CASE REPORT

Paediatric idiopathic intracranial hypertension

José Nuno Galveia, Ana Paula Mendonça, João Marques Costa

Department of Ophthalmology,
Hospital Egas Moniz,
Lisbon, Portugal

Correspondence to
Dr José Nuno Galveia,
jgalveia@gmail.com

SUMMARY

Idiopathic intracranial hypertension (IIH) can occur in paediatric age with clinical characteristics that may differ from adult presentation. The authors present a case of an 11-year-old boy, presenting with severe holocranial headaches for the past 4 weeks. Best-corrected visual acuities (BCVA) were 20/200 bilaterally and the fundus examination showed marked bilateral optical disc and macular oedema. CT scan with contrast as well as MRI showed no space occupying lesions, normal permeability of the dural venous sinuses and a partially empty sella. Lumbar puncture revealed an opening pressure of 540 mm Hg, with clear cerebrospinal fluid, with normal biochemistry and cytology. The patient was treated medically and subsequently submitted to a ventriculoperitoneal shunting procedure. 3 months after surgery the symptoms got completely resolved and his BCVA were 20/20 bilaterally.

BACKGROUND

Idiopathic intracranial hypertension (IIH) was characterised by papilloedema and elevated intracranial pressure (ICP) in the presence of normal neuroimaging and cerebrospinal fluid (CSF) studies.^{1–3}

The incidence of IIH in the general population is estimated to be 1 of 100 000. In the paediatric population there is a greater incidence of IIH in adolescents (12–15 years) as compared to children (2–12 years).³

CASE PRESENTATION

An 11-year-old boy presented with headache (severe, holocranial) for the past 4 weeks, accompanied by tinnitus and marked decrease in best-corrected visual acuity (BCVA). No relevant medical history was present. No medications were being taken regularly.

At presentation his BCVA were found to be 20/200 bilaterally. The patient correctly identified 1 out of 13 Ishihara plates for the right eye (RE) and none for the left eye (LE).

Slit lamp examination of both the eyes revealed no anterior segment abnormalities. Fundus examination showed a bilateral disc and macular oedema (figure 1).

Both optical discs were oedematous with multiple cotton-wool spots in a radiate disposition as well as flame-shaped haemorrhages. Marked venous and capillary tortuosity and distension were also present.

A careful examination of the macular area showed a radiate disposition of the hard exudates along the Henle's layer forming a subtle macular star figure, predominantly on the nasal side (figure 1).

Neurological examination revealed no other abnormalities besides the ones described.

At presentation the weight was 44 kg, the height 125 cm and the body mass index was 28.21, placing it above the 95th percentile for his age group. The patient had gained 5 kg in the last 6 months. Blood pressure was 110/60 mmHg. He had already reached puberty and could be classified as having achieved stage III in Tanner classification.

INVESTIGATIONS

Static computerised perimetry (SCP) (26/10/2011) (figure 2):

- ▶ Bilateral enlargement of the blind spot, nasal depression and decreased central sensitivity, predominantly in the left eye.

Optical coherence tomography (figure 3): macular oedema with subretinal fluid, and central foveal thickness of 523 µm for the right eye and 554 µm for the left eye.

CT with contrast (25/10/2011): no space occupying lesions, adequate development of brain structures; normal permeability of the ventricular system; intraocular protrusion of the optical nerve; partial empty sella; normal filling of the dural venous sinuses (figure 4).

MRI (18/11/2011): no space occupying lesions, normal permeability of the ventricular system; no ectasia of the optical nerve sheath; partial empty sella (figure 5).

Lumbar puncture: opening pressure of 540 mm H₂O, CSF, with normal protein and glucose values, leucocytes <3 cells/µl and negative microbiology.

Laboratory workup:

- ▶ General—normal cell blood count, negative CRP
- ▶ Endocrinology—normal seric values of luteinising hormone, parathyroid hormone, thyroid stimulating hormone, unbound thyroxine, insulin-like growth factor 1, adrenocorticotrophic hormone, and cortisol
- ▶ Immunology—negative antinuclear body test; tests for *Bartonella*, toxoplasmosis and treponemal antibodies were negative

DIFFERENTIAL DIAGNOSIS

- ▶ Idiopathic intracranial hypertension
- ▶ Cerebral venous sinus thrombosis
- ▶ Neuroretinitis

Idiopathic hypertension is a diagnosis of exclusion. The modified Dandy criteria³ must be met in order to make the diagnosis. These include: (1) general signs and symptoms of elevated ICP or papilloedema, (2) elevated cerebrospinal fluid pressure

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Figure 1 Bilateral optical disc oedema with hard exudates, as well as bilateral macular oedema with incipient macular star formation.

(>250 mm H₂O), measured in lateral decubitus position, with normal composition, (3) no evidence of hydrocephalus, mass, structural or vascular lesion on MRI or contrast-enhanced CT for typical patients, and MRI and MR venography for all others and (4) no other identified cause of intracranial hypertension. These criteria are only defined clearly for the adult.

In children, despite a great number of limitations (restlessness, need for sedation, greater range of reported normal values), the upper limit of normal values for CSF opening pressure are defined as 180 mm H₂O in ages up to 8 years of age and 250 mm H₂O afterwards.³ The patient fulfils all the inclusion criteria.

Figure 2 Static computerised perimetry at presentation.

Figure 3 Optic coherence tomography at presentation.

Figure 4 CT scan after contrast injection showing complete perfusion of the lateral cerebral venous sinuses as well as an intraocular protrusion of the optical nerve head (papilloedema).

The patient's weight is above the 95th percentile for the age group and a significant recent weight gain was also mentioned. Epidemiologically in adults idiopathic central hypogonadism is more common in middle-aged, obese women.^{3–5} In paediatric ages, however, the association with weight and sex is proven only for postpubescent children, such as our patient.³

Severe depression in visual acuity is usually not a part of the clinical presentation of paediatric IIH. Visual acuity in this group of patients

usually ranges between 20/30 to 20/60.² The patient had 20/200 bilaterally due to marked papilloedema with macular extension.

Cerebral venous thrombosis presents with a similar clinical picture and must be pondered in this setting. The diagnosis is made by neuroimaging. The current gold standard is the combination of MRI to visualise the thrombosed vessel and magnetic resonance venography to detect the non-visualisation of the same vessel.⁶ The neuroimaging examination performed allowed us to exclude this diagnosis.

Neuroretinitis is characterised by optical disc swelling and macular oedema with macular star formation. Several infectious agents, most commonly *Bartonella henselae*, *Treponema pallidum* and *Toxoplasma spp* can cause it. We excluded this diagnosis based on the absence of prodromic systemic symptoms as well as a negative serological testing.

TREATMENT

Given the magnitude of the ICP elevation, as well as the signs of optical nerve dysfunction, surgical treatment was deemed mandatory.

Before surgery, the patient was treated with acetazolamide 1 g/day, divided in two doses. Weight loss was recommended.

Medical treatment led to a symptomatic reduction in the headaches as well as the BCVA (after 1 month 20/50 for the right eye and 20/60 for the left eye and the patient identified 3 out of 13 Ishihara plates for the RE 1 for the LE).

The patient was referred to neurosurgery and was submitted to a ventriculoperitoneal shunting procedure, 1 month after the initial observation.

OUTCOME AND FOLLOW-UP

Two weeks after the surgery the headaches had disappeared, the papilloedema was improving and the BCVA were 20/20 for the right eye and 20/25 for the left eye.

Three months after the surgery the BCVA were 20/20 bilaterally. In this evaluation the patient identified correctly 13 out

Figure 5 Sagittal T1 fluid attenuated inversion recovery at presentation, showing partial empty sella (superior arachnoidocele).

Figure 6 Optical disc 3 months after surgery, showing secondary optical atrophy and visible Patton line (vertical line, temporal to the optical disc, more readily identifiable in the left eye, reflecting the limit of previous peri-papillary retinal elevation).

of 13 Ishihara plates bilaterally. The papilloedema had completely resolved. Fundoscopy revealed a diffuse pallor of the optical disc, predominantly on the temporal portion (figure 6) and a complete resolution of the macular oedema. Static computerised perimetry (figure 7) showed a persistent central relative scotoma and nasal depression, sparing the fixation.

After 6 months the patient remains symptom free and with a stable BCVA. He changed his diet and has lost 5 kg since the initial observation.

DISCUSSION

IIH is a rare clinical entity in the general population and even less prevalent in the paediatric population. Some aspects of the epidemiology, diagnosis and management are still under investigation.

In the paediatric population, clinical presentation can vary according to the onset of puberty.

In prepubertal children IIH is rare, has no sex predilection and no association with obesity.^{2 3 7} It commonly presents with stiff neck, strabismus and papilloedema.²

In adolescents (aged 12–18 years) the incidence of IIH is higher and related to obesity and female sex³ assuming a clinical presentation that more closely resembles the one in adults. The most common symptoms in this age group include headache, transient visual obscurations, photopsia, retrobulbar pain and diplopia.

Balcer *et al*⁸ reported a prevalence of obesity of 43% in patients with IIH aged 3–11 years versus 81% of those in the age group between 12 and 14 years and 91% of those in the age group between 15 and 17 years.

Childhood obesity is becoming a major public health issue. In the UK in 2008, 31% of boys and 29% of girls aged 2–15 years were classified as being overweight.⁹ In the USA during the years 2007 and 2008, 31.7% of children and adolescents aged

2–19 years were overweight.¹⁰ Given the association between IIH in postpubertal adolescents and obesity, we are likely to witness a rise in the prevalence of this pathology.

Our patient was 11 years old, had already entered puberty and was overweight.

IIH is a diagnosis of exclusion. Our patient presented with holocranial headache for the past 4 weeks, accompanied by tinnitus and decreased visual acuity. Fundus examination revealed bilateral optical disc and macular oedema, with macular star formation. MR imaging showed a partially empty sella, no evidence of hydrocephalus, mass, structural or vascular lesion. Empty sella was postulated to result from the longstanding effects of pulsatile CSF under high pressure, leading to the herniation of an arachnoidocoele through a defect in the *diafragma sella*.¹¹ It has a sensitivity of 70% and a specificity of 95% in IIH.¹¹

Lumbar puncture revealed a marked elevation of the opening pressure (250 mm of water) but normal glucose, protein and cytology, as well as negative microbiology. Laboratory workup excluded endocrinological or infectious causes. No medication was being taken regularly.

Concerning treatment, the symptoms responded to medical therapy with acetazolamide. However, given the magnitude of intraocular pressure elevation as well as the marked visual field loss we, in conjunction with neurosurgery opted for surgical intervention in the form of a ventriculoperitoneal shunt.

This case represents a particularly florid clinical picture. Usually presentation is milder with a less marked visual acuity and visual field loss and subtler fundus findings. The severity of the clinical picture may relate to the 4 weeks of waiting before searching medical consultation or may simply be idiosyncratic. In IIH, early diagnosis and treatment are critical. If intervention is prompt most children with mild disc oedema and visual field defects experience a complete resolution of both. Macular oedema, when present,

Figure 7 Static computerised perimetry 3 months after surgery, showing improved sensitivity, with normal central sensitivity and persistent nasal depression.

usually responds to therapy although most often a few months later than the previous changes. Permanent loss of visual acuity and visual field defects are reported to occur in 0–10% and 17% of patients, respectively.³ Three months after surgery the patient had a visual acuity of 20/20 bilaterally a complete recovery from the disc and macular oedema. However, paracentral visual field depression and peripheral constriction (most marked nasally) still persisted.

Learning points

- ▶ Idiopathic intracranial hypertension (IIH) is a diagnosis of exclusion.
- ▶ IIH in the paediatric population assumes different clinical characteristics in prepubertal children and postpubertal adolescents.
- ▶ In postpubertal adolescents IIH favours the female sex and is related with obesity.
- ▶ A common associated neuroradiological sign is the presence of an empty *sella turcica*.
- ▶ Early diagnosis and treatment are paramount to a good outcome

Competing interests None.

Patient consent Obtained.

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