

Management of Idiopathic Intracranial Hypertension: Experience of a North African Center

Ibrahim Assoumane^{1,2} Mahdia Touati¹ Harissou Adamou³ Nadia Lagha¹ Ibrahim Baaré⁴
Samuila Sanoussi⁵ Abderahmanne Sidi Said¹ Abdelhalim Morsli¹

¹Department of Neurosurgery, CHU Bab El Oued, Algiers, Algeria

²Department of Neurosurgery, Maradi Reference Hospital, Niger Republic

³Department of General Surgery, Faculty of Health Sciences, University of Zinder, National Hospital, Zinder, Niger Republic

⁴Department of Ophthalmology, Maradi Reference Hospital, Niger Republic

⁵Department of Neurosurgery, Niamey National Hospital, Niger Republic

Address for correspondence Ibrahim Assoumane, MD, Maradi Reference Hospital, Niger Republic (e-mail: as_ibrah2006@yahoo.fr, assoubrahim18@gmail.com).

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Abstract

Background The idiopathic intracranial hypertension (IIH) is characterized by elevation of intracranial pressure (ICP) in the absence of intracerebral space occupation or venous sinus thrombosis. It usually occurs in obese young women but is very rare in childhood.

Materials and Methods We conducted a retrospective study in the neurosurgical department of Bab El Oued teaching hospital of Algiers over a period of 8 years from January 2008 to December 2015. We analyzed clinical data of 10 patients with IIH diagnosis, and the surgical technique consisted of an insertion of a lumbo-peritoneal shunt.

Results In our study, 80% were women with sex ratio M/F of 0.25; the mean age was 32 years and 60% of our patients were obese. For most of our patients, the post-operative outcome was marked by the disappearance of the headache immediately, disappearance of strabismus and diplopia in the following week, followed by improvement of visual function.

Conclusion The IIH is predominantly a disease of women in the childbearing age; surgical treatment is a good option for patients who resisted medication or did not tolerate it as well as for ICH fulminous in emergency cases.

Keywords

- ▶ idiopathic
- ▶ intracranial hypertension
- ▶ lumbo-peritoneal

Introduction

Idiopathic intracranial hypertension (IIH), was first described by Quincke in 1893 after the introduction of the lumbar puncture.^{1–5} It was also called pseudotumor cerebri by Nonne in 1904 or benign intracranial hypertension by Foley in 1955.^{1,2,4} The first series was reported by Dandy in 1937.^{2,6} The annual incidence of IIH is 1 to 3 per 100,000 in the general population. It is elevated to 20 per 10,000 among the female population of childbearing age.^{1,7} The etiology

of IIH is not completely understood; however, female sex, obesity, endocrine disorders, and high-levels of vitamin A are risk factors incriminated.^{4,8} The IIH is characterized by elevation of intracranial pressure (ICP) in the absence of intracerebral space occupation or venous sinus thrombosis. IIH leads to permanent vision loss and refractory headaches that had failed medical management.¹ It is a therapeutic emergency to preserve vision and reduce other symptoms.^{1,3,4} The aim of our study was to assess the clinical and therapeutic aspects of the idiopathic intracranial hypertension in our department.



Materials and Methods

We conducted a retrospective study in the neurosurgical department of Bab El Oued teaching hospital of Algiers over a period of 8 years from January 2008 to December 2015. We analyzed medical data of 10 patients presenting severe ocular signs associated to headaches not relieved by medical treatment and iterative lumbar punctures. The analysis included: the body mass index (BMI): below 18.5 = underweight, 18.5–24.9 = normal weight, above 25 Kg/m² = obesity, neurologic examination, visual function tests, neuroimaging studies (brain computed tomography [CT] scan, brain magnetic resonance imaging [MRI]), cerebrospinal fluid (CSF) studies (cytology and biochemistry), ICP measurement, and treatment and outcome. The surgical technique consisted of an insertion of a lumbo-peritoneal shunt. Under general anesthesia and lateral position, the first step is a median skin incision from L3 to L5, and lumbar puncture at L4 L5 level to collect CSF for cytology follow by the intradural insertion of a T catheter at that level. The second step is a paraumbilical skin incision, exposition of the peritoneum, tunneling of the lumbar catheter to join the paraumbilical incision, verification of the leakage of CSF, and introduction of the catheter inside the peritoneum. The end of the procedure is the closure of the two incisions (lumbar and paraumbilical). The median follow-up is 12 months.

Results

During our study, we registered 10 patients admitted for IIH. The mean age was 32 years (range: 9–48 years). There were eight women and two men with a sex ratio M/F of 0.25. Most of our patients were obese, 60% ($n = 6$), 20% ($n = 2$) are overweight, and only 20% ($n = 2$) had normal weight. The most frequent clinical signs were headaches and decrease of visual acuity, representing 100% for each followed by transient visual obscurations, diplopia, and strabismus with 50, 30, and 10%, respectively.

We summarize the socio-epidemiological, clinical, and paraclinical characteristics and outcomes of patients in **Table 1**.

The fundus examination objectified bilateral papilloedema in 80%, while bilateral optic atrophy was present in 20%. Visual field was constricted in seven cases which represent 70% but normal in just one case; the test was impossible in two cases (**Fig. 1**).

We performed brain MRI for all our patients: this investigation objectified empty sella in five patients (50%), tortuosity of the optic nerve in six patients (60%), protrusion of optic nerve papillae into vitreous in all the patients (100%), and flattening of the posterior sclera in two patients (20%) (**Fig. 2**).

We operated 10 patients under general anesthesia on lateral position and performed the insertion of a lumbo-peritoneal shunt for all of them. The postoperative outcome was good for the majority of patients (8 = 80%) because all of them experienced disappearance of the headache immediately, and disappearance of strabismus and diplopia

in the following week. The visual function improved for eight cases representing 80%, but there was bilateral optic atrophy, leading to blindness in two cases (20%). We noticed postoperative complications obstructing the peritoneal catheter (two cases), which represented 20% of our series at 12 months postoperation and the migration of the catheter in one case (10%) 3 weeks after surgery. We managed the complications by revising the shunt in two cases of obstruction and changed the whole shunt in the case of the intra-abdominal migration.

Discussion

The incidence of IIH is approximately 1 to 3 per 100,000 in the general population.^{4,9-11} The most reported risk factors are female sex and obesity. In our study, there were eight women and two men with a sex ratio M/F of 0.25, mean age of 32 years, and obesity representing 60%. Many authors reported especially high rates among young women suffering from obesity^{11,12}; Ali et al⁸ reported childbearing age in 99%. For Grant et al,⁹ there is no incidence of sex in a review of 79 cases in infancy and childhood. In our study, the most frequent clinical signs are headaches and decrease of visual acuity, as reported by many authors.^{1,4,12,13} The main neuroimaging findings that may accompany IIH are empty sella (70%), distension (45%), and tortuosity (40%) of the perioptic subarachnoid space, flattening of the posterior sclera (80%), and protrusion of optic nerve papillae into the vitreous and transverse sinus stenosis (90%).^{5,14,15}

IIH corresponds an increased ICP of unknown cause (thereby excluding secondary intracranial hypertension).^{4,7} In IIH, the CSF composition is normal.^{1-3,5,7,9,10,12-21} The pseudotumor name cerebri seems incorrectly understood, since it includes secondary causes of intracranial hypertension with the exception of lesions occupying space.^{3,4} It is more logical to call them pseudotumor cerebri primitive.⁴

Despite the number of pathogenesis theories that have been proposed, the cause of IIH is still unknown. Basic mechanisms that could explain a sustained intracranial hypertensive state in otherwise healthy subjects include: (1) increased CSF production rate, (2) cerebral edema, and (3) reduced CSF absorption.^{5,10} IIH is believed to result from irregularities of CSF production, distribution, and absorption; however, etiopathogenesis still remains unclear.^{4,10,21}

Diagnostic criteria for IIH was developed in 1937 by Dandy and modified in 2002 by Friedman and Jacobson²² who defined the disorder according to the following characteristics:

- ▶ Symptoms related to increased ICP with unknown etiology and no neurologic signs of focalization.
- ▶ Normal brain imaging.
- ▶ Elevated CSF pressure: > 20 mm Hg for nonobese patients and 25 mm Hg for obese patients.
- ▶ Normal composition of the CSF.
- ▶ No other etiology of ICH.^{1,13,15,21}

The advance of imaging has significantly modified the evaluation of patients with possible IIH.¹³

Table 1 The socio-epidemiological, clinical, paraclinical characteristics and outcomes of patients

No.	Age (y)	Sex	BMI	Clinical signs	Results of MRI	Complications	LOS
1	48	F	22.5	Headaches Decrease of VA TVO	Empty sella Tortusity ON Protrusion ON	Obstruction of the abdominal catheter	10
2	32	F	28.3	Headaches Decrease of VA TVO	Tortusity ON Protrusion ON FPS	None	10
3	9	F	31.4	Headaches Decrease of VA Diplopia	Empty sella Protrusion ON	None	8
4	15	F	32.7	Headaches Decrease of VA Diplopia	Empty sella Protrusion ON	None	10
5	26	M	27.8	Headaches Decrease of VA Strabismus	Tortusity ON Protrusion ON FPS	None	10
6	30	F	39.4	Headaches Decrease of VA TVO	Empty sella Protrusion ON	Migration of the catheter in the abdomen	10
7	34	F	35.8	Headaches Decrease of VA TVO	Tortusity ON Protrusion ON	Obstruction of the abdominal catheter	10
8	38	M	38.6	Headaches Decrease of VA	Tortusity ON Protrusion ON	None	10
9	40	F	23.1	Headaches Decrease of VA TVO	Tortusity ON Protrusion ON	None	10
10	28	F	37.7	Headaches Decrease of VA Diplopia	Empty sella Protrusion ON	None	10

Abbreviations: FPS, flattening of the posterior sclera; LOS, length of stay (days); N, normal weight; OB, obese; ON, optic nerve; OV, overweight; TVO, transient visual obscuration; VA, visual acuity.

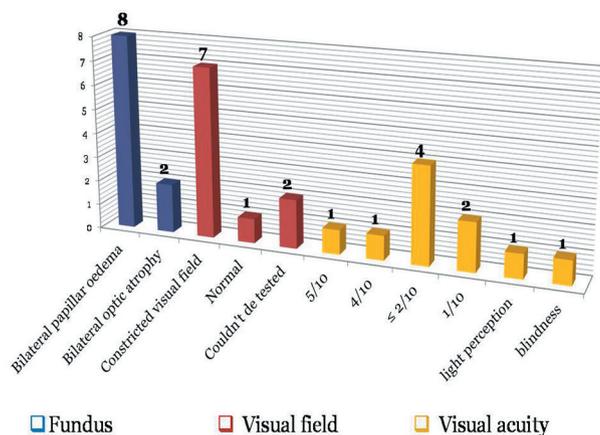


Fig. 1 Repartition of the patients by visual function test results.

The main goal of the treatment was to reduce ICP in order to avoid blindness by papillary edema in emergency. There exist many options for the treatment of IIH even if there are no precise guidelines for that nowadays. IIH does not currently have a well-established management algorithm, given the paucity of studies on the issue.^{1,3,8,17,19} Kalyvas et al¹⁵ in a systemic review of literature declare that treatment options range from conservative to interventional and surgical.

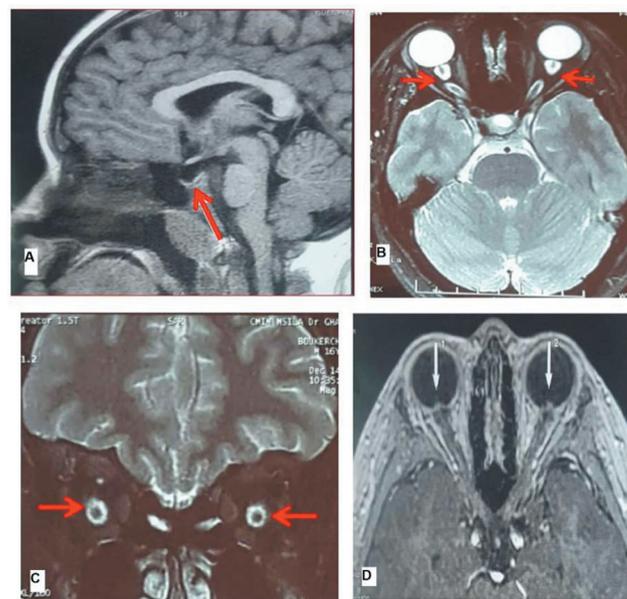


Fig. 2 (A) Empty sella (red arrow). (B) Optic nerve tortuosity (red arrow). (C) Distension of perioptic subarachnoid space. (D) Protrusion of optic nerve papillae into vitreous (arrow).

Conservative options include weight loss, drugs (acetazolamide, topiramate, furosemide, or octreotide), and serial lumbar punctures. Interventional and surgical options are

CSF diversion procedures (ventriculo-peritoneal or lumbo-peritoneal shunting), optic nerve sheath fenestration (ONSF), venous sinus stenting, and bariatric surgery.¹⁵ Headache disappears immediately after the lumbo-peritoneal shunt insertion for all our patients, but there was bilateral optic atrophy leading to blindness in two cases (20%). Moataz et al¹⁹ reported the improvement of headaches in 10 patients (45.5%) with complete resolution of papillary edema, gradually in 100% after 4 weeks. The most frequent complication in our series is the obstruction of the peritoneal catheter in 20% of our series followed by the migration of the catheter in the abdomen in 10%. Being an extra-cranial procedure, lumbo-peritoneal shunt insertion has a lower complication rate. Shunt obstruction and infections are the most common complications of this operation.¹⁸

Conclusion

IIH is characterized by elevated CSF pressure of unknown cause; it is predominantly a disease of women belonging to the childbearing age. It became clear that loss of visual function is common and patients might progress to blindness if untreated. Surgical treatment is allowed for patients who resisted medication or did not tolerate it as well as for ICH fulminant in emergency cases.

Conflict of Interest

None declared.

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