

Idiopathic intracranial hypertension: 33 observations during 5 years in the neurology department of the Mercy Hospital Metz- France and analysis of the literature

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Introduction

First described in 1897 by Heinrich Quincke as “serous meningitis”, HTICI remains a poorly understood pathology. Idiopathic intracranial hypertension (IIH) results from an isolated increase in cerebrospinal fluid (CSF) pressure of unknown etiology.^{1,2} The term “idiopathic” is imperfect because it classically includes forms without an obvious cause and patients in whom a triggering factor, especially medication, is suspected.² For this reason, the diagnosis is based on the 2013 modified Dandy criteria,⁴ which include: headache, nausea, vomiting, intracranial hypertension (CSF pressure greater than 250 mm water), normal biochemical analysis of lumbar puncture fluid, without clinical signs of focalization, with normal neuroimaging and bilateral papilledema. The majority of authors recommend the use of the revised diagnostic criteria suggesting the use of the generic term “pseudotumor cerebri syndrome” instead of “HTICI”.³ Its mechanism is not clearly known. Mostly of primary origin, some cases have been reported after treatments: Tetracycline, isotretinoin,⁵ corticosteroids.⁶ HTICI mainly affects young, overweight or obese women.¹ The dramatic increase in the prevalence of obesity worldwide has led to a doubling of the incidence of CTEI in recent years, from 2 per 100,000 people in the general population in the early 2000s to more than 4 per 100,000 currently in some countries.⁷ The risk of occurrence or recurrence of HTICI increases with the degree of obesity and with the existence of recent weight gain, even moderate (5-15% of body weight). Nevertheless, it is important to emphasize that the vast majority of young overweight women with headaches do not have HTICI. Benign essential headaches are common and the diagnosis of HTICI should not be made in the absence of definite papilledema.² CTEI without papilledema is very rare and is only diagnosed if strict criteria are met.³ When faced with a clinical picture suspicious of ICH, some complementary examinations have a key role in the positive and differential diagnosis: it is indeed urgent to eliminate a secondary ICH by obtaining an injected brain imaging. The assessment of visual function is essential, as it guides the initial management without replacing the analysis of the CSF. It is also imperative to look for a possible triggering or aggravating factor of idiopathic ICH, as well as factors of poor visual prognosis that will condition the delay of the rest of the management. Despite the often exhaustive workup, the etiologies and pathophysiological mechanisms of this disease remain unknown.

We report here the diagnostic, therapeutic and evolutionary management of cases of idiopathic intracranial hypertension followed in the Neurology Department at the CHR Metz.

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Materials and methods

In this retrospective study, we recorded for each patient an overweight and the use of a product that favored it. Imagery, fundus and cerebrospinal fluid were analyzed.

Results

Thirty-three patients met the diagnostic criteria, thirty-two women and one man. Twenty-five were overweight, all our patients had papilledema at the initial consultation. Thirty-one complained of headache, twenty-seven of visual disturbances, five cases of tinnitus and followed by three cases of diplopia and two cases of cranial nerve VI paralysis. It was also noted that four cases were taking favourable treatments (isoretinoid, penicillin, contraceptives).

Medical treatment was focused on oral acetazolamide in our patients and lumbar puncture for evacuation. Surgical treatment was performed in some patients, depending on the case: angioplasty of the venous sinus with stent placement (3%) and internal cerebrospinal fluid bypass (6%). And 6% of patients benefited from bariatric surgery. Follow-up with a dietician, endocrinologist and psychologist was requested for some patients. Improvement or stabilization of visual function and headaches were obtained for more than 80% of patients at 6 months after diagnosis.

Discussion

In our series of studies we note a female predominance in women of childbearing age. Obesity plays a major role in the condition as 70% of women with HTICI are obese. The reasons for consultation are headaches and visual disorders. The diagnosis is based on a combination of clinical and radiological findings and analysis of the lumbar puncture with measurement of the CSF opening pressure. It should be noted that each step contributes to the diagnosis of HTICI. The multidisciplinary nature of the diagnostic management of HTICI

in an emergency context requires a codified reference system to rapidly institute the treatment best adapted to the severity of the optic neuropathy. The clinical triad (papilledema, headache, visual signs) associated with the terrain (young obese woman) confirms the data in the literature. The treatment is based on the intake of Diamox and Epiteo usually associated with a balanced diet. A weight loss of 3.5% of BMI can cause a significant decrease in intracranial pressure. The absence of any of these symptoms should lead to a differential diagnosis.

Conclusion

Through the study of our 33 series of observations and analysis of the literature, we have tried to achieve a general approach to HICI. It remains a pathology involving a multidisciplinary management associating ophthalmologist, radiologist, neurologist. The rapidity of the diagnostic and therapeutic strategy allows to improve or stabilize the great majority of patients. Finally, the therapeutic management remains very much debated, in particular surgical intervention or interventional neuroradiology.

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None.

Conflicts of interest

The authors declare no conflict of interest.

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