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Comparison of Lumbarperitoneal Shunt with the Stereotactic Ventriculoperitoneal Shunt for the Treatment of Refractory Idiopathic Intracranial Hypertension：A Case Report and Literature Review

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Case report

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Abstract

Background

Idiopathic intracranial hypertension (IIH) or pseudotumor cerebri (PTC), is a disorder of young obese females and characterized by headache, papilledema with raised intracranial pressure in the absence of known pathological cause. Due to the uncertainty of etiology, it may lead to misdiagnosis and poor outcome. IIH is typically treated with shunts (lumbarperitoneal and ventriculoperitoneal shunting), but shunts are prone to malfunction and infection, resulting in many patients experiencing recurrent headaches post-treatment.

Case presentation

We report a case of 41 years old obese female (BMI:30.9) with IIH who exhibited a history of hypertension (BP:150/100mmHg) and documented elevated intracranial pressure (OP:450mm H_2O). After the failure of several medical treatments, the patient was offered LPS due to vision loss and headache, but postoperative symptoms (intermittent headache-mainly whole craniocerebral prickling pain accompanied by neck and shoulder pain) did not significantly relieve lasting for 11 years. Therefore, we considered the blockage of the primary shunt tube in the patient, and the patient with a small ventricle leads to some difficulty in ventricle puncture, then we had to treated with the stereotactic VPS□VPS□for her exacerbation symptoms. More surprisingly, the hypertension was within the normal range (<115/80mmHg) after the surgery (without antihypertensive medication).

Purpose

To compare the surgical therapeutic effects and complications of lumbarperitoneal shunt (LPS) and the stereotactic ventriculoperitoneal shunt (SVPS) for idiopathic intracranial hypertension.

Conclusion

CSF diversion with a ventriculoperitoneal shunt (VPS) or lumboperitoneal shunt (LPS) is usually performed when the main symptom is vision loss and it also stabilizes headache and papilledema. LPS significantly alleviates symptoms in the short term, but due to excessive shunt of LPS long time, it is easy to be complicated with iatrogenic Chiari malformation and slit ventricle syndrome. Therefore, we are encouraged to apply the SVPS on our patients for the favorable long-term outcome.

Background

Idiopathic intracranial hypertension (IIH) is a disorder characterized by elevated intracranial pressure, can lead to symptoms such as severe headache, papilledema, and vision loss, in the absence of an identifiable cause. The incidence of IIH ranges from 1 to 3 patients per 100,000 people per year, and predominantly affects young adult women under 45 year of age [1].

The initial treatment of choice for IIH is conservative, including weight loss and conservative treatment. Advanced cases of severe visual loss and/or progressive IIH may not respond to conservative treatment. Such advanced cases occur in 10% to 20% of patients with IIH and may progress to permanent blindness through optic nerve atrophy at highly variable rates (weeks to years) if left untreated. Therefore, surgery is recommended in the case of advanced IIH cases [2]. Common surgical treatments for idiopathic intracranial hypertension include LPS and VPS [3]. Because the ventricles are not enlarged, the LPS is usually preferred over the VPS [4]. However, due to excessive shunt of LPS long term, it is easy to be complicated with iatrogenic Chiari malformation and the slit ventricle syndrome. In addition, the SVPS was noted to have a higher medium-term patency rate than lumboperitoneal shunt [5]. Therefore, we compare the surgical therapeutic effects and complications of LPS with the stereotactic VPS for the treatment of refractory IIH.

Case Presentation

Eleven years ago, a 41 years old obese woman (BMI 30.9) with IIH who exhibited a history of hypertension (BP:150/100mmHg) and documented elevated intracranial pressure (OP:450mm H_2O). After the failure of several medical treatments, then the patient was presented to the neurosurgery department for intermittent binocular vision loss and amaurosis lasting for one year. The condition was further aggravated and accompanied by intermittent headache.

At admission, the patient was fully conscious, and hemodynamically stable. On examination, her visual acuity was 90/100 in her right eye and 20/100 in the left eye. Neurological examination revealed bilateral abducens nerve palsy, optic fundi showed severe papilledema (grade 4) with unclear boundaries (Fig.1).

In view of the possibility of an intracranial mass lesion, cranial MRI was performed which evidenced no structural alterations (Fig.2A, B). Non contrast MRI of brain and cerebrospinal fluid (CSF) analysis were normal. Repeated lumbar puncture showed pressure >330mmH_2O. Other cranial nerves examination showed no abnormalities. These findings were consistent with idiopathic intracranial hypertension.

The patient was prepared for surgery and underwent lumbarperitoneal shunting on the 18th day of her referral, with improved postoperative vision and reduced optic papillomedema. She was discharged with a diagnosis of idiopathic intracranial hypertension. During follow up, her vision was improved significantly in both eyes.

However, during the 11 years following LPS, the patient was admitted to the hospital several times for intermittent headache—mainly whole craniocerebral prickling pain accompanied by neck and shoulder pain, with no significant relief with painkillers, and the cerebrospinal fluid pressure was measured by multiple lumbar puncture, and the overall pressure showed an upward trend, the last pressure was 350mmH_2O. According to the lumbar puncture pressure of the patient, we considered the blockage of the primary shunt tube in the patient, and due to the small ventricle of the patient (Fig.3A) leads to some difficulty in ventricle puncture, and MRI brain showed iatrogenic Chiari malformation on T1 image

(Fig.3B). Therefore, we decided to perform VPS in stereotaxic manner(preoperative BP:>150/100mmHg). She experienced instant relief (the headache was significantly relieved) after the surgery, and documented intracranial pressure (OP:140mm H_2O). CT and MRI of the brain are shown after surgery (Fig.4A, B and C). More surprisingly, the patient's hypertension was within the normal range (<115/80mmHg) by multiple measurements over several days after the surgery (without antihypertensive medication). Finally, she was discharged with a diagnosis of refractory IIH.

Discussion

Idiopathic intracranial hypertension (IIH) is a rare condition that causes elevated intracranial pressures of unknown etiology [6].The incidence of IIH ranges from 1 to 3 patients per 100,000 people per year [1],and primarily affects obese women as approximately 90% of those diagnosed with IIH are obese female [6].It is unclear exactly why obesity is so closely related to the development of IIH, however, it is thought that the increased intra-abdominal pressure could lead to an increase in the filling pressure of the right heart, resulting in increased venous pressure [7].

Typically, the three symptoms representative of IIH include daily severe headache, papilledema, and vision loss [8].Papilledema is generally considered to be the primary symptom of IIH and can lead to severe morbidity if left untreated [9],and advanced cases occur in 10% to 20% of patients with IIH and may progress to permanent blindness through optic nerve atrophy at highly variable rates (weeks to years)[8].Although headaches are the most reported symptom, visual loss remains the most feared complication [10].

The exact cause of IIH is unknown, but there have been several proposed mechanisms attempting to explain its occurrence, such as CSF hypersecretion, normal CSF access blocked, CSF absorption disorder, venous sinus thrombosis and some unknown reasons. IIH is predominately a diagnosis of exclusion [11] (see Table 1 for the modified Dandy diagnostic criteria).

For patients with simple vision loss, medication may be effective for a short period of time, but long-term use may lead to gradual vision loss and even the risk of blindness. Therefore, surgical treatment should be performed for patients who fail to respond to conservative treatment. Dandy [12] believes that increased cerebral blood flow or cerebrospinal fluid volume leads to increased intracranial pressure, which leads to symptoms related to intracranial hypertension and optic papillomedema, leading to progressive vision loss and blindness. Therefore, IIH is typically treated with shunts, and both LPS and VPS can reduce intracranial pressure and have a certain effect on visual impairment by shunting CSF to reduce compression of the arteries and veins around the optic nerve. They are safe, effective and has a good short -term effect for LP and VP shunts in the treatment of IIH-induced headache and papilledema, as well as the improvement of visual acuity loss and/or visual field defects.

Complications of LPS surgery mainly include decreased intracranial pressure caused by excessive shunt, blockage of shunt tube, infection, and cerebrospinal fluid leakage [13]. In addition, due to excessive shunt

of LPS, it is easy to be complicated with iatrogenic Chiari malformation and slit ventricle syndrome. Moreover, several reports show that the LPS technique is associated with a 60% rate of revision [14,15], a 4% rate of iatrogenic Chiari malformation [16], and an 18.5% rate of iatrogenic radicular pain [15].Therefore, in order to avoid the formation of iatrogenic Chiari malformation and slit ventricle syndrome, the postoperative CSF pressure of LPS was higher than VPS, so VPS has better effect on headache and vision recovery in patients with idiopathic intracranial hypertension than LPS. Moreover, complications following LPS have been reported [17] in 18% to 85% of cases, and the need for multiple revision surgeries and frequent wound complications have prompted many to abandon this procedure for the treatment of idiopathic intracranial hypertension, in favor of VPS. However, shunt tube blockage is a common complication of VPS. Ventricular end obstruction is mainly caused by excessive cerebrospinal fluid shunt, smaller ventricle, and the ventricle end of the shunt tube is buried in the brain tissue. It is possible that shunt failure rates may be similar between LP and VP shunts, but reasons for revision, removal, or replacement may differ. Therefore, It is necessary to find a solution to the blockage of the shunt tube—the SVPS technology.

Nowadays, as rates of obesity rise globally, IIH is expected to become increasingly common [18]. CSF shunting currently remains the primary treatment for acute reduction of symptom severity. The effectiveness and safety of VPS in the presence of IIH has been greatly increased with the advent of image guidance. Image guidance allows the procedure to be performed safely and accurately, and some series use image guidance to achieve 100% placement of a VPS with only one pass of the catheter needed on each patient [19].And the SVPS is noted to have a higher medium-term patency rate than LPS [5].Consequently, it might reduce the rate of shunt revision, particularly those due to proximal obstructions [20].More surprisingly, the associated concomitant symptoms/triggers of IIH are addressed, such as hypertension. So, we need to further investigate the relationship between hypertension and IIH in hypertensive IIH patients.

Conclusion

CSF diversion with a VPS or LPS is usually performed when the main symptom is vision loss and it also stabilizes headache, papilledema. LPS significantly alleviates symptoms in the short term, but due to excessive shunt of LPS long term, it is easy to be complicated with iatrogenic Chiari malformation [16]and slit ventricle syndrome. In addition, we believe the SVPS technique to be the first surgical option for our patients because of its relative simplicity, minimal invasiveness, easily managed morbidities, and long-term efficacy when compared with more invasive surgical procedures. Therefore, we are encouraged to apply the SVPS on our patients for the favorable long-term outcome.

Abbreviations

IIH Idiopathic Intracranial Hypertension

LPS Lumbarperitoneal shunt

VPS Ventriculoperitoneal shunt

SVPS Stereotactic Ventriculoperitoneal shunt

CSF Cerebrospinal fluid

CT Computer tomography

MRI Magnetic resonance imaging

Declarations

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Not applicable.

Authors' contributions

CG is the lead author of this case report. HZ and JF reviewed and supervised the writing of CG. Both have no conflict of interest. The concerned patient gave permission to publish this article and gave the authors admission to her hospital files. The authors did not receive funding for writing or publishing this article. All authors read and approved the final manuscript.

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Ethics approval and consent to participate

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Consent for publication

The patient provided verbal and written consent for this case report.

Competing interests

The authors declare that they have no competing interests.

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Table

Table 1: Modified Dandy Criteria for the diagnosis of IIH.

Modified Dandy Criteria
1.Signs and symptoms of increased ICP
2.No localizing neurologic finding (except possible 6th nerve palsy)
3.Normal CT/MRI findings
4.Increased CSF opening pressure (>200 mmH2O in non-obese patients, >250 mmH2O in obese patients),but normal CSF composition
5.No other identifiable cause of increased ICP

Figures

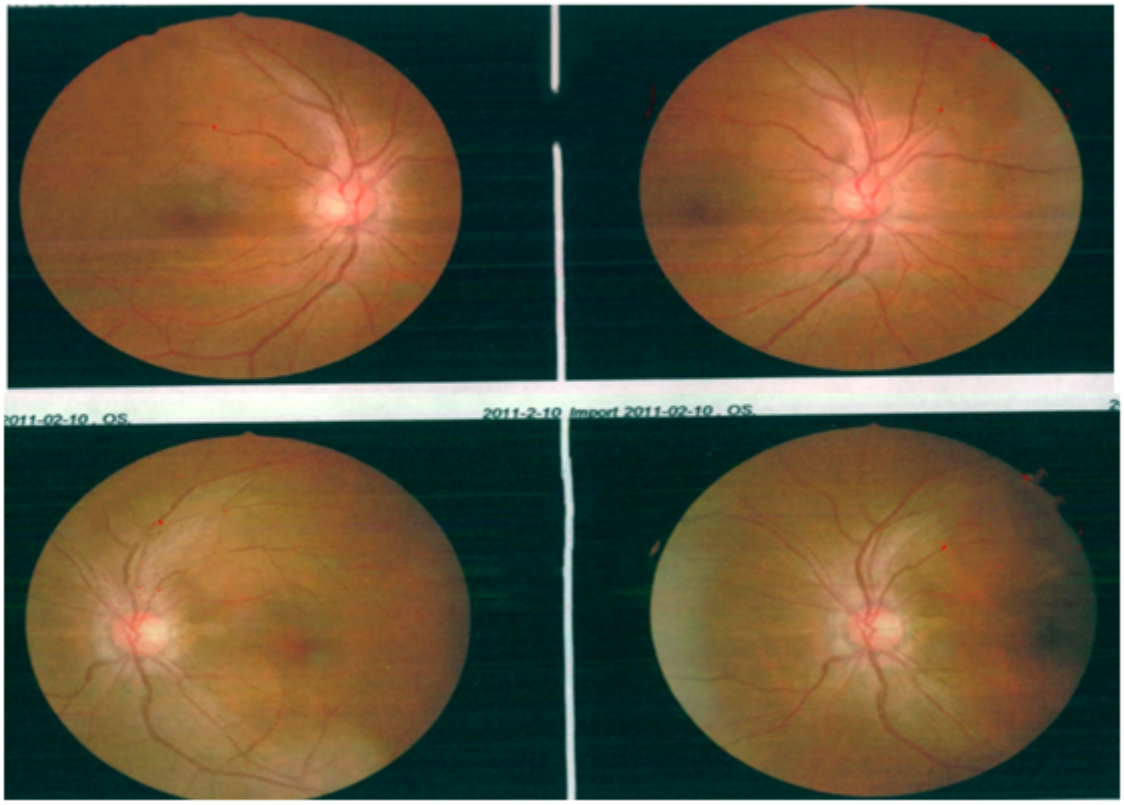

Figure 1

Bilateral optic fundi showed papilledema

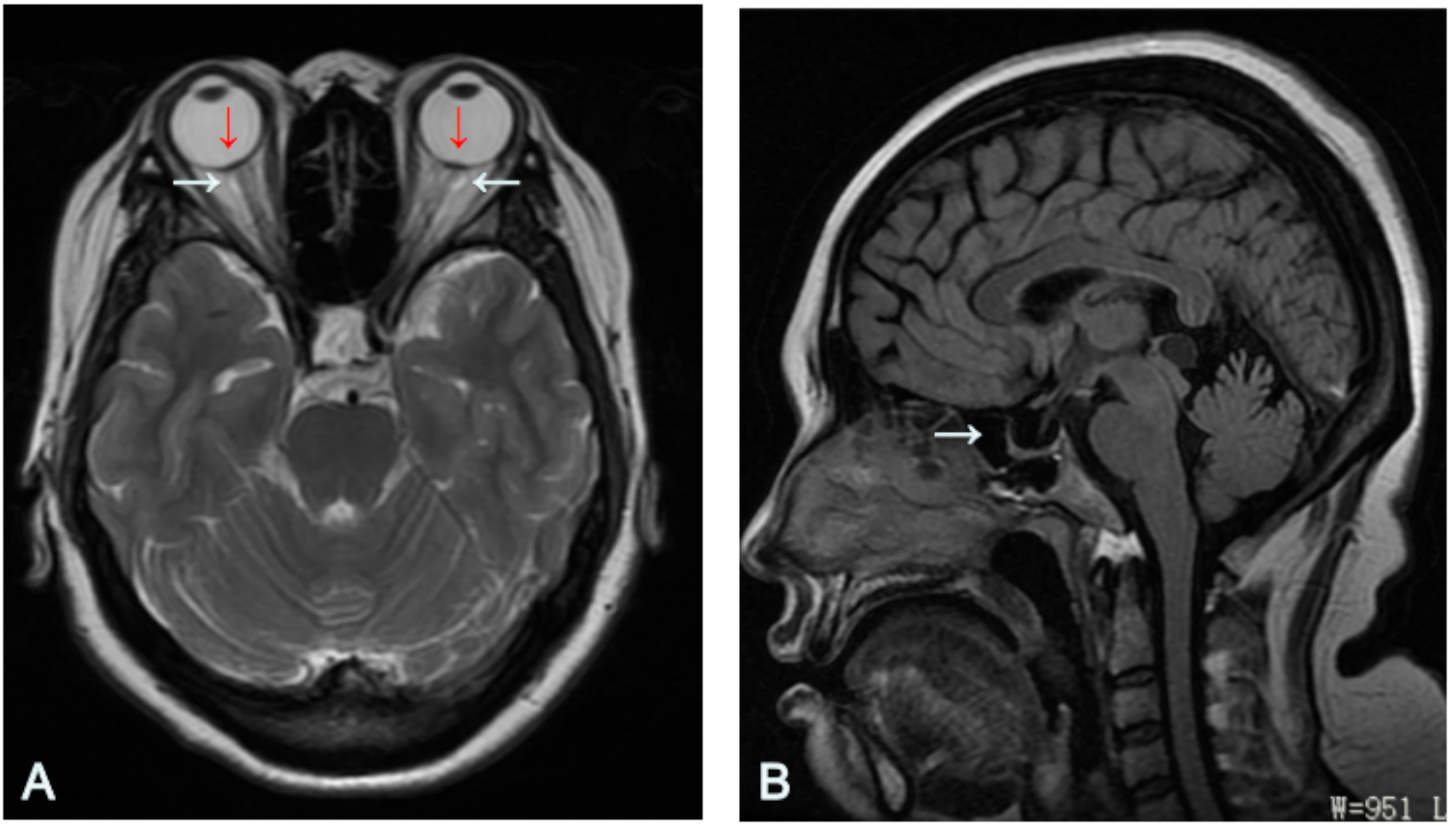

Figure 2

A Axial T2 weighted image showing increased transverse dimension of bilateral optic nerve/sheath complexes (white arrows) and bilateral optic nerve papillae protrusion into the vitreous space of the globes with flattening of the bilateral posterior sclera (red arrows). B Sagittal T1 image showing empty sella (white arrow).

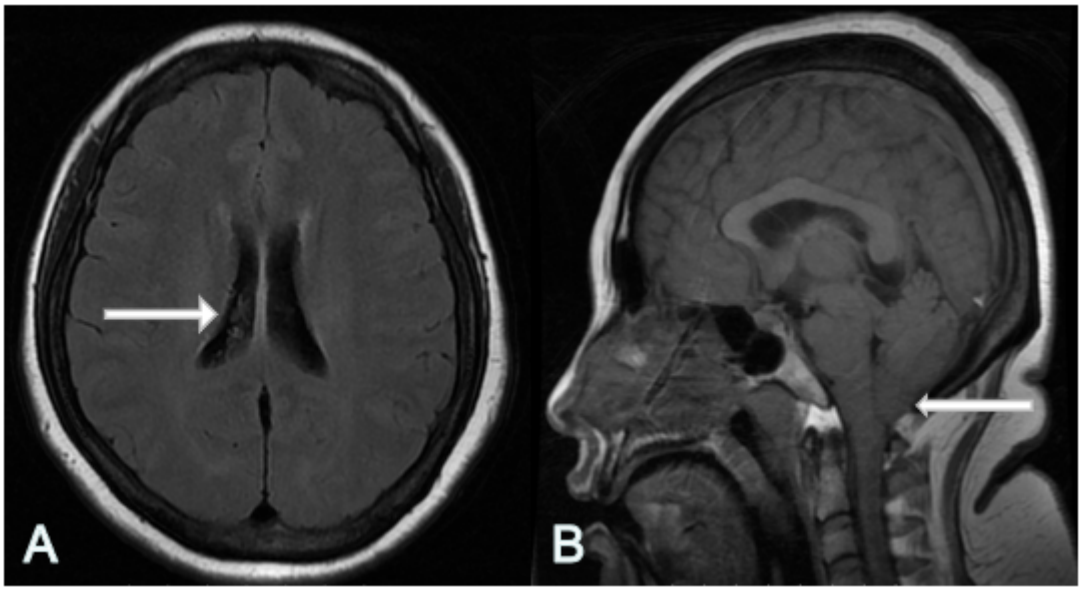

Figure 3

A Axial T1 image showing normal volume of lateral ventricle (white arrow). B Sagittal T1 image showing iatrogenic Chiari malformation (white arrow).

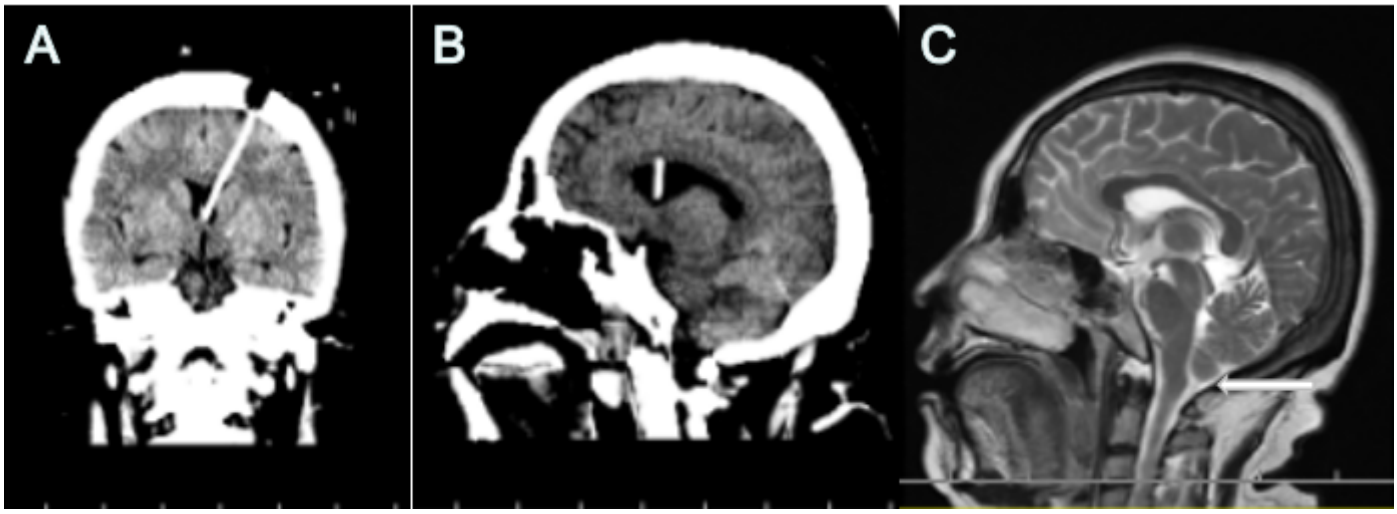

Figure 4

A. Coronal CT image showing the ventricular shunt tube was accurately located in the ventricle; B Sagittal CT image showing the ventricular shunt tube after the stereotactic VPS; C Sagittal T2 image showing the iatrogenic Chiari malformation was slightly relieved (white arrow).