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Case Reports

Idiopathic Intracranial Hypertension in Children: Case Report and Estimation of Local Incidence

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Abstract Idiopathic intracranial hypertension (IIH) is a rare disease and local epidemiological data on childhood IIH is limited. Presenting symptoms are often nonspecific and may mimic migraine. We report an adolescent with IIH who did not have classical predisposing risk factors. Annual incidence in Hong Kong is estimated to be 0.78 per 100,000 paediatric admissions to public hospitals. Enhanced awareness and prompt treatment is important as delayed diagnosis is associated with high risk of permanent visual damage. Urgent surgical intervention is necessary for those who do not response to medical treatment.

Key words Idiopathic intracranial hypertension; Papilloedema; Pseudotumor cerebri

Introduction

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is a rare condition characterised by raised intracranial pressure (ICP) with normal cerebrospinal fluid (CSF) composition and absence of hydrocephalus or space occupying lesions. It predominantly affects adult population, particularly overweight women of child-bearing age. The condition is rare in children.[1] As symptoms are non-specific and parents often reject invasive lumbar puncture (LP), diagnosis may be delayed resulting in permanent visual damage. We report an adolescent who did not have classical predisposing risk factors, and review the recent literature in the clinical management of IIH. Annual incidence is estimated from local epidemiological data.

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Case Report

A 14-year-old Chinese girl presented with bilateral peripheral visual loss and frontal headache for one year. Her headache was throbbing in nature and was aggravated by exercise. It was progressive with increasing frequency up to several times per day. Her headache was associated with transient visual obscurations, described as bilateral peripheral visual field loss that occurred for five to eight seconds each time. There were no other associated symptoms. No preceding head injury, viral illness or use of medication was found. Her birth history and past health were otherwise unremarkable.

Physical examination revealed a normal build girl with body weight at 25th percentile, and height at 50 to 75th percentile. Body mass index was 18 kg per m^2 (10 to 25th percentile), blood pressure 118/80 mmHg. There were no focal neurological deficits; systemic examination was unremarkable. Ophthalmologic examination revealed bilateral grade 4 papilloedema (Figure 1a). Extraocular

movement was normal. Goldman visual field test showed normal visual field on both eyes but mildly enlarged blind spot on right eye. Optical coherence tomography demonstrated thickening of nerve fibre layer on right eye. Visual evoked potential was normal.

Magnetic resonance imaging (MRI) of brain showed no ventricular dilatation (Figure 1b). Magnetic resonance venography showed no abnormalities in all major draining sinuses. Blood gas, serum cortisol, thyroid function, antinuclear antibodies, rheumatoid factor, protein C, protein S and lupus anticoagulant were all normal. Urine metabolic screening was unremarkable. LP revealed clear CSF with raised opening pressure of 33.5 cmH_2O. CSF cell count and biochemistry were normal; microbial smear and culture were negative.

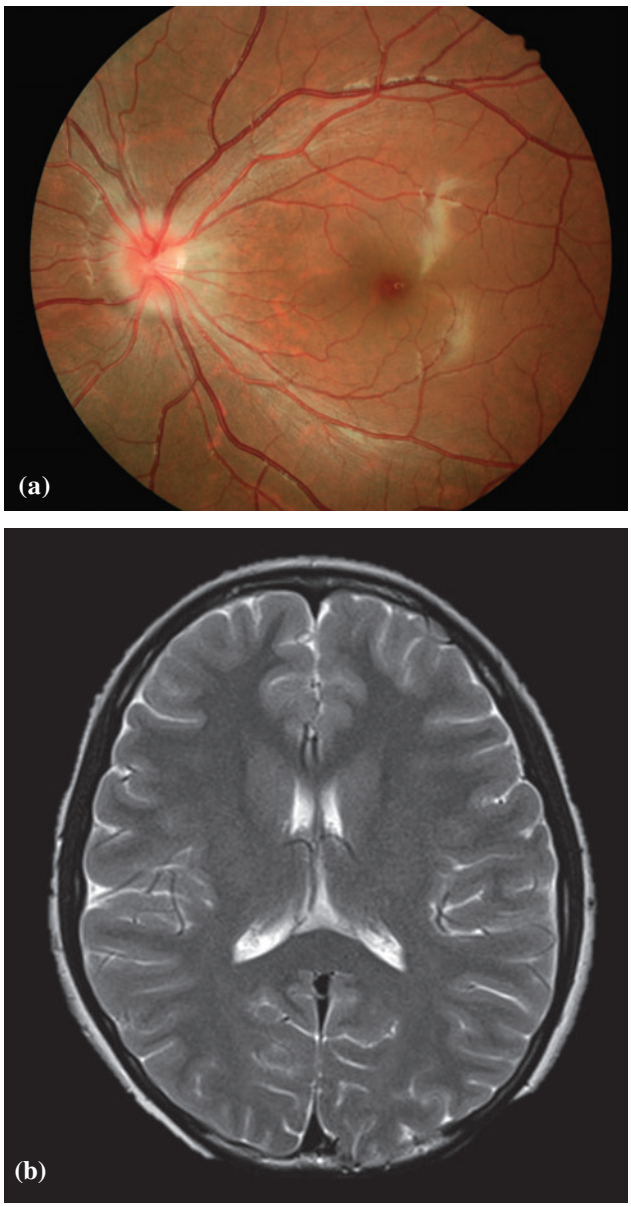

Figure 1 (a) Grade 4 papilloedema showing total obscuration on the disc of a segment of a major blood vessel on the disc. (b) Magnetic resonance image of brain showed normal size of ventricles.

Diagnosis of IIH was established according to the Modified Criteria of Dandy, which include symptoms and signs of raised intracranial pressure, absence of localising neurological signs, raised cerebrospinal fluid pressure with normal CSF composition, and normal sized ventricles.[3] She was started on acetazolamide 500 mg twice daily (25 mg/kg/day) and gradually stepped up to 1 gram twice daily. Two weeks later there was significant improvement in headache and visual obscurations, with partial resolution of papilloedema to grade 2. Follow up blood tests showed normal acid-base and electrolyte levels. However, further dose escalation was precluded by side effects including tingling sensation in the toes, anorexia and abdominal cramps. She was subsequently put on combination treatment with furosemide, with resolution of symptoms and papilloedema after four months.

Estimated Incidence in Hong Kong Children

We estimated the local disease incidence among children aged 0 to 18 years old by retrieving Hospital Authority (HA) clinical disease analysis and registration system (CDARS) data from 1st May 2007 to 1st May 2012. Of a total of 434,655 paediatric hospital admissions to all HA hospitals during the five year period, a total of 17 patients (9 males, 8 females) were diagnosed with IIH, yielding an annual incidence of 0.78 per 100,000 admissions.

Discussion

IIH is a rare condition in childhood. A recent study in United Kingdom has reported an estimated annual incidence of 0.8 per 100,000 child population aged 1 to 16 years,[1] which is concordant with our current estimated local incidence of 0.78 per 100,000 admissions. However, the true incidence may be underestimated as patients from private sectors and outpatient clinics were excluded in our calculation. Other factors such as coding errors in CDARS, presence of non-specific symptoms and refusal of LP might also contribute to underestimation of local incidence.

Heightened physician awareness is important to prevent visual sequelae in childhood IIH. Unfortunately, the presenting symptoms are often non-specific and may mimic migraine. Headache is typically generalised throbbing on presentation, aggravated by Valsalva manoeuvre. These may be associated with nausea, vomiting, stiff neck, retro-orbital pain, photophobia, pulsatile tinnitus and dizziness. Neurological examination is typically normal, with the exception of unilateral or bilateral sixth nerve palsies, which can occur in up to 50% of paediatric patients.[2] Previous studies had shown that around 60% of paediatric patients with IIH were older than 10 years old.[3] Unlike in adults and postpubertal subjects, female gender and obesity were not found to be risk factors among prepubertal children.[4] In the absence of common predisposing factors (Table 1), diagnosis may be delayed till the onset of visual complications. The latter may range from transient visual obscurations (2-53%), visual field defects (up to 90%), to irreversible deterioration in visual acuity (20%).[2,3] The overall risk of visual impairment is up to 40%, with blindness in 4% of patients.[3,4] High grade papilloedema, marked visual field loss, subretinal haemorrhage, hypertension and recent weight gain are risk factors for rapid progression and poor visual outcome.[2,3]

In children suspected of IIH, cranial imaging should be performed prior to LP to exclude space occupying lesion in the brain. MRI is superior to computer tomography for detecting pathologies such as posterior fossa lesions, isodense tumours, meningeal infiltration and cerebral venous sinus thrombosis. Subtle radiological findings indicative of increased ICP include flattening of posterior sclera, empty sella or distension of perioptic subarachnoid space.[5]

Data on normal range of CSF opening pressure is limited in children. A recent review defines CSF opening pressure of 25 cmH_2O as diagnostic in children of 8 years old or older, whereas pressure of 20 cmH_2O is used in those younger than 8 years of age.[3] The degree of raised CSF pressure at presentation is not an important prognostic factor.[6] Repeated LP is not recommended, unless there is relapse of disease or rapid visual deterioration.

Treatment of IIH is indicated in patients with loss of visual field or acuity, persistent headache and moderate to severe papilloedema.[2] Acetazolamide, a carbonic anhydrase inhibitor, is the first line medication in IIH. Variable effectiveness of 47-67% was reported in childhood cases.[4] Recommended starting dose is 25 mg/kg/day given in three to four divided doses, with maximum dose of 100 mg/kg/day or 2 grams /day.[4] As in the present case, dose escalation is often limited by side effects such as loss of appetite, fatigue, altered taste, paraesthesia of fingers and toes and abdominal cramps. Laboratory monitor is required to look for metabolic acidosis, acute tubular necrosis, potassium and magnesium depletion. Furosemide helps to reduce reabsorption of sodium and decrease total extracellular fluid. Combination therapy with acetazolamide has been shown to be effective in reducing CSF pressure with good clinical response.[4] Topiramate, with its carbonic anhydrase inhibitor activity and anorexic effect, has also been utilised in treatment for IIH. However, its efficacy in children has not been established.[4] Therapeutic LP is used as an acute relief of symptomatic IIH. However, ICP may quickly return to pre- treatment level after one to two hours.[4] LP is also poorly tolerated and difficult to perform in agitated children.

Majority of patients with IIH respond well to medical treatment. Most children have resolution of symptoms and papilloedema after one week to six months of treatment.

Table 1 Known medical conditions and drugs associated with idiopathic intracranial hypertension (IIH)[5,7]

Drugs	Tetracycline and derivatives
	Nitrofurantoin
	Nalidixic acid
	NSAIDs
	Corticosteroid therapy or withdrawal
	Growth hormone
	Oral contraceptive pills
	Lithium
	Vitamin A analogues
	Penicillin
Systemic diseases	Systemic lupus erythematosus
	Ulcerative colitis
	Iron deficiency anaemia
	Sickle cell disease
	Chronic renal failure
	Migraine
Endocrine	Diabetes mellitus
	Thyroid disease
	Hypoparathyroidism
	Polycystic ovarian syndrome
	Cushing's disease
	Adrenal insufficiency
Infection	Lyme disease
	HIV

NSAIDs: Nonsteroidal anti-inflammatory drugs; HIV: Human immunodeficiency virus

While the optimal treatment duration has not been established, in general medication may be tapered after 2 months with symptom resolution.[4] Medication failure ranges from 18% to 22%, and a third of patients may have persistent papilloedema after 10 months.[2,4] Recurrence of IIH, usually peaks during first 18 months of diagnosis, is reported in 5-22% of cases.[3,4,6]

Urgent referral to neuro-ophthalmologist and neurosurgeon is indicated if there is severe or rapid visual loss at onset, severe papilloedema with macular oedema and exudates, and intractable headache.[2,3] Children who do not respond to medication with worsening of symptoms and signs are also potential candidates for surgery. Surgical interventions include CSF diversion procedures such as lumboperitoneal or ventriculoperitoneal shunt, and optic nerve sheath fenestration.[2-4] To date, no data are available to compare efficacy of different surgical options. Choice of surgery mainly depends on availability of expertise and patient's preference.

Conclusion

IIH is rare in children, with an estimated annual incidence of 0.78 per 100,000 paediatric admissions in Hong Kong. Heightened physician awareness is important for early detection and prompt treatment to prevent irreversible visual loss.

Declaration of Interest

We declare that we have no conflict of interests.

References

1. Matthews YY, Deans F, Matyka K, et al. UK surveillance of Childhood Idiopathic Intracranial Hypertension (IIH). Arch Dis Child 2012;97(Suppl 1):A1-A186.
2. Spennato P, Ruggiero C, Parlato RS, et.al. Pseudotumor cerebri. Childs Nerv Syst 2011;27:215-35.
3. Standridge SM. Idiopathic intracranial hypertension in children: a review and algorithm. Pediatr Neurol 2010;43:377-90.
4. Matthews YY. Drugs used in childhood idiopathic or benign intracranial hypertension. Arch Dis Child Educ Pract Ed 2008; 3:19-25.
5. Shin RK, Balcer LJ. New developments in idiopathic intracranial hypertension. Curr Neurol Neurosci Rep 2001;1:463-70.
6. Soiberman U, Stolovitch C, Balcer LJ, Regenbogen M, Constantini S, Kesler A. Idiopathic intracranial hypertension in children: visual outcome and risk of recurrence. Childs Nerv Syst 2011;27:1913-8.