

Okposio M M
Abhulimhen-Iyoha BI

Idiopathic intracranial hypertension with altered consciousness in a Nigerian school girl: a case report and review of the literature

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Okposio M M (✉)
Department of Pediatrics, Mariere
Memorial Central Hospital,
Ughelli, Delta State, Nigeria
Tel: +2348034042120
E-mail: mattokmatok@yahoo.com

Abhulimhen-Iyoha BI
Department of Child Health,
University of Benin Teaching
Hospital,
Benin City, Nigeria

Abstract Idiopathic intracranial hypertension (IIH) is a clinical condition of increased intracranial pressure (ICP) without an obvious underlying pathological brain lesion. It is usually characterized by headache, neck pain, vomiting, visual disturbances, papilledema, cranial nerve palsy or a combination of these signs and symptoms. The diagnosis of IIH is often made in a patient with intact consciousness. We present a teenager who

developed altered consciousness while being treated for suspected meningitis and later found to have IIH. This case brings to the fore the need for a high index of suspicion even in situations where features appear atypical to avoid the unpleasant consequences of a misdiagnosis.

Keywords: Idiopathic intracranial hypertension, Altered consciousness, adolescent girl, Nigeria.

Introduction

Idiopathic intracranial hypertension (IIH) is a neurological disorder which has undergone a series of name changes since it was first described as meningitis serosa by Quinke in 1893.¹ It was last known as benign intracranial hypertension, however, because of the potential visual loss that accompany the papilledema, Corbett and Thompson substituted benign with idiopathic.² Idiopathic intracranial hypertension is a clinical condition of increased intracranial pressure (ICP) without an obvious underlying pathological brain lesion. It is a rare disorder in children and very few cases have been reported in Nigeria.³ It is usually characterized by headache, neck pain, vomiting, visual disturbances, papilledema, cranial nerve palsy or a combination of these signs and symptoms.⁴ These clinical features can also be present in some other central nervous system morbidities seen in our region such as meningitis, as a result IIH can easily be misdiagnosed especially by non specialist practitioners. This is more so in children where atypical features of IIH have been reported.⁵ Therefore, a high index of suspicion is required to accurately make the diagnosis of idiopathic intracranial hypertension.

We present the case of a 13-year-old school girl with idiopathic intracranial hypertension whose level of consciousness deteriorated while being treated for suspected meningitis.

Case Report

O.O is a 13-year-old school girl who was referred by a private medical practitioner and presented at the Emergency Unit of Lily Hospital Warri, a private secondary health care facility in Delta State, Nigeria. She was on admission at the referring hospital for seven days having presented there with a five-day history of generalized throbbing headache. The headache was worse in the morning and aggravated by straining but was transiently relieved by analgesic. There was an associated neck pain and blurring of vision; however, there was no vomiting and no fever. There was also no history of seizures or trauma to the head and she was not on any medication prior to the onset of symptoms. She was placed on anti malaria medication on an outpatient basis, however, she represented to the same hospital two days later because of worsening symptoms. She was subsequently admitted after re-evaluation and treatment was commenced for suspected meningitis with intravenous ceftriaxone and analgesics. The patient's condition, however, progressively worsened with restlessness, collapsing episodes, followed by deterioration in the level of consciousness which necessitated her referral.

At presentation, we found an obese young adolescent (BMI 33.1), with altered consciousness, a Glasgow coma scale of 12/15. She was afebrile (T ° 37 °C), not pale, not jaundiced, pupils were dilated and sluggishly reactive to light. Fundoscopy revealed fundi with blurred disc margin and engorged retinal vessels sugges-

tive of bilateral papilloedema. She had bilateral abducent nerve palsy and right sided facial nerve palsy. Tone and tendon reflexes were normal, but there was nuchal rigidity. Her pulse rate was 86 beats per minute and blood pressure 120/80mmHg. Further systemic examination was largely unremarkable.

A provisional diagnosis of raised intracranial pressure ? cause was made. The patient was then commenced on intravenous furosemide 60mg 12 hourly and intravenous dexamethazone 4mg 6 hourly. A computerized tomography (CT) scan was done and revealed a normal cranial vault, sella and suture lines. The brain slices revealed normal tissues as well as ventricular system and there was no areas of hyperattenuation (Figure 1,2&3). Lumbar puncture was done using a 22G, 3.5-inch spinal needle with the patient in the lateral decubitus position, relaxed and legs extended after the CT scan ruled out an intracranial mass lesion. The cerebrospinal fluid (CSF) was clear and colourless and was under pressure. The biochemical and microbiological studies were all normal. The complete blood count, platelet count, renal and liver function tests were normal. The prothrombin time and partial thromboplastin time were not prolonged. A diagnosis of Idiopathic Intracranial Hypertension was therefore made.

Intravenous furosemide was discontinued on the third day of admission after she became fully conscious and oral acetazolamide 500mg 12hrly was commenced. She made remarkable clinical improvement with resolution of the headache and was discharged home after 12 days of hospitalization. She was on two weekly follow-up visit during which an assessment of cranial nerves 6 and 7 function were done. In addition, visual acuity and visual field assessment were also done because of residual visual disturbances. By the fourth follow-up visit, the cranial nerves dysfunction as well as the visual obscuration had completely resolved.



Fig1

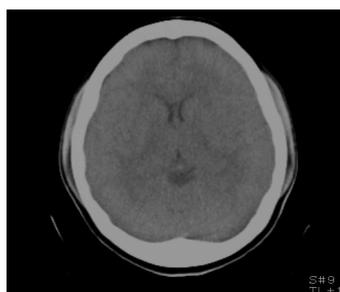


Fig 2

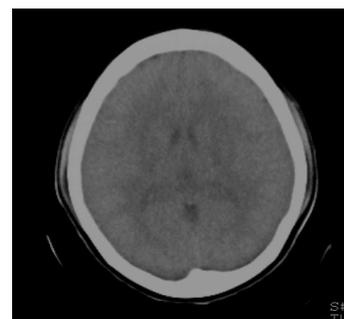


Fig 3

Discussion

Idiopathic intracranial hypertension is a neurological disorder that is characterized by increased ICP in the absence of an intracranial space-occupying lesion. It is a diagnosis of exclusion made largely based on clinical parameters but radiological and laboratory studies have a role in confirming the diagnosis.⁶

The overall annual incidence is 0.9 per 100,000 and there is a strong female predilection.⁷ In young adults whose body weight is 10-20% above normal, the incidence may be as high as 19.3% per 100,000.⁸ However, in the paediatric age group, especially in pre-pubertal children, it is considered relatively rare and the incidence is equal among boys and girls.⁹ After puberty, predilection for female and association with obesity increases, likely as a result of hormonal changes.¹⁰ Our patient was an obese young adolescent school girl with a BMI of 33.1 and therefore had risk factors for IIH.

The pathogenesis of IIH is still not clear. Many postulations have been put forward based on findings of neuroradiologic research and studies of cerebrospinal fluid hydrodynamics. These include a possible decreased cerebrospinal fluid absorption by the arachnoid villi, or an increased cerebrospinal fluid production by the choroid plexus. It is also postulated that perhaps, an increased water volume of the brain content diminishes its compliance, explaining the normal or reduced ventricular size.¹¹

Headache is the commonest symptom of IIH in both children and adult. It is usually worse in the morning, sometimes awakens the patient from sleep and increased with valsalva manoeuvre. The headache may be associated with nausea and vomiting and very frequently there are ophthalmological symptoms such as decreased or blurred vision, diplopia and transient visual obscurations.¹²

The diagnosis of IIH is often made based on modified Dandy's criteria¹³ and these include symptoms of increased intracranial pressure, no or false localizing neurological signs, normal brain imaging result, an awake and alert patient, normal cerebrospinal fluid finding and no identifiable cause of increased ICP. Our patient met

the diagnostic criteria except that she had impaired consciousness at presentation. Although, she was conscious and alert when she was first seen at the referring hospital, the subsequent alteration of consciousness makes this case somewhat atypical. It is important to note that specific paediatric diagnostic criteria have not been established. Interestingly, children with IIH may display a greater spectrum of clinical presentation than adults. These may include irritability, apathy, somnolence, ataxia, dizziness neck pain, stiff neck and seizure¹⁴ Impairment of consciousness in our patient was preceded by a series of collapsing episodes. This, we thought could have been due to a plateau wave; a phenomenon first described by Lundberg.¹⁵ It is characterized by an acute elevation of ICP in a situation where baseline ICP is already moderately elevated. A plateau wave develops as a result of a rapid increase in intracerebral blood volume. These acute elevations of ICP have been observed in patients with brain tumour, subarachnoid haemorrhage, acute hydrocephalus and idiopathic intracranial hypertension.¹⁶ and may be triggered in ambulatory patient by postural changes. It follows a reduction in intracranial compliance. The normal process of cerebral autoregulation is the result of a slow rise in cerebral blood volume as a result of vasodilatation which occurs because of a reduction in cerebral perfusion pressure (CPP). However, when CPP drops below a critical level, the rate of vasodilatation also increases dramatically, resulting in a rapid increase in cerebral blood volume and a sudden sustained increase in ICP. This may produce paroxysmal symptoms in patient with IIH and these include among others alteration in consciousness, postural and motor control¹⁷

Multiple cranial neuropathies was another unusual finding in our patient. Most cases of IIH presents with abducent nerve palsy.¹² In this case, both abducent nerves and the right facial nerve were affected. Facial nerve palsy in IIH has been described in the literature in about 2-6% of cases.¹⁸ Some reports suggest that increase pressure at the posterior fossa and enlargement of the fallopian canal may be responsible.¹⁹ Other atypical features of IIH that have been reported include hypoglossal nerve palsy, hyperreflexia with positive Babinski sign, choreiform movement and nystagmus.²⁰

The prior use of an antibiotics in this patient could have been a limitation because of the possibility of a partially treated meningitis; given the atypical features seen this patient. However, this was less likely because there is no significant effect of previous antimicrobial therapy on CSF cell count or glucose and protein among patients with positive or negative CSF culture.²¹ This was the finding of Shohet et al in a review of 115 children with clinical diagnoses of meningitis, of which 47 had received antimicrobial therapy before hospitalization. Our patient had normal microbiological and biochemical CSF results .

The choice of neuroimaging studies for suspected cases of IIH is still the biggest controversy among physicians.¹³ Although, magnetic resonance imaging (MRI)

and magnetic resonance venography (MRV) are recommended,²⁰ cranial tomography scan can be used especially in places where MRI is not available. It can exclude hydrocephalus and most mass lesions as well as venous sinus thrombosis, meningeal infiltrations and isodense tumours.^{13, 14} Neuroimaging are mandatory not only to exclude potential secondary causes of elevated ICP but the possibility of herniation prior to a lumbar puncture.. The result of neuroimaging should be normal in IIH, however, signs of increased intracranial pressure may be found such as flattening of the posterior globe where the optic nerve inserts in 80% of patients, empty sella in 70% and distension of perioptic nerve sheath in 45%.¹³ In addition, a lumbar puncture in a patient who has IIH should display an increased opening pressure as well as normal cell count, normal glucose concentration, normal or low protein content and the absence of infection. In our patient neuroimaging studies as well as CSF biochemical and microbiological studies were all normal fulfilling a major diagnostic criterion. Although manometry could not done to determine the exact CSF pressure because of non availability, the flow rate was suggestive of a CSF pressure that was very high. Robert et al showed in a study that CSF drops counts per given time can be used to estimate CSF pressure with a reasonable degree of accuracy.²²

The key to a good outcome in IIH is early recognition and prompt treatment. This is to forestall permanent optic nerve death and subsequent visual loss. Weight reducing measures should form part of the initial treatment regimen.²³ Drugs that have been used with success include acetazolamide,, a sulfa-derived diuretic and carbonic anhydrase inhibitor that reduces CSF production. The starting dose in children is 25mg/kg/day which may be increased to 100mg/kg/day (maximum, 2g/day).²⁴ Furosemide, a loop diuretic, can be used in combination with or as an alternative to acetazolamide at a dose of 1mg/kg/dose. Corticosteroids have also been used especially in patient with rapid visual deterioration.²⁵ Other interventions found useful in selected cases include serial lumbar puncture, although this is technically difficult in children and their pressure lowering effect is only temporary. Surgery may be indicated such as Bariatric surgery, CSF diversion procedures and optic nerve decompression.²⁶

Idiopathic Intracranial Hypertention is a rare disease especially in children and may be easily confused with more common central nervous system morbidities. A high index of suspicion and appropriate investigations are therefore needed to reduce the attendant sequelae of a misdiagnosis.

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