Corbett JJ. Problems in the diagnosis and treatment of pseudotumour cerebri. *Am J Neurol Sci* 1983;10:221–9.
- 21 Rowe FJ. Assessment of visual function in idiopathic intracranial hypertension. *Br J Neurosurg* 2011;25:45–54.
- 22 Mauriello JA, Shaderowsky P, Gizzi M, et al. Management of visual loss after optic nerve sheath decompression in patients with pseudotumour cerebri. *Ophthalmology* 1995;102:441–5.
- 23 Liu GT, Glaser JS, Schatz NJ. High-dose methylprednisolone and acetazolamide for visual loss in pseudotumour cerebri. *Am J Ophthalmol* 1994;118:88–96.
- 24 Maher CO, Garrity JA, Meyer FB. Refractory idiopathic intracranial hypertension treated with stereotactically planned ventriculoperitoneal shunt placement. *Neurosurg Focus* 2001;10:1–4.

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