CASE REPORT

Guiding follow-up of paediatric idiopathic intracranial hypertension with optical coherence tomography

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SUMMARY

Idiopathic intracranial hypertension (IIH) is uncommon in the paediatric population. Papilloedema is the hallmark sign and patients can suffer permanent vision loss as a consequence. We describe the role of optical coherence tomography (OCT) in the follow-up of two paediatric patients with newly diagnosed IIH. Patient A presented with vomiting and examination showed ophthalmoplaegia and papilloedema. She was treated with acetazolamide, furosemide and therapeutic lumbar punctures. Patient B presented with incidental papilloedema and was treated with acetazolamide and she reported intermittent headache during follow-up. Fundoscopic examinations for both patients showed persistent blurred disc margins but OCT examinations documented improvement of average retinal nerve fibre layers. OCT may be of value in monitoring for recurrence in paediatric IIH.

BACKGROUND

Idiopathic intracranial hypertension (IIH) is an uncommon syndrome in the paediatric population. Patients may be asymptomatic or present with symptoms of raised intracranial pressure (ICP) or ophthalmic symptoms. Typical signs include papilloedema and esotropia from abducens nerve palsy.^{1 2} IIH is diagnosed using the modified Dandy criteria, which include: (1) signs and symptoms of ICP; (2) absence of localising neurological signs except abducens nerve palsy; (3) elevated cerebrospinal fluid (CSF) opening pressure of more than 25 cm H₂O with normal composition and (4) normal or small ventricles on brain imaging.^{1 2} It is more challenging to diagnose in the paediatric group compared to adults as there are age-dependent characteristics.^{1 2} Treatment is directed at relieving symptoms of raised ICP and to prevent permanent loss of vision. Close surveillance is required as the disease course can vary in duration and severity, and recurrence of IIH is possible. Objective assessments entail invasive procedures such as lumbar puncture (LP) to follow-up IIH and evaluate treatment response. Monitoring with clinical signs and subjective evaluation of papilloedema are imprecise. Objective non-invasive investigations would be helpful in the assessment and follow-up of paediatric patients with suspected IIH.

Optical coherence tomography (OCT) is a non-invasive imaging technique that produces high-resolution cross-sectional images of the retina. With OCT technology advancing from frequency and

spectral domains to the latest swept source techniques, the quality of the images allows distinct evaluation of the separate layers of the retina and precise measurements of each layer. Increase in perineural pressure is postulated to cause papilloedema via stasis of axoplasmic flow, and thus axonal swelling and thickening of the retinal nerve fibre layer (RNFL). Peripapillary RNFL therefore reflects the severity of the papilloedema.^{3–5} Commercially available machines can perform measurements of RNFL thickness, ganglion cell layer and total retinal thickness.^{3–5} Most commercial machines now also have a 'retest' function that ensures follow-up measurements are taken from the same area of the retina as the baseline examination, enabling us to compare measurements taken at different times.⁶

In this case series, we describe the use of OCT as an objective and non-invasive tool in the follow-up of two paediatric patients with IIH.

CASE PRESENTATION

Patient A

A 15-year-old Indian girl presented with 3 days of vomiting, and neck and back pain. She had a past medical history of chronic back pain. On admission, she was afebrile with normal clinical parameters and had a body mass index of 31.1 (>97th centile). Physical and neurological examinations were normal apart from painful limitation of neck movements. On the eighth day of admission, she reported double vision and was found to have an incomitant esotropia with mild limitation of abduction in both eyes. Visual acuity was 6/6 on the right and 6/9 on the left. Funduscopy also showed bilateral papilloedema; she was referred to neurology and ophthalmology for further investigation.

Patient B

A 9-year-old Chinese girl with no medical history was seen for myopia and was incidentally found to have asymmetric blurring of her optic disc margins (right more than left). Visual acuity, colour vision and Goldmann visual fields were normal. B-scan ultrasonography showed right optic nerve head drusen and initial impression was right pseudopapilloedema. She was reviewed 7 months later and fundoscopic examination showed increased blurring of the left disc margin. She did not report any symptoms of raised ICP and physical examination did not reveal any neurological deficit. OCT

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examination (using Spectralis, Heidelberg Engineering, Germany) showed increased average RNFL of 178 μm (right eye) and 147 μm (left eye); she was referred to neurology for further management.

INVESTIGATIONS

Patient A

Haematological investigations showed a normal full blood count consisting of haemoglobin of 12.9 g/dL, total white cell count of $6.91 \times 10^9/\text{L}$, platelet count of $290 \times 10^9/\text{L}$ and a normal renal panel with urea of 2.5 mmol/L, creatinine of 68 $\mu\text{mol/L}$, sodium of 138 mmol/L, potassium of 3.5 mmol/L, chloride of 105 mmol/L and bicarbonate of 20.2 mmol/L. X-ray and MRI of the cervical, thoracic and lumbar spine were normal. The patient underwent urgent MRI and MR venogram of the brain when she was found to have esotropia and papilloedema (Frisen grade 2) and both scans were normal. LP revealed an opening pressure of more than 55 cm H_2O (the upper limit of CSF manometer markings) and closing pressure was 47.5 cm H_2O . Microscopic examination of the CSF was normal with protein of 0.26 g/L (normal range 0.10–0.40 g/L) and glucose of 2.5 mmol/L, without pleocytosis. Bacterial and virology studies were also negative.

Patient B

Full blood count was normal with haemoglobin of 13.1 g/dL, total white cell count of $5.98 \times 10^9/\text{L}$ and platelet count of $351 \times 10^9/\text{L}$. Erythrocyte sedimentation rate and C reactive protein were also within normal limits. MRI of the brain was normal and LP showed an elevated opening pressure of 27 cm H_2O with normal CSF composition and no oligoclonal band seen.

TREATMENT

Patient A

The patient was started on oral acetazolamide at 250 mg two times a day (7.5 mg/kg/day). The headache and cranial nerve palsies persisted and she underwent a second therapeutic LP 5 days later. However, there was no improvement of her papilloedema and this was followed by lumbar drain insertion on day 24 of admission, which was retained for 5 days, draining 800 mL of CSF before it had to be removed. The patient underwent two further therapeutic LPs with the last puncture recording an opening pressure of 37 cm H_2O and closing pressure of 24 cm H_2O . Her symptoms resolved and examination also showed improvement of ophthalmoplaegia. Papilloedema, however, was still present (figure 1). Confrontation and

Humphrey visual fields remained normal and her visual acuity was 6/6 bilaterally. She was discharged on acetazolamide treatment and started on a weight management programme.

Patient B

This patient was started on acetazolamide 250 mg two times a day (11.4 mg/kg/day) and did not require further therapeutic LP.

OUTCOME AND FOLLOW-UP

Patient A

The patient reported of worsening headache and recurrence of diplopia 6 weeks later. Funduscopy showed persistent bilateral papilloedema. Recurrence of IIH was diagnosed and a further LP was performed with an opening pressure of 45 cm H_2O . Acetazolamide was increased to 500 mg four times a day (15 mg/kg/day) and furosemide 20 mg once daily (0.3 mg/kg/day) was added. She was discharged on acetazolamide and furosemide treatment.

The patient was closely monitored with monthly ophthalmology and neurology follow-up. She was interviewed for symptoms of raised ICP, orthoptic assessment for esotropia and optic disc examination at each visit. Serial OCT examinations (using Cirrus HD-OCT, ZEISS, Germany) were introduced to monitor her progress quantitatively. She did not report any headache, vomiting, diplopia or vision loss. Visual acuity was stable at 6/6 bilaterally. Optic disc examination continued to show disc swelling, which appeared to be improving, and this was confirmed by the subsequent OCT examinations, which documented a gradual improvement of the papilloedema except on the first measurement (at 10 weeks from presentation), likely due to inaccurate segmentation of retinal layers as a result of severe papilloedema (table 1). On follow-up 6 months after diagnosis, the average RNFL thickness recorded was 87 μm (right eye) and 85 μm (left eye), and resolution of papilloedema was seen on fundus examination (figure 2). The patient remained well on follow-up and there was no recurrence of symptoms or papilloedema.

Patient B

During outpatient follow-ups, the patient reported intermittent non-specific headache and funduscopy examinations showed persistent blurred disc margins bilaterally. OCT examinations showed gradual improvement of the average RNFL with treatment (table 2). She did not require further invasive investigation and her medication was subsequently reduced to 125 mg ON (2.8 mg/kg/day) due to gastrointestinal adverse effects.

Figure 1 Serial OCT measurements paired with coloured photographs of the optic disc (A) and cross-section of circle scan of the retina (B) at 10 weeks, 12 weeks and at 24 weeks, showing a general improving trend of OCT measurements but persistence of papilloedema in patient A. OCT, optical coherence tomography.

Table 1 Serial average retinal nerve fibre layer thickness measurements of patient A, using Cirrus HD-OCT

	Average retinal nerve fibre layer thickness	
Time from presentation (weeks)	Right eye (μm)	Left eye (μm)
10	122	161
12	135	138
14	120	116
20	91	93
24	87	85
36	75	73
72	66	65

DISCUSSION

OCT is of value to paediatric neurologists in the non-invasive follow-up monitoring of IIH in children. To our knowledge, this is the first report of such use of this modality in the BMJ case reports literature. The overall annual incidence of IIH is 0.9 per 100 000 individuals, based on adult studies.² These patients can present with symptoms secondary to raised ICP (such as headache) or visual problems (such as blurred vision or diplopia).^{1 2 7-11} Infants may present with non-specific symptoms such as irritability and somnolence.^{2 7} However, children with IIH can be asymptomatic and present only with papilloedema on ophthalmic examination, which is the hallmark for IIH.^{2 8 10} IIH without papilloedema has also been reported but is very rare.^{2 3 8 9} Ophthalmoplaegia is often due to abducens

Table 2 Serial average retinal nerve fibre layer thickness measurements of patient B, using Spectralis

Time from presentation (weeks)	Average retinal nerve fibre layer thickness	
	Right eye (µm)	Left eye (µm)
0	147	178
4	150	173
11	130	129
23	129	128
35	132	131

nerve palsy, but other patterns of cranial nerve palsies or ophthalmoplaegia have been described.^{2 9} The remainder of the neurological examination findings should be normal. Loss of visual acuity is often a late finding. Permanent decreases in visual acuity and visual field defects have been reported in up to 10% and 17% of children with IIH, respectively,^{2 8 11} and can occur at any point in the disease: at presentation, during treatment or even following recurrences.⁸

The aim of treatment is to relieve symptoms and preserve the vision by normalising the ICP.^{2 10} LPs are diagnostic and therapeutic. Acetazolamide, loop diuretics and topiramate are adjunct treatments, and corticosteroids may be considered in patients with rapid vision loss.^{2 8 10 11} Surgical interventions are reserved for patients who have failed medical therapy and for those who presented with rapid or severe vision loss, and are directed at decreasing ICP via a shunt procedure or to reduce the degree of papilloedema by optic nerve sheath fenestration, respectively.^{2 8 10 11} Definitive evidence on the best treatment modality is lacking, especially in the paediatric age group.^{7 8}

Clinical evaluation of IIH during outpatient paediatric neurology follow-up is often subjective. A recurrence rate of up to 22% has been reported, so close surveillance of affected children is vital in detecting recurrence.⁸ Regular assessment for symptoms and signs of raised ICP and visual loss is carried out during follow-up visits.² Direct ophthalmoscopic examination of the fundus is performed to document the progress of the papilloedema. This technique has certain limitations as it requires the patient's cooperation, and it is a subjective assessment based on the training and experience of the physician.^{3 8} Repeated LPs are the only way to objectively determine the intracranial pressure, but are invasive and emotionally traumatic for children. A non-invasive adjunct such as OCT would therefore be valuable in the evaluation and follow-up of paediatric patients. In a study of using OCT to follow-up adult patients with IIH, OCT examination documented a gradual decrease in the RNFL after treatment. This result coincided with resolution of papilloedema and impaired visual field defects.³ We postulate that earlier introduction of OCT for patient A after her diagnosis of IIH may have assisted monitoring of her treatment response and therefore earlier detection of the recurrence of her IIH.

There are limitations to using OCT including: (1) the reduction of RNFL thickness can be a result of optic atrophy and not necessarily due to a decrease in oedema, although optic atrophy would be an obvious clinical finding, (2) the reduction in papilloedema and hence OCT measurement always lags behind the decrease in ICP, (3) the retina is too thick for light to penetrate in severe papilloedema and the readings are thus inaccurate and (4) patients need to be cooperative and sit up and fixate for the

scan to be carried out.^{3 4 11} Recently, hand-held OCT has been shown to provide reproducible results but requires patients to be sedated for the test.⁶ It must be borne in mind that Humphrey Visual Field testing requires a certain level of understanding in children and is not always easily performed by younger children. Clinical diagnosis is the gold standard and OCT is an important adjunctive tool in the follow-up of IIH.

Learning points

- ▶ Paediatric patients with idiopathic intracranial hypertension require close monitoring for detection of recurrence of disease.
- ▶ Detection of recurrence can be challenging as clinical evaluation (eg, funduscopy) is subjective, and objective assessment (eg, lumbar puncture) is invasive and emotionally traumatic.
- ▶ Measurement of the retinal nerve fibre layer using optical coherence tomography coincides with severity of papilloedema, which can monitor effect of treatment and detect recurrence of disease.
- ▶ Clinical diagnosis is the gold standard in diagnosing recurrence of paediatric idiopathic intracranial hypertension, and optical coherence tomography can serve as an important adjunctive tool in detection of the disease by providing non-invasive and objective assessments.

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