

Case Report

Attachment of a pressure gradient chamber to existing lumbar theco-peritoneal shunt in a patient with pseudotumor cerebri with low pressure headaches, a novel procedure: a case report

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ABSTRACT

Pseudotumor cerebri is a syndrome which causes intracranial hypertension with no associated mass lesion. It is managed both medically as well as surgically. Cerebral spinal fluid diversion using theco-peritoneal shunt is commonly performed to decrease intracranial tension. We present a case of a middle age lady who had severe low pressure headaches following theco-peritoneal shunt for pseudotumor cerebri. She was managed by attaching pressure gradient chamber to the pre-existing theco-peritoneal shunt. Patient had marked improvement in headache, which gradually subsided. Attaching pressure gradient chamber to the pre-existing theco-peritoneal shunt helps to improve low pressures headaches in pseudotumor cerebri. It is an alternative procedure considering other extensive surgical options.

Keywords: Idiopathic intracranial hypertension, Programmable chamber, Pressure gradient chamber, Pseudotumor cerebri, Theco-peritoneal shunt

INTRODUCTION

Quincke, a German physician in 1890 described pseudotumor cerebri (PTC) which is also known as idiopathic intracranial hypertension.¹ PTC is rare with incidence of 1 to 5 cases per 1 lakh people in the general population.^{2,3} Clinically they present with varying degrees of headache, vomiting and visual disturbances. Fundoscopy reveals papilledema suggesting high intracranial pressure (ICP) and is confirmed by lumbar or ventricular puncture. The pathophysiology is unclear and mostly seen in middle-aged obese women. Management aims to rule out other causes of raised ICP. Improvement in headache and vision following CSF drainage helps to consider CSF diversion procedure for candidates who do not improve with medical measures.

CASE REPORT

A 39-year-old obese lady presented to us with headache for 1½ years, intermittent vomiting for 1 year and progressive blurring of vision in both eyes for 6 months duration. On examination she had visual acuity of 6/9 in both the eyes and bilateral papilledema (right>left). Computed tomography, magnetic resonance imaging (MRI) of brain along with MR venogram, (Figure 1) revealed normal ventricular size and ruled out intracranial space occupying lesion. MR venogram was normal. Lumbar cerebrospinal fluid (CSF) pressure was high (29 cm of water). She was diagnosed to have pseudotumor cerebri which did not improve with medical measures. In view of her persisting symptoms, she underwent lumbar theco-peritoneal shunt (valve-less or no pressure gradient) (Figure 2). Post operatively she developed

severe low-pressure headaches. To regulate the CSF flow, a high-pressure gradient chamber (Figure 3) was additionally connected to existing lumbar theco-peritoneal shunt (TP Shunt). The patient improved following the procedure and on follow up over a year, her vision marginally improved with no headache and vomiting.

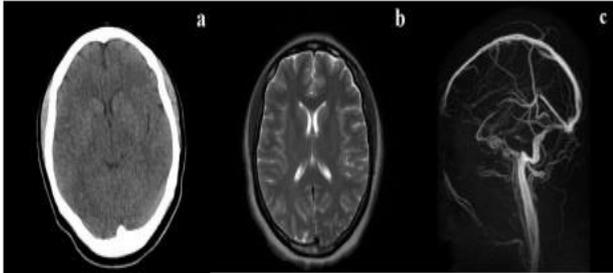


Figure 1: Plain CT, MRI brain axial section (Fig 1a & 1b) showing normal size ventricles and normal MR venogram (Fig 1c).

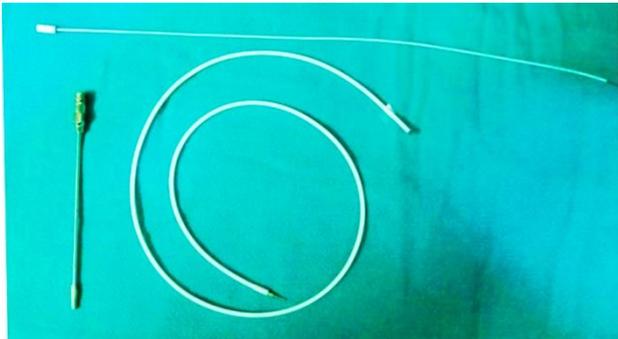


Figure 2: Theco-peirtoneal shunt (valve-less or no pressure gradient).

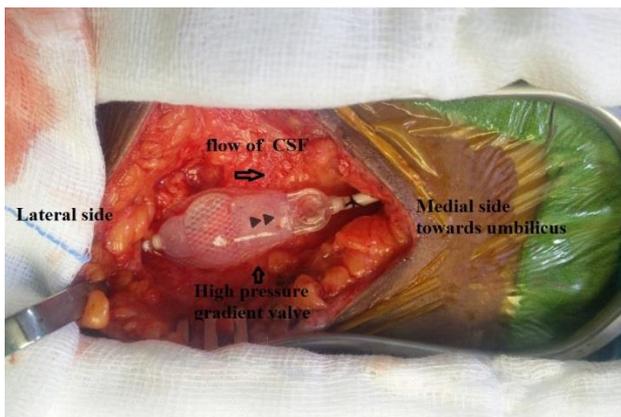


Figure 3: Abdominal transverse incision with attachment of high pressure gradient VP shunt valve to the existing theco-peritoneal shunt.

DISCUSSION

Patients with severe headaches associated with visual disturbance are often treated by various surgical options

which include optic nerve sheath fenestration and CSF diversion procedures namely theco-peritoneal shunt or ventriculo-peritoneal shunt. Theco-peritoneal shunting has been the main stay of treatment for pseudotumor cerebri in patients not responding to medical measures.^{4,5} Ventriculo-peritoneal shunt (VP Shunt) also an option however, it carries risk of intracranial bleed and difficulty in negotiating ventricular catheter in narrow ventricles.⁶ The main advantage of TP shunt is that it is extracranial however, it is also associated with complications like over or under drainage and shunt related complications requiring shunt revision.^{7,8}

In the present case, patient developed low pressure severe headaches following TP shunt which did not respond to conservative treatment. The conventional theco-peritoneal shunt are valve-less and had the major disadvantage of adjusting the CSF flow.^{9,10} Patients with these shunts frequently experienced over-drainage symptoms.

In our part of the world, pressure gradient theco-peritoneal shunts are not easily available, so we had the option of either remove/knot, the insitu theco-peritoneal shunt and replace with pressure gradient ventriculo-peritoneal shunt accepting the risks involved in the procedure. According to Wang et al.¹⁰ Introduction of valve reservoir have significantly decreased the incidence over-drainage and related complications. After their introduction, programmable TP shunt has been widely used for pseudotumor cerebri and also for other conditions like intraventricular hemorrhage and subarachnoid hemorrhage.^{12,13}

Considering risk of insertion of a fresh pressure gradient VP shunt with or without removal of in situ TP shunt, we opted to attach high pressure gradient VP shunt valve to the existing TP shunt. The procedure required opening of subcutaneous plane in the abdomen, identifying the tube and dividing it just before it enters the peritoneum and attaching pressure gradient VP shunt chamber. It was a short procedure which only required small incision in subcutaneous plane in the abdomen and attaching the pressure gradient VP shunt chamber. In developing countries where programmable theco-peritoneal shunts are unavailable, this novel procedure can be used as an alternative.

CONCLUSION

Low pressure headaches following lumbar theco-peritoneal shunt is a known complication. It can be managed by various methods. Attaching programmable chamber to the existing theco-peritoneal shunt is alternate, short, day care procedure, cost effective and involves minimal operative risk.

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