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Case Report Open Access

An Unusual Presentation of Atypical Idiopathic Intracranial Hypertension in a Pre-Pubertal Child

Sarah YF Chan*

Greenslopes, Queensland, Australia

***Corresponding author:** Sarah YF Chan, Greenslopes, Queensland, Australia, Tel: + 61410954081; +610400343772; E-mail: nahcharas@hotmail.com

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Abstract

Purpose: In paediatrics, Idiopathic Intracranial Hypertension (IIH) presents differently in pre- and post-pubertal children. This case report aims to bring awareness to uncommon presentations of IIH in a pre-pubertal child, highlighting the variability and exceptions of an atypical condition.

Observations: A 7-year-old female with a BMI in the 84th percentile presented to Rockhampton Base Hospital (Queensland, Australia) with a 2-month history of intermittent blurry vision, headache, vomiting, bi-temporal hemianopia and bilateral swollen discs. There were no common findings of obesity, neck stiffness, strabismus, abducens nerve palsy or common field defects such as enlarged blind spots, peripheral constrictions or paracentral scotomas. MRI brain/orbits with contrast performed had no MRI features of IIH, and lumbar puncture revealed an opening pressure of 6 cm H_20 with normal cerebrospinal fluid composition. Ophthalmologist, Paediatricians and Neurologist at Rockhampton Base Hospital and Lady Cilento Children's Hospital Brisbane (Queensland, Australia) were involved early on in the management of this patient. The patient's retinal nerve fibre layer thickness and field defects were found to be improving on formal HD-OCT and Humphrey Visual Field tests following early medical treatment with oral acetazolamide.

Conclusion: This report is important to increase awareness of uncommon presentations of an atypical condition and encourage early involvement of multidisciplinary teams in the management and treatment of early adolescent children with this condition.

Keywords: Paediatric idiopathic intracranial hypertension; Pre-pubertal idiopathic intracranial hypertension; Atypical idiopathic intracranial hypertension

Introduction

Idiopathic Intracranial Hypertension (IIH) is originally defined by Quincke as increased intracranial pressure (ICP) without a brain tumour [1]. This was later updated to a set of criteria called the Modified Dandy criteria where typical IIH manifests in young obese females who present with headaches and poor vision accompanied with elevated intracranial pressure, normal CSF constituents, and papilloedema in the absence of mass lesions, infective or vascular causes [1].

In paediatrics, IIH shares some but not all features of adult IIH and presents differently in pre-pubertal and post-pubertal children [2]. There are differences in the sex distribution at onset, and absence of significant correlation with obesity as incidences in children seem to peek at between 4-6 years and the other at 13-14 years [3]. In addition, majority of children have identifiable causes of intracranial hypertension or associated conditions which makes the diagnosis not quite idiopathic [4,5]. It is also known that sometimes children have signs that allude to a possible posterior fossa lesion such as ataxia, facial palsy, nuchal rigidity, malaise, irritability, torticollis or Babinski sign [1].

The characteristics and visual defects of IIH in paediatrics have been reviewed by various authors as IIH in children is rare compared to the incidence of IIH in the general population of 0.9: 100,000 [6]. This case report serves to highlight the variability and exceptions of a condition that is at present already known to be atypical.

Case Report

A 7-year-old female was referred to the Ophthalmology Outpatient Department at Rockhampton Base Hospital (Queensland, Australia) by a local optometrist who found bilateral swollen optic nerve heads. This patient presented with a 2-month history of intermittent blurry vision every few days, noted whilst she was looking at her teacher in front of the classroom or reading a book. About a month after noticing the intermittent blurry vision, she noticed intermittent frontotemporal headaches that occurred every other day on waking or after school. She denied any other associated symptoms such as nausea, vomiting, recent head injury, recent falls, and family history of blindness or vasculitis. This is on a background of no previous history of headaches, no significant past medical history, immunisations which are up to date, and a family history of a grandmother, a great-grandmother and an aunt with migraines.

On examination, she demonstrated a body mass index of 18.6 kg/m^2 (84th percentile) and her visual acuities were (R) 6/5 and (L) 6/5 with IOPs of (R) 10 mmHg and (L) 14 mmHg. She had normal optic nerve functions, no relative afferent pupillary defect, full extraocular muscle movement and normal anterior segment examination. Her visual fields on confrontation revealed a distinct right more than left bi-temporal hemianopia. And her posterior segment examination revealed bilateral

swollen optic nerve heads in the left greater than the right, with no haemorrhages, vasculitis or vitritis. Her blood tests returned negative for any inflammatory, infectious or autoimmune causes (Table 1).

CRP	<2.0 (<5.0 mg/L)
Free T4 & TSH	9.6 (7.5–17 pmol/L) & 1.4 (0.7– 4 mU/L)
ANA & ENA	Negative
dsDNA (RIA)	0 (<7 IU/mL)
P-ANCA & C-ANCA	Negative
MPO-ANCA	<3 (<20 CU)
PR3-ANCA	<2 (<20 CU)
C3 & C4	1.15 (0.9–1.8 g/L) & 0.18 (0.1–0.4 g/L)
Anti-Cardiolipin (IgG) Abs (aCL-IgG)	7 (<20 CU)
Anti-beta 2 glycoprotein I (IgG)	10 (<20 G units)

Table 1: Laboratory Investigations April 2016.

OCT retinal nerve fibre layer thickness measured with ZEISS CIRRUS HD-OCT Model 4000 Dual-core (Software 7.0.1.290) gave an average of 152 µm on the right and 217 µm on the left (range of normal 72.5-136.9 µm) (Figure 1).

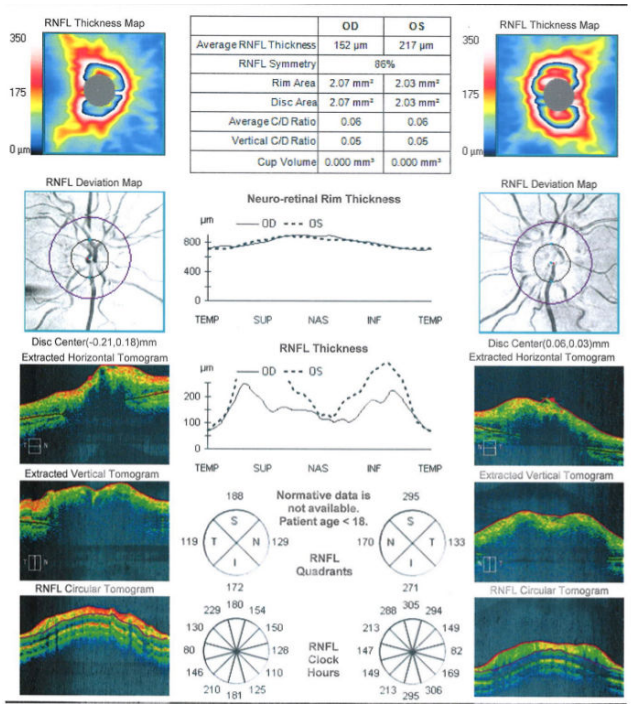


Figure 1: Optic Nerve Head and Retinal Nerve Fibre Layer Analysis May'2016 prior to commencement of oral Acetazolamide.

MRI brain/orbits with contrast was performed using SIEMENS MAGNETOM Verio 3T MRI where T1 and T2 weighted images of the brain and T1 post contrast images of the orbits showed no abnormalities. Lumbar puncture revealed an opening pressure of 6 cm H_2O with no organisms seen on CSF gram stain, normal protein levels, normal glucose levels and no growth on culture.

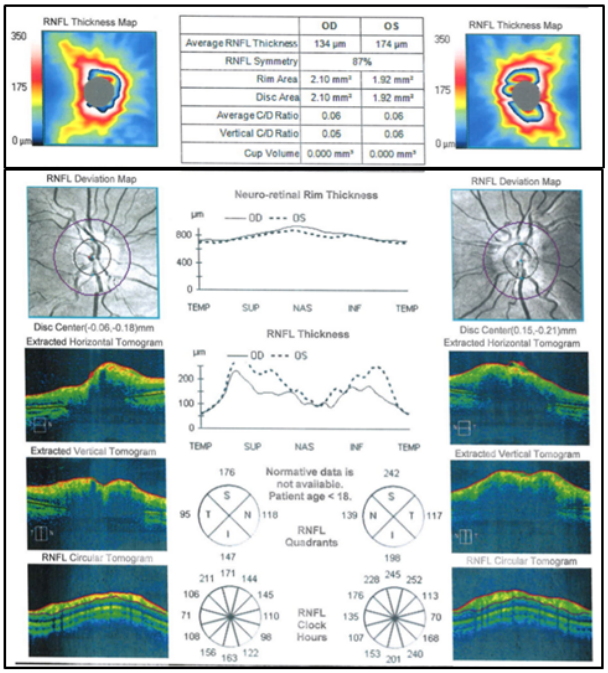
In view of her visual field defects, intermittent headaches and bilateral swollen optic nerve head, she was further referred on to the Paediatricians at Rockhampton Base Hospital who referred her on to Lady Cilento Children's Hospital Brisbane (Queensland, Australia) for opinions from the Paediatric Neurology and Ophthalmology teams there. She was then commenced on PO Acetazolamide 125 mg BD with no improvements in her headache. Unfortunately, she did not tolerate an increased dose of PO Acetazolamide as she experienced adverse side effects such as nose bleeds and per vaginal bleeding despite no evidence of blood dyscrasias (Table 2).

INR	1.1 (0.9–1.2)
Prothrombin time	13 (9–13 s)
APTT	33 (24–39 s)
Fib (derived)	3.4 (1.7–4.5 g/L)
Platelets	315 x 10^9 /L (150–400 x 10^9 /L)
Lupus A/C screen	Negative
Factor 8:C	0.57 (0.5–1.5 U/mL)
Factor 9	1.04 (0.5–1.5 U/mL)
von Willebrand Factor Antigen (Ag)	0.54 (0.5–2.0 U/mL)
Von Willebrand Factor RCOF	0.51 (0.5–2.0 U/mL)
Von Willebrand Factor Collagen Binding	0.53 (0.5–1.5 U/mL)

Table 2: Further laboratory investigations April 2017.

This patient's headaches continued to increase in frequency despite increasing PO Acetazolamide to 250mg BD. However, her swollen discs bilaterally gradually reduced in retinal nerve fibre layer thickness measurements (Figure 2) and her visual field defects which was objectively measured using the 30-2 ZEISS Humphrey Field Analyzer HFA3 seemed to have returned grossly normal (Figure 3 and Figure 4). She has also maintained the same good visual acuities with intact optic nerve function throughout.

At this stage, she is being regularly reviewed every 3 months by a multidisciplinary team of Ophthalmologists, Paediatricians & Neurologists at Rockhampton Base Hospital as well as Lady Cilento Children's Hospital Brisbane to monitor resolution or progression of her optic nerve head swelling, visual acuities, visual fields and optic nerve function. There is also a plan to consider addition of Topiramate to her current Acetazolamide treatments should there be ongoing poor response to acetazolamide or worsening of any of her examination parameters.



	OD	OS
Average RNFL Thickness	134 µm	174 µm
RNFL Symmetry	87%	
Rim Area	2.10 mm²	1.92 mm²
Disc Area	2.10 mm²	1.92 mm²
Average C/D Ratio	0.06	0.06
Vertical C/D Ratio	0.05	0.06
Cup Volume	0.000 mm³	0.000 mm³

Figure 2: Optic Nerve Head and Retinal Nerve Fibre Layer Analysis May'2017 post commencement of treatment with oral Acetazolamide.

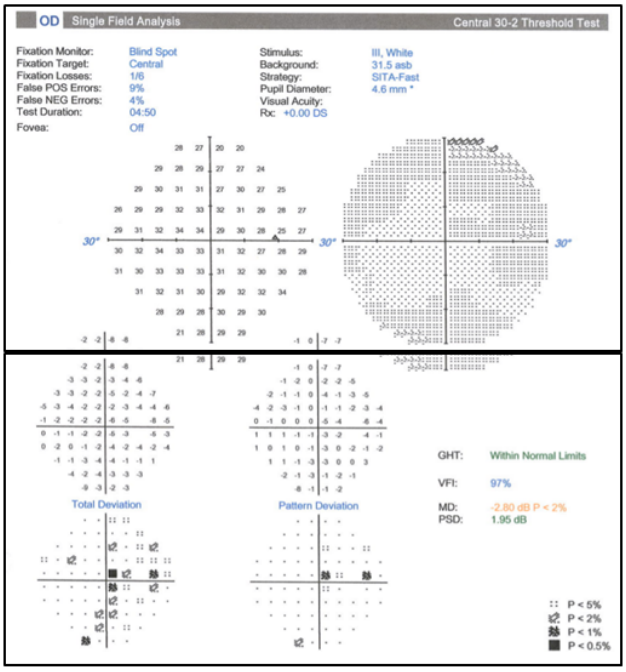


Figure 3: 30-2 Humphrey Visual Fields OD post commencement of treatment with oral Acetazolamide.

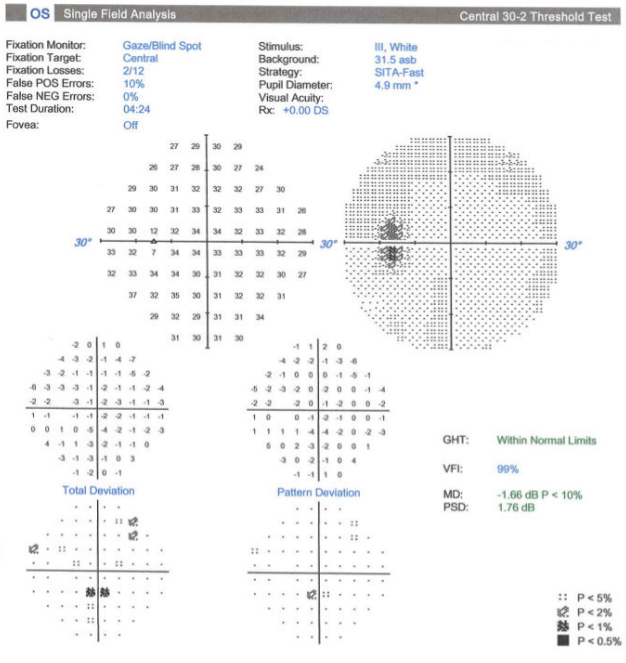


Figure 4: 30-2 Humphrey Visual Fields OS post commencement of treatment with oral Acetazolamide.

Discussion

Although it has been documented that IIH is considered atypical in children and men according to the Modified Dandy Criteria 1985 [7], we believe that this case has an atypical presentation due to a multitude of uncommon features.

Firstly, the patient's age in itself is an uncommon feature as this patient was a 7-year-old female [7-9]. Multiple studies have reported IIH in children being more common between the ages of 11-16 [7-9]. In fact, more than 60% of children with IIH are older than 10 years of age [7].

Secondly, this patient is a female who falls in the pre-pubertal age group of IIH which has shown to usually have a male predominance and often associated with adiposity that exceeds the threshold for obesity; this goes against the current consensus that there is no sex predilection and no association to obesity between this subgroup of early adolescent children [2,3,5,7].

Thirdly, even though there has been another documented study stating that the likelihood of obesity in children with IIH are between the ages of 7-12 as 70% of this study's children fulfilled the criteria for obesity [8], our patient in this case report had a BMI in the 84th percentile which is considered within a normal range [10].

Fourthly, the most common features of IIH in pre-pubertal children are known to involve neck stiffness, strabismus or abducens nerve palsy, but in this case, this patient only had symptoms of headache,

vomiting and blurring of vision [1,11]. Visual field defects in IIH is as common as 60% of pre-pubertal cases, and usually presents in the form of enlarged blind spots, peripheral constrictions or paracentral scotomas secondary to swollen discs [11]. This patient on the other hand had a bi-temporal hemianopia with swollen discs bilaterally. Even though her visual fields were examined by confrontation and not on formal visual field testing, a field defect pattern such as this appears unusual and not found or recorded in literature search thus far.

Furthermore, MRI scans are usually the traditional method of neuroimaging to exclude other causes of raised intracranial pressure or space occupying lesions [12]. There are 6 neuroimaging features of raised intracranial pressure and the presence of 3 or more of these MRI features is 95% specific in predicting IIH (Table 3) [12].

1	Flattening of posterior sclera
2	Enhancement of prelaminar optic nerve
3	Distension of peri-optic subarachnoid space
4	Intraocular protrusion of the prelaminar optic nerve
5	Vertical tortuosity of orbital optic nerve
6	An empty sella

Table 3: The 6 neuroimaging features of raised intracranial pressure [12].

This patient's MRI on the other hand was essentially unremarkable and revealed no evidence of flattening of posterior sclera, enhancement of prelaminar optic nerve, distension of peri-optic subarachnoid space, intraocular protrusion of the prelaminar optic nerve, vertical tortuosity of orbital optic nerve or an empty sella. This further supports the emphasis of such an unusual presentation of an existing atypical condition as neuroimaging in itself did not demonstrate any predictive features of IIH.

Lastly, lumbar puncture opening pressures in pre-pubertal IIH patients have been documented to be elevated and at least >18 cm H_2O [6,13,14]. This is another unique finding as this patient's opening pressure was recorded at 6 cm H_2O and instead of giving relief to the patient, she suffered a great deal after from post lumbar puncture headaches; refuting the possibility of a technical error in this procedure.

Given all these uncommon findings, the patient's retinal nerve fibre layer thickness and field defect resolved on formal field testing following medical treatment of acetazolamide which is consistent with studies documenting that early medical treatment is usually successful in most paediatric IIH patients [3,7,11].

Conclusion

In summary, this report was to make known the variable presentations of an uncommon condition like atypical IIH. Thus, increasing awareness and encouraging detailed examination, investigations as well as early involvement of multidisciplinary teams in the management and treatment to achieve the best outcome in early adolescent children.

Patient Consent

This report does not contain any identifiable information and therefore no consent was obtained to publish this report.

Disclosures

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