

Tension Pneumocephalus Following LP Shunt due to Congenital Bone Defects: A Case Report

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Abstract

A 72-year-old man who had undergone a lumboperitoneal shunt for idiopathic normal pressure hydrocephalus was admitted to our emergency department with fever and disturbance of consciousness 8 days after placement. Computed tomography scan showed pneumocephalus and a right-sided temporal porencephalic cyst with a small bone defect in the right petrous bone. Shunt valve pressure was raised from 145 mmH₂O to “virtual off” setting. After 2 weeks, follow-up computed tomography showed improvement of pneumocephalus, and the shunt valve pressure was lowered to 215 mmH₂O. Since that time, the patient has a good clinical course without recurrence. Tension pneumocephalus following shunt placement for idiopathic normal pressure hydrocephalus is rare and has never been reported in the early postoperative stage after lumboperitoneal shunt, except for the present one. Temporary raising shunt valve pressure is effective in improving the pneumocephalus. Preoperative screening for congenital bone defects by thin-slice computed tomography may be useful for selecting types of shunt valve and determining postoperative pressure setting.

Keywords: tension pneumocephalus, LP shunt, iNPH, congenital bone defect

Introduction

Tension pneumocephalus (TP) is a neurosurgical emergency that causes an increase in intracranial pressure (ICP) due to air or gas accumulation in the dura mater, subarachnoid space, lateral ventricles, or parenchyma.¹⁾ It has often been reported following head trauma and as a complication of neurosurgical procedures. In rare cases, it can occur after shunt placement for hydrocephalus. In this report, we present a rare case of TP observed in the early postoperative stage after lumboperitoneal (LP) shunt placement for idiopathic normal pressure hydrocephalus (iNPH) caused by congenital petrous bone defects.

Case Report

Presenting with progressive cognitive dysfunction and gait disturbance, a 72-year-old man had no history of

trauma or intracranial disease. Computed tomography (CT) scan showed tight high convexity cerebrospinal fluid (CSF) spaces, ventriculomegaly, and enlarged Sylvian fissures that suggested disproportionately enlarged subarachnoid space hydrocephalus (Fig. 1). His gait disturbance and cognitive disorder improved after CSF tap test. The initial cerebrospinal pressure was 105 mmH₂O. Therefore, he was diagnosed with iNPH, and, after the consent from him was obtained, an LP shunt with a pressure programmable valve was placed (Codman CERTAS Plus Programmable Valve with SiphonGuard, Integra LifeSciences Switzerland Sàrl, Rue Girardet, Switzerland). The shunt valve pressure was set at setting 5 (\approx 145 mmH₂O in a lying position) with reference to body weight and height.

The patient's gait was gradually stable, and he was discharged ambulatory on the eighth day without any complications. On the same day, he was found lying down at home and was taken to the emergency department again.

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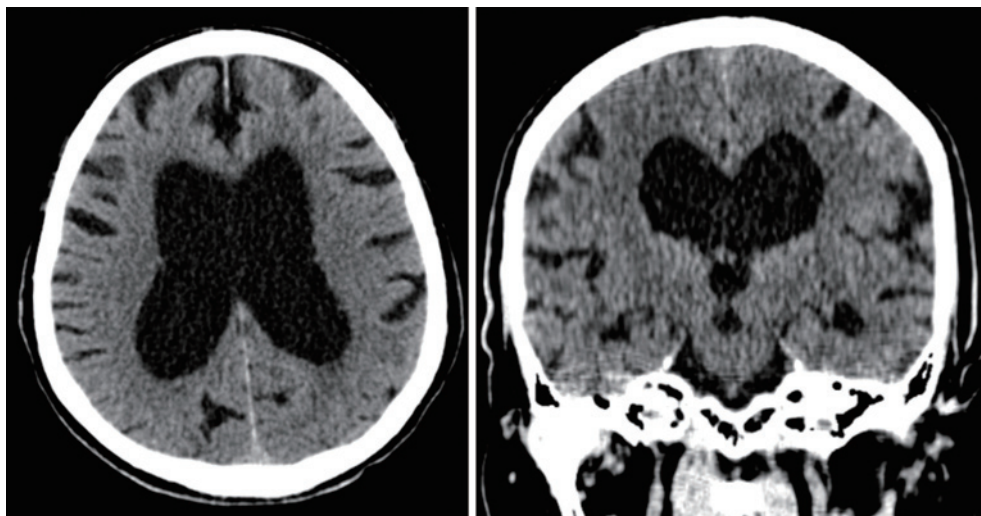


Fig. 1 Axial and coronal views of preoperative CT scan. These showed tight high convexity CSF spaces, ventriculomegaly, and enlarged Sylvian fissures that suggested disproportionately enlarged subarachnoid space hydrocephalus.

At readmission, he had a fever of 37.4°C and slightly impaired consciousness. CT scan and MRI showed significant air accumulation in the lateral ventricles and subarachnoid space and a right temporal porencephalic cyst that extended to the inferior horn of the right ventricle (Fig. 2A, B). A bone window setting of high-resolution CT scan showed that a part of the air cell at the right petrous bone was opened into the subdural space (Fig. 2C). Retrospectively, preoperative CT showed a finding of suspected small encephalocele through the petrous bone defects (Fig. 2D). Based on these findings, the patient was diagnosed with a TP caused due to the right petrous bone defect. His fever was suggested to be due to bacterial meningitis, but clinical and image findings showed no CSF leakage. First, he was placed on bed rest, and the shunt pressure was changed to setting 8 (virtual off setting; limiting flow through the valve). Antibiotics were given for 2 weeks. The postprocedural course was uneventful, and the CT scan showed gradual improvement of pneumocephalus. Then, the shunt pressure was decreased to setting 7 (\approx 215 mmH₂O) on the 14th day after readmission. He was transferred to another hospital for rehabilitation and discharged ambulatory on the 54th day. No intracranial air was detected in the follow-up CT scan 3 months after readmission (Fig. 3), and his gait and cognitive disorder improved with the shunt pressure setting 7.

Discussion

TP following shunt placement was first reported by Pitts et al. in 1975.²⁾ More than 10 cases of TP as a complication of shunt placement for hydrocephalus associated with brain tumor have been reported.³⁾ However, the case of iNPH is rare, especially following LP shunt placement. To the best of our knowledge, there is no reported case of TP

following LP shunt for iNPH, while 6 cases of TP following ventriculoperitoneal (VP) shunt placement following iNPH have been reported (Table 1).^{4,9)} There are two reports of TP associated with intraoperative procedure within 1 day after VP shunt placement.^{6,7)} On the other hand, in case of congenital bone defect, TP tended to develop in the chronic postoperative stage, several months or years after VP shunt placement. The present case is the first report of TP that developed following LP shunt placement due to congenital bone defect in an early postoperative stage.

According to previous reports, congenital bone defects of the skull base,^{4,5,8,9)} inadequate repair of frontal sinus opened during surgery,¹⁰⁾ or a skin fistula just above the burr hole^{7,11)} can cause the air inflow. In the present case, petrous bone defects were confirmed by thin-slice CT, and congenital bone defects were considered the cause of pneumocephalus since there was no history of trauma, tumor, or surgery. The pneumocephalus following shunt replacement due to congenital bone defects like our case is considered to be caused by the following three factors: (1) ICP increased by hydrocephalus thins the dura above the bone defects, resulting in a partial tear of it and fistula formation. (2) The brain parenchyma invaginated into the fistula acts like a plug and prevents the air inflow and CSF leakage before shunting. (3) The pressure gradient due to shunting leads to the drawing of air from the mastoid cells into the brain parenchyma through the bone defects. Like a one-way valve, this process is repeated because the brain parenchyma pressed by the air re-plugs the fistula. As a result, a porencephalic cyst is often formed, and as the cyst grows and penetrates into the ventricle, TP develops like in the present case. In cases with petrous bone defects, it is common to form a porencephalic cyst in the temporal lobe.²⁾ Ahren C et al. and Lang DV et al. reported that more than 20% of the general population have congenital

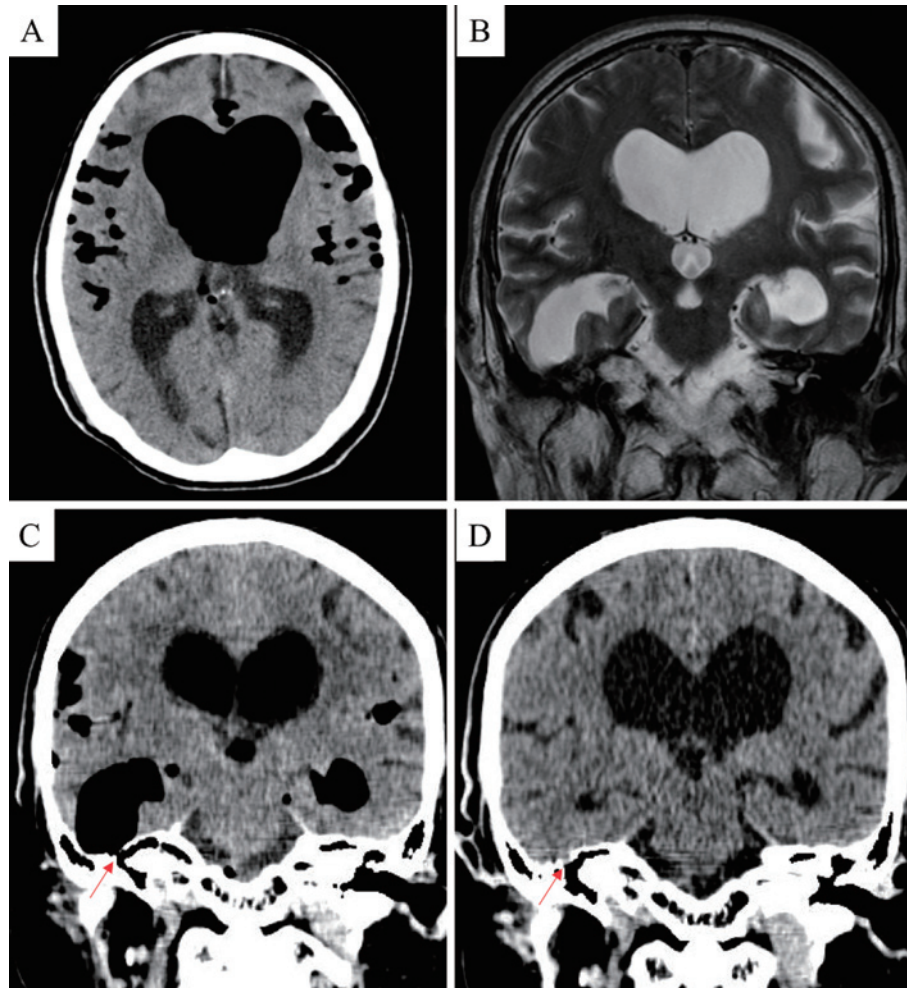


Fig. 2 Image findings related to TP due to congenital bone defects.

A) CT scan revealed significant air accumulation observed in the lateral ventricles and subarachnoid space.

B) MRI revealed a temporal porencephalic cyst extended to the inferior horn of the right ventricle.

C) Postoperative thin-slice CT scan showed a part of the air cell (red arrow) at the right petrous bone (coronal view).

D) Preoperative thin-slice CT revealed a small encephalocele through the petrous bone defects (red arrow).

bone defects of the tegmen tympani.^{12,13}) Once the porencephalic cyst is formed, bone defects can be identified. However, it is difficult to identify while congenital bone defects are asymptomatic. In the present case, thin-slice CT was helpful in detecting bone defects that caused pneumocephalus (Fig. 2C). Preoperative CT scan revealed a small encephalocele, suggesting petrous bone defects as the cause of air inflow (Fig. 2D). However, no findings suggestive of otitis media were noted. In addition, there is no history of meningitis nor intracranial hypotension although small encephalocele via a petrous bone defect is often the cause of them. These findings support the mechanism described above, that encephalocele might act like one-way valve. Considering that the bone defect in this case was extremely small, we speculated that the encephalocele might have become apparent after hydrocephalus onset. And, due to ICP increased by hydrocephalus, the pressure difference between intra and extracranial might not have

become large enough to allow air inflow in even when standing before shunting.

Compared with VP shunt cases, the earlier onset of TP in the present case also provides insight into the pathophysiology. In the seated position, the CSF pressure increased in the lumbar region corresponding to the distance from the cisterna magna,^{14,15} while ICP decreased compared to those in the lying position.¹⁶) These findings indicated that the CSF pressure gradient may fluctuate depending on the posture after LP shunt, unlike VP shunt. Although the shunt pressure is often set at the recommended setting referring to body weight and height,¹⁷ the higher pressure setting is desirable to prevent overdrainage, especially in patients with congenital bone defects, where, depending on the posture, the CSF pressure applied to the shunt valve placed at the lumbar region can be higher than the initial pressure measured in the lying position. Preoperative screening for congenital bone defects by

thin-slice CT may be useful for selecting types of shunt valve and determining postoperative pressure setting. In



Fig. 3 Axial view of CT scan after TP improvement.

other words, for shunting in iNPH patients with congenital bone defects, it is recommended to use pressure-adjustable valves, which can achieve “virtual off setting,” so that minimally invasive treatment can be selected when TP occurs. In addition, it is also recommended to set the shunt pressure higher than usual and adjust the pressure checking the signs of TP at narrow intervals in order to prevent severe TP.

Direct closure of the dural fistula^{2,4,8,9)} or removal of the shunt system^{3,5)} has been reported as a treatment option for TP following shunt placement. However, recently introduced pressure-adjustable valves can change the CSF pressure to the same CSF level as the condition with the shunt removal, “virtual off setting,” without surgical intervention. As the ICP rises again, air is pushed out of the skull over a few weeks. In the process of improving pneumocephalus, such as traumatic CSF leakage, the fistula may be smaller due to the granulation associated with meningitis. To prevent CSF leakage and improve the symptoms of hydrocephalus, readjustment of shunt pressure should be considered once pneumocephalus has improved. Reducing the ICP even slightly can lead to promoting remodeling in the dura mater. Regarding the target shunt pressure after the improvement of TP, it is not necessary to lower to the preoperative CSF pressure. In patients with iNPH who underwent LP shunt, it has been reported that approximately

Table 1 Previous case reports on shunt-associated tension pneumocephalus in patients with idiopathic normal pressure hydrocephalus

Author (year)	Age (years)/ Sex	Shunt type	Duration of shunting	Location of air entry	Porencephalic cyst	Management of shunt	Management of fistula
Kanner et al. (2000) ⁴⁾	63/M	VP	30 months	Congenital petrous bone defect	-	-	Mastoidectomy and fat packing
Nagai et al. (2007) ⁵⁾	78/M	VP	20 days	Congenital petrous bone defect	Temporal lobe	Valve temporary raised at 200 mmH ₂ O	-
Barada et al. (2009) ⁶⁾	79/F	VP	Within 1 day	Dura opening for placement of the ventricular catheter	-	Valve temporary raised at 140-170 mmH ₂ O	-
Aydoseli et al. (2013) ⁷⁾	75/M	VP	Within 1 day	Wound defect on ventriculostomy incision	-	-	External ventricular drainage and wound closure
Martinez-Perez et al. (2017) ⁸⁾	75/M	VP	2 months	Congenital petrous bone defect	Temporal lobe	LP shunt removal (LP for ethmoidal CSF leakage 15 years before VP)	Closing defects of the bone and dura
Verhaeghe et al. (2018) ⁹⁾	81/-	VP	10 months	Congenital petrous bone defect	Temporal lobe	-	Covering with fascia of the temporal muscle
Present case	72/M	LP	8 days	Congenital petrous bone defect	Temporal lobe	Valve temporary raised at “virtual off”	-

15% of patients had their optimal pressure even above 210 mmH₂O.¹⁸⁾ In the present case, the symptoms of iNPH improved at 215 mmH₂O, and there were no signs of otitis media associated with CSF leakage. Therefore, the patient could be followed up for one year without further reduction of shunt pressure. It should be noted that there is still a risk of repeated pneumocephalus without shunt removal or consistent “virtual off setting.” While on the one hand asymptomatic congenital bone defects are difficult to detect preoperatively and are often discovered only after the onset of pneumocephalus with meningitis, direct closure of the dural fistula should be performed to avoid this risk. Closing the dural fistula with craniotomy during the acute phase of infection itself carries the risk of infection. Therefore, if the general condition is good and neurological abnormalities are minor, as in this case, conservative treatment may be an option. If conservative treatment does not effect within a few days, or if neurological symptoms are severe, direct dural closure is necessary even in the acute phase.⁷⁾ Considering that TP occurs from several days to 30 months after shunting, careful follow-up to check the signs of pneumocephalus is essential for avoiding aggravation not only in the short term but also in the long term, even if conservative treatment is successful, as in this case. Without shunt revision or consistent “virtual off setting,” direct closure of the dural fistula should be considered to prevent recurrence in the chronic phase.

Conclusion

The present case is the first report of TP that developed due to congenital skull base defects in an early postoperative stage after LP shunt for iNPH. Preoperative screening for congenital bone defects by thin-slice CT may be useful for selecting types of shunt valve and determining postoperative pressure setting. Temporary raising shunt valve pressure, “virtual off” setting, is one of the effective treatments for TP.

Conflicts of Interest Disclosure

The authors declare no conflict of interest.

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