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Research Article

PSEUDOTUMOR CEREBRI: A CASE REPORT AND REVIEW OF THE LITERATURE

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Abstract

Idiopathic intracranial hypertension (IIH) is a syndrome in which there is an increase in intracranial pressure (ICP) of unknown etiology. Previously known as pseudo tumour cerebri or benign intracranial hypertension, this condition frequently occurs in obese women of childbearing age. It is defined by the revised modified Dandy criteria. Our objective is to share this clinical observation received in our department in order to keep attention of Scientifics on the consequences that it can make in children mostly vision problem.

Keywords: Idiopathic Intracranial Hypertension, Case Report.

INTRODUCTION

Clinical observation

An 18-year-old asthmatic patient complaining about retro orbital headaches that had been evolving for 3 months, of progressive intensity associated with visual blur and intermittent vomiting, as well as photophobia and phonophobia. She consulted the ophthalmology department where a stage II papilledema was diagnosed and a cerebral CT scan was performed; given the results, she was referred to our department for better management.

Examination

Intracranial hypertension syndrome, obesity, ophthalmological examination: visual acuity reduced to 1/20, absence of the blink reflex at the threat, a lazy photomotor reflex, normal ocular tension, fundus shows stage III papilledema with invasion of the peripapillary retina (picture in favour of stasis oedema). Cerebral CT scan normal and CT scan of the facial massif: in favour of bilateral maxillo ethmoid sinusitis. Treatment with acetazolamide tablet, lumbar puncture for evacuation and analgesia was initiated. She then underwent a ventriculo-peritoneal shunt. The clinical evolution was marked by an improvement of the headaches and the persistence of visual disorders.

DISCUSSION

Epidemiology

Idiopathic intracranial hypertension is commonly seen in obese women of childbearing age, with a worldwide incidence of about 12-20 per 100,000 people per year in this group, 5-7 but only 0.5-2 per 100,000 people per year in the general population [Table 1]. In the absence of large epidemiological studies, prevalence is poorly documented in male and paediatric populations. Obesity rates are reportedly lower in children with idiopathic intracranial hypertension than in women, and more atypical features tend to be present in these groups.

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Although no UK-wide assessment of the prevalence or incidence of idiopathic intracranial hypertension has taken place, small retrospective case reviews have been carried out in Sheffield and Northern Ireland. The reported incidence rates were 1 - 56 per 100,000 people per year in Sheffi eld and 0.5 per 100,000 people per year in Northern Ireland. One of the largest retrospective reviews of idiopathic intracranial hypertension was conducted in Israel, with an incidence of 2.02 per 100,000 people per year in the population, rising to 5.49 per 100,000 per year in women of childbearing age. The mean age of diagnosis is between 25 and 36 years, with a clear female predominance; a large retrospective study of 721 patients with idiopathic intracranial hypertension reported only 66 [9%] as male. Of the total cohort of all studies reviewed, 57-100% of people with idiopathic intracranial hypertension are obese (Keira *et al.,* 2016; Biousse and Bousser, 2006).

Pathogeny

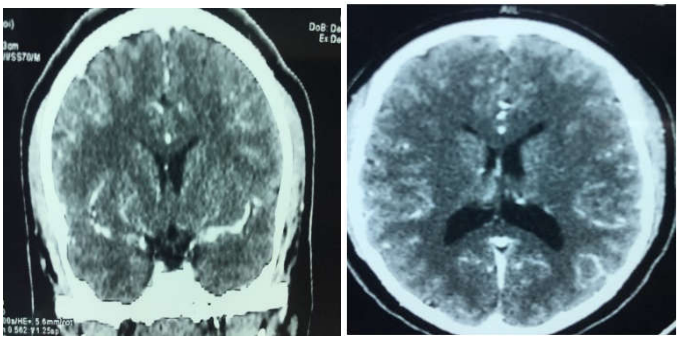
The pathogenic mechanisms of idiopathic intracranial hypertension are currently unknown (Matthew *et al.,* 2010; Keira *et al.,* 2016; Biousse and Bousser, 2006). Several hypotheses have been put forward: hyper secretion of cerebral spinal fluid (CSF), venous pressure of the sinuses, cerebral oedema, obesity, hormones (GH, TSH), secondary causes (tetracyclines, minocyclines, doxycillin, nitrofurantoine, sulphonamides, nalidixic acids, vitamin A deficiency, corticoids, anemia, Addison's disease, cushing's disease, respiratory disorders, chromosomal disorders, renal failure, autoimmune disorders (Keira *et al.,* 2016).

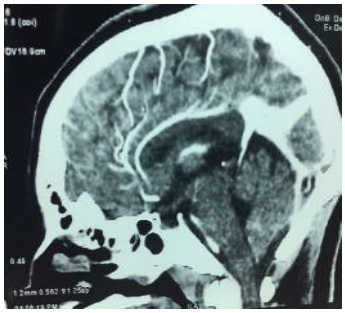
Clinical

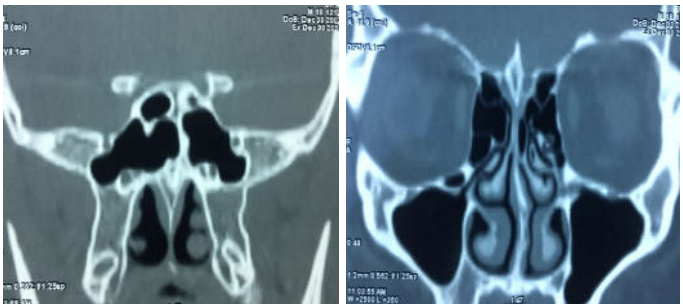
The clinical presentation of idiopathic intracranial hypertension is highly variable, which can lead to significant delays in diagnosis.

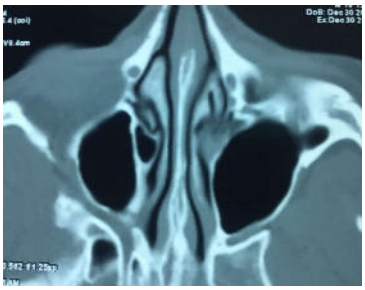
- Headache is a predominant feature of the disease, very heterogeneous. Pressure headache, holo cranial, frontal or retro-orbital, worse on awakening or with Valsalva type manoeuvres, and improved with lumbar evacuation. Many report migraine-like features, including unilateral or focal flutter, accompanied by nausea, photophobia and phonophobia (Keira *et al.,* 2016).

- Unilateral or bilateral transient visual obscurations, described as a brief (less than 60 s) loss of vision, occur with a postural challenge, Disruption of the microcirculation to the optic nerve head, caused by tissue engorgement leading to transient ischaemia, is described as the cause of this event. They are not specific to papilledema, but occur with other optic nerve disorders (Keira *et al.,* 2016).
- Pulsatile tinnitus: Turbulent flow in the transverse venous sinus, probably secondary to venous sinus stenosis, is thought to cause pulsatile tinnitus. Compression of the interior jugular vein may therefore relieve tinnitus (Keira *et al.,* 2016).









- Other symptoms include dizziness, visual loss, horizontal diplopia and cognitive impairment. Cognitive impairment, affecting reaction time and processing speed, rhinorrhea (Keira *et al.,* 2016).
- Ophthalmological examination: Abnormalities identified on any visual function test. Idiopathic intracranial hypertension causes significant visual morbidity through loss of visual field, visual acuity or both. Loss of acuity is thought to occur late in the disease. Papilledema refers to swelling of the intraocular tract, the (pre-laminar) part of the optic nerve head. Although usually bilateral, papilledema can be unilateral (Keira *et al.,* 2016).
- In our series, the examination included an intracranial hypertension syndrome (headache, vomiting, visual disturbances), obesity, photophobia and phonophobia. Ophthalmological examination: visual acuity reduced to 1/20, absence of the blink reflex at the threat, a lazy photomotor reflex, normal eye pressure, fundus evidences stage III papilledema with invasion of the peripapillary retina (picture in favour of stasis oedema)

Additional examinations

By definition, in idiopathic ICH, brain imaging is intended to rule out an expansive process, hydrocephalus, and cerebral thrombophlebitis or dural fistula.

- MRI, with T1, T2 and gradient echo sequences in particular, is essential for the diagnosis of idiopathic ICH, especially to rule out cerebral venous thrombosis (Matthew *et al.,* 2010; Keira *et al.,* 2016; Biousse and Bousser, 2006; Bidot, 2016; Deborah and Daniel, 2004; Susan *et al.,* 2018; Peter, 2015; Beau *et al.,* 2018; Degnan *et al.,* 2011).
- It can be coupled with venous magnetic resonance angiography (MRA) or CT angiography, thus avoiding the need for conventional angiography in most cases (Matthew *et al.,* 2010; Biousse and Bousser, 2006).
- In no case can a brain X-ray be considered sufficient (Matthew *et al.,* 2010; Keira *et al.,* 2016; Biousse and Bousser, 2006; Beau *et al.,* 2018; Degnan *et al.,* 2011).
- Suspicion of a dural fistula may also justify conventional arteriography (Biousse and Bousser, 2006).
- Once the diagnosis of idiopathic ICH is confirmed, an endocrine and metabolic workup is usually performed to look for a contributing factor. The search for anaemia and sleep apnoea syndrome completes the evaluation of these patients (Biousse and Bousser, 2006).
- Lumbar puncture then confirms the diagnosis of idiopathic ICH and further investigations are sometimes performed to investigate the causes (Matthew *et al.,* 2010; Keira *et al.,* 2016; Biousse and Bousser, 2006; Bidot, 2016; Deborah and Daniel, 2004).
- In our series, only the facial and cerebral scans were performed and showed a normal cerebral appearance and bilateral maxillo-ethmoid sinusitis.

Diagnosis

The diagnosis is based on the Dandy criteria modified in 2013 (Matthew *et al.,* 2010; Keira *et al.,* 2016; Biousse and Bousser, 2006; Bidot, 2016; Deborah and Daniel, 2004; Susan *et al.,* 2018; Peter, 2015; Beau *et al.,* 2018; Degnan *et al.,* 2011; Deborah *et al.,* 2002):

A definite diagnosis can be made if the patient meets criteria A to E. The diagnosis is likely if the patient meets criteria A to D, but the CSF pressure is < 25 cm of water.

a. Papillary stasis oedema
b. Normal neurological examination, except for VI paralysis
c. Neuroimaging: normal cerebral parenchyma without hydrocephalus, intracranial expansive process, or structural

abnormality, and absence of meningeal contrast on MRI without and with contrast. Venous MRI angiography is necessary in atypical patients; if MRI is unavailable or contraindicated, a cerebral CT scan without and with contrast medium can be performed in association with a venous CT angiography with contrast medium Normal composition of the LCS

d. Elevated CSF opening pressure (≥ 25 cm of water in adults and ≥ 28 cm of water in children [25 cm of water if unsedated and non-obese]) obtained from a lumbar puncture performed in lateral recumbency.

According to the modified Dandy criteria, our patient was A, B, C and D.

f. Treatment

There is no consensus on the best management strategy for idiopathic intracranial hypertension, mainly due to a lack of solid evidence (Keira *et al.,* 2016).

For optimal management of patients with HII, one should (Deborah and Daniel, 2004):

- ✓ Clear communication between clinicians for seamless joint care between different specialties.
- ✓ Weight loss reduces ICP and has been shown to improve papilledema and headaches.

The main principles of management of benign intracranial hypertension are (Deborah and Daniel, 2004):

1. Treating the underlying disease
2. Protecting vision
3. Minimise the morbidity of headaches.

But an evidence-based approach may begin to be adopted given recent trials that have evaluated the efficacy of acetazolamides and weight loss in patients with intracranial hypertension (Keira *et al.,* 2016).

- ✓ Weight loss (Biousse and Bousser, 2006; Bidot, 2016; Deborah and Daniel, 2004; Susan *et al.,* 2018)
- ✓ Lumbar evacuation and exploration puncture
- ✓ Medical treatment:
  - Acetazolamide: Frequently used in the management of idiopathic intracranial hypertension as it is thought to alter CSF secretion at the choroid plexus. It inhibits the enzyme carbonic anhydrase, which catalyses the conversion of water and carbon dioxide to bicarbonate and hydrogen ions, ion carriers. The resulting reduction in movement of ions and water through the choroid plexus subsequently reduces CSF secretion (Matthew *et al.,* 2010; Keira *et al.,* 2016; Biousse and Bousser, 2006; Bidot, 2016; Deborah and Daniel, 2004; Susan *et al.,* 2018).
  - Furosemides: Sometimes used to treat intracranial hypertension, but very little evidence to support its clinical use (Keira *et al.,* 2016; Biousse and Bousser, 2006; Bidot, 2016; Deborah and Daniel, 2004; Susan *et al.,* 2018).
  - Topiramate: An antiepileptic and migraine prophylactic, is becoming increasingly popular as a management option in idiopathic intracranial hypertension. The advantages of this drug include its weak action as a carbonic anhydrase inhibitor, its prophylactic effect against migraine and its side effect of appetite suppression (Keira *et al.,* 2016).
  - Octreotide is a somatostatin analogue mainly used to manage hormone-releasing tumours (Keira *et al.,* 2016).

- ✓ Surgical treatment:

Surgical procedures for idiopathic intracranial hypertension include:

- ❖ Optic nerve sheath fenestration (ONSF): Investigations into the safety and efficacy of ONSF have historically been small, uncontrolled retrospective studies, which often omitted statistical evaluation in favour of percentage stability or subjective patient assessments (Matthew *et al.,* 2010; Keira *et al.,* 2016; Biousse and Bousser, 2006; Bidot, 2016; Deborah and Daniel, 2004; Susan *et al.,* 2018).
- ❖ Spinal fluid diversion: CSF diversion can be performed by lumbo-peritoneal shunts or ventriculo-peritoneal and ventriculo-jugular shunts and a ventriculo-auricular shunt can sometimes be performed (Matthew *et al.,* 2010; Keira *et al.,* 2016; Biousse and Bousser, 2006; Bidot, 2016; Deborah and Daniel, 2004; Susan *et al.,* 2018). Only ventriculo-peritoneal shunting was performed in our series.
- ❖ Endovascular stenting for venous sinus: was first performed by Higgins and 118 results from their case series of 12 patients with refractory idiopathic intracranial hypertension showed improvements in venous pressure gradients after the procedure. Resolution of tinnitus, headache, visual function and papilledema (Keira *et al.,* 2016; Biousse and Bousser, 2006).
- ❖ Bariatric surgery: has proven to be very effective in achieving long-term weight loss.125 Laparoscopic gastric bypass (LGB) and laparoscopic adjustable gastric banding (LAGB) are the main procedures performed (Keira *et al.,* 2016; Deborah and Daniel, 2004).
- ❖ The choice of procedure for visual loss therefore depends on local availability.

Pronostic

- ✓ The visual prognosis is generally good in idiopathic intracranial hypertension, but risk of permanent vision loss exists for a subset of male patients.
- ✓ Surgical complications: Complications may include traumatic optic neuropathy, retinal vascular occlusion, pupil dilation and diplopia, shunt blockage, infection, abdominal and back pain, intracranial hypotension and tonsillar herniation, restenosis and vascular perforation have been reported as complications (Keira *et al.,* 2016; Deborah and Daniel, 2004).
- ✓ The clinical evolution of our patient was marked by an improvement of the headaches despite the persistence of visual disorders.

Conclusion

Idiopathic intracranial hypertension is suggested by an unusual headache and a combination of fundus, CSF pressure measurement and MRI. The increasing prevalence of the obese population is leading to an increase in the prevalence of IHD. The sometimes severe visual prognosis justifies optimising the management of idiopathic intracranial hypertension**.**

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