

Case Reports & Case Series

Is the adjustable lumbo-peritoneal shunt a valid option for managing symptomatic idiopathic intracranial hypertension? A case report of a multidisciplinary management

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ARTICLE INFO

Keywords:

Lumbo-peritoneal shunt
Idiopathic intracranial hypertension
Pseudotumor cerebri

ABSTRACT

Background: Idiopathic intracranial hypertension is a rare disorder, characterized by raised intracranial pressure with no recognizable cause. Headache, decreased visual acuity and papilledema are the main clinical manifestations.

Case description: A case of a 27-year-old obese woman affected by idiopathic intracranial hypertension is reported. She was initially treated conservatively, but, due to severe headache refractory to medical therapy, an adjustable lumbo-peritoneal shunt was implanted.

Conclusion: Gold standard treatment of idiopathic intracranial hypertension is not defined yet, but the lumbo-peritoneal shunt could represent a valid option for managing the disease in properly selected patients.

1. Introduction

Idiopathic Intracranial Hypertension (IIH), also known as pseudotumor cerebri, is characterized by an increase in intracranial pressure with no specific etiology such as intracranial mass, hydrocephalus, infection, or hypertensive encephalopathy [1,2].

The incidence peaks among obese female patients [1]. The pathogenesis is likely due to multiple causative factors determining a dysregulation in the cerebro-spinal fluid (CSF). Obesity, being one of the main risk factor, is also argued to eventually determine an altered CSF homeostasis through different mechanisms [1].

The most common presenting symptom is headache, followed by ophthalmic-related symptoms such as transient visual obscuration, reduced visual acuity, diplopia and complete visual loss. In the past, the presence of these symptoms with the absence of other neurological signs, the increase in CSF pressure and the demonstration of normal ventricles were diagnostic (Dandy Criteria). Nowadays, instead, the diagnosis is based on more quantifiable criteria, such as the identification of papilledema with the use of OCT, and the neuroimaging [2]. The presence of venous sinuses stenosis, and an empty sella, are specific

and sensitive imaging findings in IIH patients [1,2]. The high diagnostic yield of OCT in recognizing papilledema and an objective evaluation of the improvement of intracranial pressure, followed by Retinal Nerve Fiber Layers (RNFL) thinning after medical or surgical therapy, are also reported [1,3].

Different management strategies are suggested but there is no consensus yet on which is the optimal. Preservation of visual function, long-term control of headache, papilledema and restoration of CSF pressure are considered the main aims of treatment [4]. Behavioral changes (weight loss) and medical therapy (acetazolamide) should be the first line approach. If symptoms are not controlled, lumbo-peritoneal (LP), ventricular-peritoneal shunting (VP), venous sinus stenting and Optic Nerve Sheath Decompression are potential interventional options [4].

2. Case report

A 27-year-old obese (type I, BMI 30.1) woman was admitted to the Emergency Department because of an uncontrolled headache, a rapid decrease of visual acuity and diplopia. Her history was characterized by

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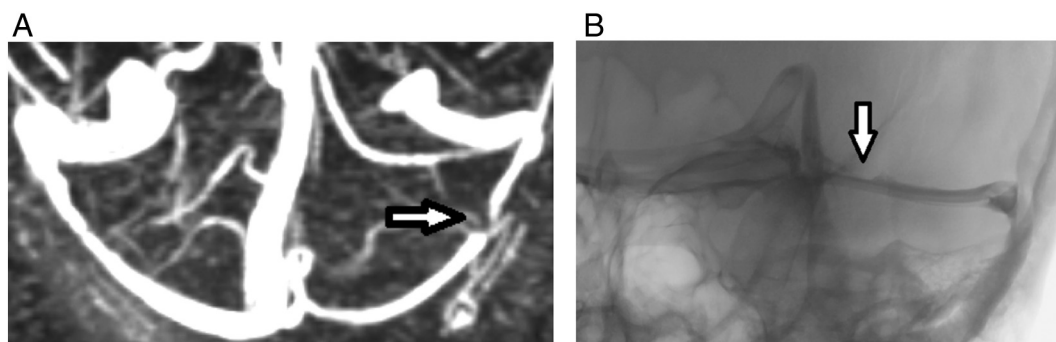


Fig. 1. Venous angiograms.

A. Shows the angio-MRI, with a narrowing of in calibre of the left transverse sinus (arrow) is found.

B. Reports the angiography scan that seems to confirm the narrowing of the sinus (arrows). However, the venous sinus stenting was not applied due to the absence of pressure gradient.

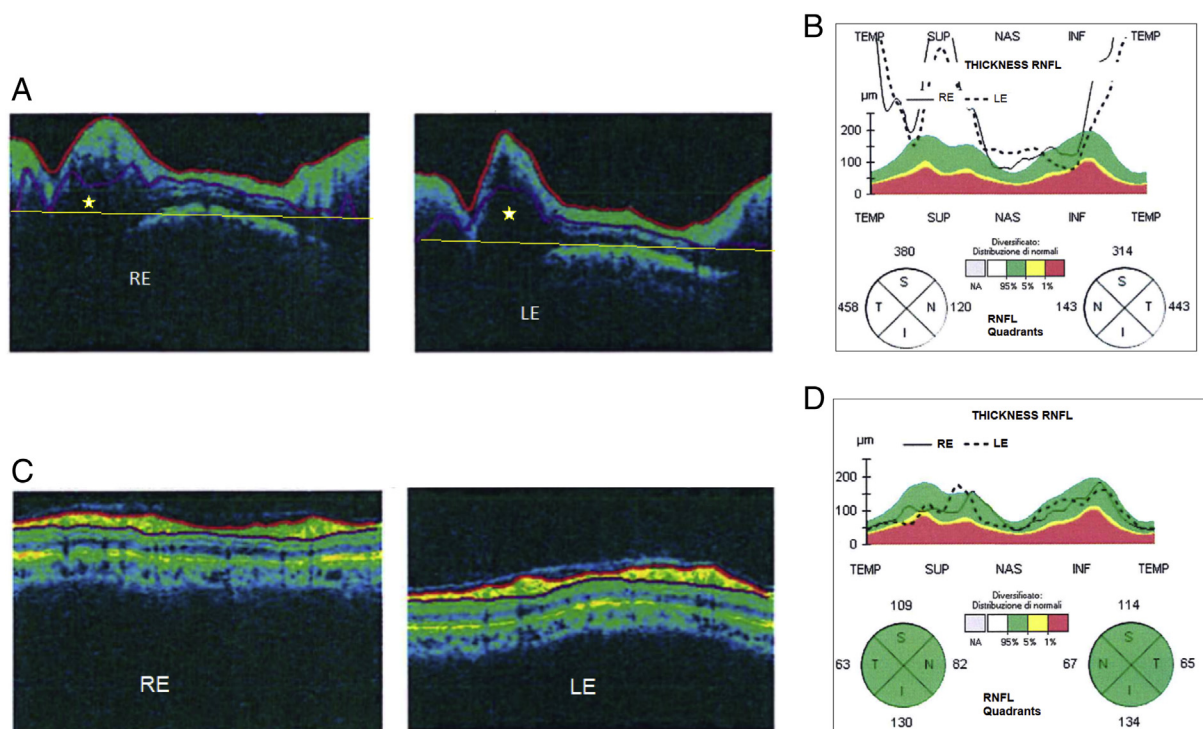


Fig. 2. Quantification of disc swelling using optical coherence tomography (OCT) before [A–B] and after [C–D] the procedure.

A. Circular OCT of the right eye (RE) and left eye (LE).

It is possible to appreciate the presence of subretinal hyporeflective space corresponding to an accumulation of oedema (star) and the detachment of the inner limiting membrane (ILM) (red line) from the base line (yellow line).

B. The papilledema is demonstrated by the retinal nerve fiber layer (RNFL) thickness, which exceeded the normative database values in all quadrants tested.

C. Circular OCT of the right eye (RE) and left eye (LE). A clear improvement of the papilledema was recognizable, as the retinal thickness was turned to normal and no subretinal hyporeflective space in peripapillary region was evident, beside the presence of bilateral retinal microcystic changes.

D. In the graphical representation is possible to appreciate the re-establishment of the normal thickness of RNFL. The RNFL measurements were in the green area for all the quadrants indicating normal RNFL thickness. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

familiar hypertension and migraine. She referred a weight gain of 10 kg over the last year. Physical examination was otherwise negative for pathological findings. Neurological examination revealed a reduced visual acuity and an altered afferent pupillary defect on the left eye.

Visual evoked potentials showed a demyelinated pattern of the optic nerve: a treatment with intravenous methylprednisolone was set up and resolution of diplopia were obtained without improvement of the visual acuity.

A brain MRI was then performed, ruling out focal lesions or enlarged ventricles. The patient was referred to the Neuro-Ophthalmology Unit with a diagnosis of Bilateral Papilledema (Frisén Classification,

class 4). A lumbar puncture showed a normal CSF composition and an opening pressure of 178 mmH₂O. A treatment with acetazolamide and topiramate was started. Due to persistent holocranic headache, resulting refractory to medical therapy, a brain angioMRI was performed. A stenosis of the left sigmoid sinus [Fig. 1] was found. A stenting procedure of the sigmoid sinus was attempted via angiography, unsuccessfully. Given the persistence of symptoms, an adjustable lumboperitoneal shunt (Strata NSC valve, Medtronic, Inc.; calibrated at the performance level 1.5, corresponding to an activity pressure range 90–110 ± 25 mmH₂O) was implanted [5]. The patient early reported a marked improvement of the symptomatology. The follow-up OCT

evidenced a decrease of the papilledema and visual field improvement [Fig. 2].

3. Discussion

The management of IIH is controversial, with indications for interventional treatments not fully cleared. It is therefore needed to identify clinical and instrumental findings that can help in the selection of the most appropriate treatment according to the patient pathological characteristics.

A multidisciplinary approach has a crucial role in the decision making. An ophthalmologist objectively confirmed the papilledema by OCT. Brain MR imaging showed no hydrocephalus or other pathology. An endocrinological work-up then ruled out other endocrinological disturbances, which could mimic IIH and help in the life-style modification. With symptoms refractory to medical therapies, the failure of the endovascular sinus stenting and the objective assessment of worsening papilledema, a surgical option was considered in this case. An adjustable LP shunt was chosen as the most effective and rapidly acting solution.

CSF shunting is the most common surgical approach for the treatment of IIH, with no superiority of LP or VP yet established [1,4,5]. We employed a LP shunt due to the minimally invasive nature of the procedure. The LP approach avoids major intracranial risk, such as haemorrhages, seizures, infections. IIH patients, different from those with hydrocephalus, have slim ventricles; this anatomical feature can cause difficulties in cannulating with a ventricular catheter, increasing the surgical risks. The conventional LP shunt is valveless, leading to one of the reported major limit, that is the overdrainage [4]. In 1967 a valve was incorporated reducing the likelihood of overdrainage. We opted for LP shunt with Strata NSC adjustable valve, that allows the calibration of the performances and the avoidance of the backflow of the CSF [4]. Despite that the patient was obese, as most of the patients with IIH are as previously mentioned [1], the placement of the lumbar and peritoneal catheter as well as the positioning of the valve on the flank occurred eventless; in particular, while working on the abdomen, large retractors were used to grant optimal control of the peritoneal fascia. Two suture stitches, one at the peritoneal and one at the muscular fascia, were placed to secure the catheter in place and to reduce the risk of displacement over time [6]. The patient herein described did not fall into the category of obesity, i.e. BMI > 35 kg/m², where medical or surgical management with bariatric surgery per se can be advocated as a first-tier therapy for signs and symptoms due to IIH [7,8], where the placement of a LP shunt can be technically problematic and more prone to failure over time [6,8,9]. The patient had a favourable outcome from

the surgery and, since the improvement of the headache and the decrease of papilledema (RNFL measured with the OCT), she could return to her normal life. The full control of symptomatology was guaranteed for 12.55 months when, due to the onset of headache refractory to analgesic drugs, an adjustment at performance level 1.0 (pressure range 35–55 ± 25 mmH₂O) was needed and lead to a complete regression of the symptomatology.

4. Conclusion

In this case, we reported the outcome of a young woman affected by IIH treated with an adjustable LP shunt. Considering the technical advantages, the optimal symptom control with no daily drugs need, the simple adjustability in response to patient's needs and the fast return to normal life, this study further proved the potential of adjustable LP shunt as a valid option in the management of properly selected subjects affected by IIH. This finding, along with previous results, can thus serve as foundation for needed confirmatory larger trials to establish the best approach in the multidisciplinary management of IIH.

Declaration of Competing Interest

No conflict of interest applies.

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