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Refractory Delayed Pneumocephalus after Transsphenoidal Cyst Drainage for Rathke's Cleft Cyst in a Patient with a Cerebrospinal Fluid Shunt

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Abstract

A 75-year-old man presented with bilateral lower limb weakness to our hospital from another clinic. Radiological examinations implied the possibilities of idiopathic normal pressure hydrocephalus (iNPH) and a suprasellar cyst, but both were observed conservatively at that time. Due to the progressive gait disturbance, a lumboperitoneal shunt was implanted 1 year later. The clinical symptoms improved, but the cyst had grown after another year, causing visual impairment. Transsphenoidal drainage of the cyst was performed, but delayed pneumocephalus occurred. Repair surgery was performed with temporary suspension of shunt function, but pneumocephalus relapsed two and a half months after the resumption of shunt flow. In the second repair surgery, the shunt was removed because it was assumed that it would prevent closure of the fistula by lowering intracranial pressure. Two and a half months later, after confirming involution of the cyst and no pneumocephalus, a ventriculoperitoneal shunt was implanted, and cerebrospinal fluid (CSF) leakage has not relapsed since then. The coexistence of idiopathic normal pressure hydrocephalus (iNPH) and Rathke's cleft cyst (RCC) is rare, but it can occur. RCC can be cured by simple drainage, but delayed pneumocephalus can occur in cases whose intracranial pressure decreases due to CSF shunting. When simple drainage without sellar reconstruction for RCC is attempted after CSF shunting for coexistent iNPH, attention should be paid to changes in intracranial pressure, and it is desirable to stop the flow of the shunt for a certain period.

Keywords

- delayed pneumocephalus
- idiopathic normal pressure hydrocephalus
- lumboperitoneal shunt
- Rathke's cleft cyst
- transsphenoidal surgery

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Introduction

The case of a patient with a Rathke's cleft cyst (RCC) and idiopathic normal pressure hydrocephalus (iNPH) is reported. Due to enlargement of the cyst after lumboperitoneal shunt (LPS) placement, it was drained by the transsphenoidal approach, but the patient developed delayed pneumocephalus. The clinical course of this case is presented, and the relationship between cyst enlargement and the intracranial environment is discussed. This is the first report of such a complicated case.

Case Report

Present History

A 75-year-old man visited a family doctor due to weakness of the lower limbs for the past 2 years. The disproportionately enlarged subarachnoid space hydrocephalus sign (►**Fig. 1A**) and a suprasellar cystic lesion were found (►**Fig. 1B**), and he was referred to our department. At the first consultation, it was decided to observe the suprasellar lesion conservatively, because ophthalmological examination showed no abnormalities. As for the probable iNPH, he was also conservatively observed because he was almost free of clinical symptoms, but 6 months later, his gait disturbance became worse due to progression of the iNPH. Since the cerebrospinal fluid (CSF) tap test was positive, LPS placement was performed using a CERTAS plus valve (Integra Life Sciences Holdings Inc., New Jersey, United States) as a pressure programmable shunt valve, and the pressure level was finally set to level 2 (corresponding to 5–7 cmH_2O).

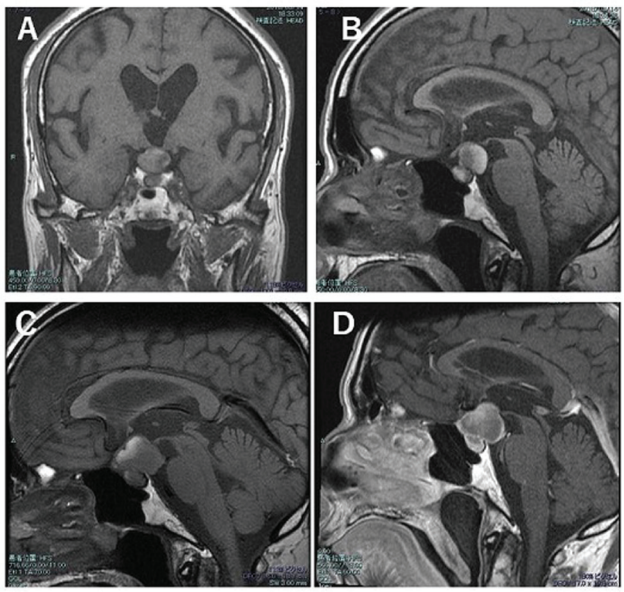

Fig. 1 Head MRI scan obtained at the initial consultation (**A**) and at the enlargement of the cyst (**B**–**D**). (**A**) Ventricular dilatation, widening of the Sylvian fissure, narrowing of the callosal angle, and high-convexity subarachnoid space tightness are seen on T1-weighted coronal MRI, compatible with the DESH sign. (**B**) A suprasellar cystic lesion is seen as a mixed intensity mass on T1-weighted sagittal MRI. (**C**) At the time of exacerbation of visual impairment, the cyst has grown larger than at the first consultation. (**D**) The wall of the cystic lesion is contrast-enhanced on gadolinium-enhanced T1-weighted sagittal scan, but the inside of the cyst is not enhanced. DESH, disproportionately enlarged subarachnoid space hydrocephalus; MRI, magnetic resonance imaging.

The patient's postoperative course was good, and improvements in gait and cognitive function were observed. One year after the operation, the patient's activities of daily living had been maintained, and no problems had occurred in the postoperative course of iNPH, but head magnetic resonance imaging (MRI) showed enlargement of the suprasellar lesion (►**Fig. 1C,D**). At the same time, the patient complained of a visual disturbance, and on ophthalmological examination, bilateral hemianopsia and decreased visual acuity in the right eye were found.

Radiological Findings

On MRI, the cyst protruded from intrasellar to suprasellar through the diaphragma sellae and pushed the optic chiasm upward. The inside of the cyst showed heterogeneous intensity. On contrast-enhanced MRI, the cyst wall was thinly enhanced, but there were no contrast-enhanced components inside (►**Fig. 1C,D**). Radiologically, RCC was highly suspected. Endocrinological examination showed no abnormal pituitary hormone levels.

Transsphenoidal Surgery

Because the RCC became symptomatic due to the enlargement of the cyst, cyst fenestration and drainage were performed by endoscopic transsphenoidal surgery (TSS). By incising the dura mater of the sella turcica, normal pituitary tissue was identified, and after splitting it, milky white liquid spilled out. Although part of the wall was collected and submitted for histological examination, it could not be diagnosed as RCC histologically, but based on the clinical and intraoperative findings, the diagnosis of RCC was considered valid. CSF leakage was not observed during the operation, and the operation was therefore finished without sellar floor reconstruction, expecting a continuous drainage effect. During the perioperative period, the setting of shunt valve pressure had been kept at level 2 (corresponding to 5–7 cmH2O).

Postoperative Course following the First Transsphenoidal Surgery

The patient's postoperative course was uneventful, and the visual impairment improved. One week after the operation, computed tomography (CT) still showed considerable air retention in the cyst, but no air in the subarachnoid cistern (►**Fig. 2A**). No CSF leak was evident both subjectively and objectively, and the patient was discharged from hospital. Two weeks after the operation, the patient's visit to the outpatient clinic was uneventful.

But approximately 1 month after the operation, the patient developed a consciousness disturbance and was again hospitalized. The patient's consciousness level at that time was Glasgow coma scale (GCS) 12 (E2/V4/M6). CT showed marked intracranial air (►**Fig. 2B**), suggesting an obvious CSF leak.

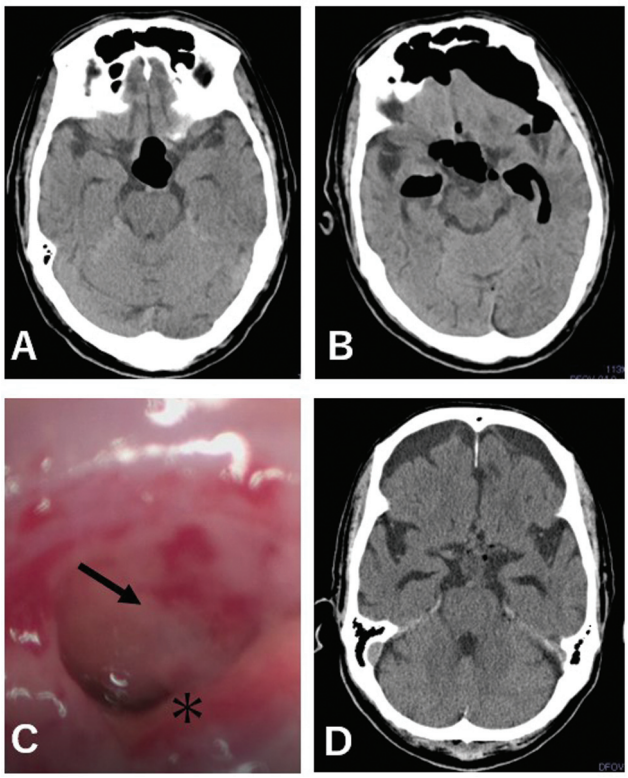

Fig. 2 (**A**) Axial CT scan 1 week after the initial TSS still shows air retention in the cyst, but no air in the cistern. (**B**) Axial CT scan at the first emergent admission after discharge shows marked air retention in both the inside of the cyst and the subarachnoid cistern. (**C**) Intraoperative findings of the sellar floor at the first repair surgery are shown. The membrane of the cyst wall has been torn (*arrow*) in the sellar region, and CSF has leaked from the fenestrated window at the sellar floor (*asterisk*). (**D**) Axial CT scan 1 month after the initial repair surgery for CSF leakage shows that the air has diminished in both the inside of the cyst and the subarachnoid cistern. CT, computed tomography; CSF, cerebrospinal fluid; TSS, transsphenoidal surgery.

Treatment of the Cerebrospinal Fluid Leak

Although there were no obvious meningeal signs on admission, antibiotics were administered prophylactically. Due to the insertion of the LPS, it was assumed that the CSF was flowing into the intraperitoneal cavity without causing obvious rhinorrhea. Since CSF shunting could further vacuum air into the intracranial cavity, the shunt valve was switched to virtual off mode to stop the flow into the intraperitoneal cavity. Although CT the next day showed decreased intracranial air, and it was assumed that the inflow of air could be minimized, it was thought necessary to reconstruct the sellar floor to stop the CSF leak completely, and repair surgery was performed by TSS. By observation of the sellar floor, CSF leakage was clearly confirmed, and when observing the inside of the cyst, part of the membrane-like substance was lacerated, and CSF leaked from there (►**Fig. 2C**). The cyst was filled with abdominal fat, and the sellar floor was reconstructed by artificial absorbable material (Lactosorb; Zimmer Biomet Holdings, Inc., Florida, United States). The sphenoid sinus was also filled with fat.

Postoperative CT showed a further decrease of intracranial air, suggesting that the CSF had stopped leaking. The patient's consciousness level improved when the virtual off mode was released immediately after the operation and the shunt was activated again. The patient's subsequent course was also uneventful, and no relapse of pneumocephalus was observed (►**Fig. 2D**). The patient was transferred to another hospital for rehabilitation.

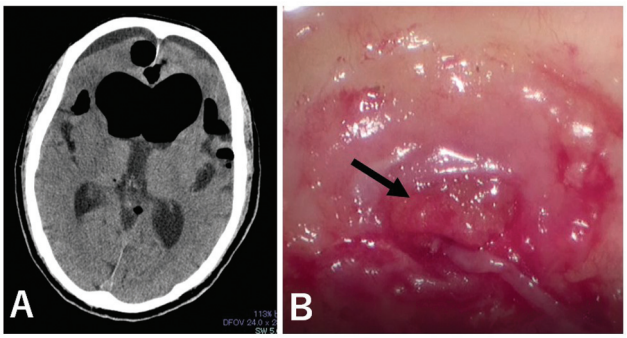

Fig. 3 (**A**) Axial CT scan at the second emergent admission shows marked air retention in the lateral ventricles and the subarachnoid cistern and ventricular dilatation. (**B**) Intraoperative findings of the sellar floor at the second repair surgery is shown. Previously filled fat tissue has been partially melted (*arrow*), and CSF has exuded from the aperture. CSF, cerebrospinal fluid; CT, computed tomography.

Recurrence of Pneumocephalus and Subsequent Treatment

However, one and a half months later, the patient's level of consciousness deteriorated again. CT showed significant intracranial air (►**Fig. 3A**). The level of consciousness was GCS 7, and he was febrile. The findings of CSF collected from the shunt valve were compatible with meningitis. Diagnosed as a relapse of pneumocephalus and shunt infection, transsphenoidal repair surgery and shunt removal were performed. Observation inside the sphenoid sinus showed that the filled fat was partially melted (►**Fig. 3B**). CSF leakage was observed from the sellar floor by the Valsalva method. After the cyst was again filled with abdominal fat, the sellar floor was reconstructed by Lactosorb and closed in a multilayered manner. Specifically, the reconstructed sellar floor was covered by rectus abdominis muscle membrane, the complex was sealed by fibrin glue, and the sphenoid sinus was also filled with fat. Subsequently, the infected shunt system was removed temporarily.

After the operation, intracranial air decreased over time on imaging, and the CSF leakage seemed to have stopped. Antibiotic treatment improved the meningitis. On the other hand, the discontinuation of the CSF shunt exacerbated ventricular enlargement (►**Fig. 4A**), and he remained in a bedridden state.

Two and a half months after the second repair surgery, a new ventriculoperitoneal shunt (VPS) was inserted after confirming the involution of the cyst and no pneumocephalus. The shunt pressure was gradually reduced while confirming that there was no recurrence of pneumocephalus, and it finally reached the previous setting. He gradually improved neurologically and was transferred to a rehabilitation hospital. CT three and a half months after the second repair surgery showed no intracranial air and a shrunken suprasellar cyst (►**Fig. 4B,C**).

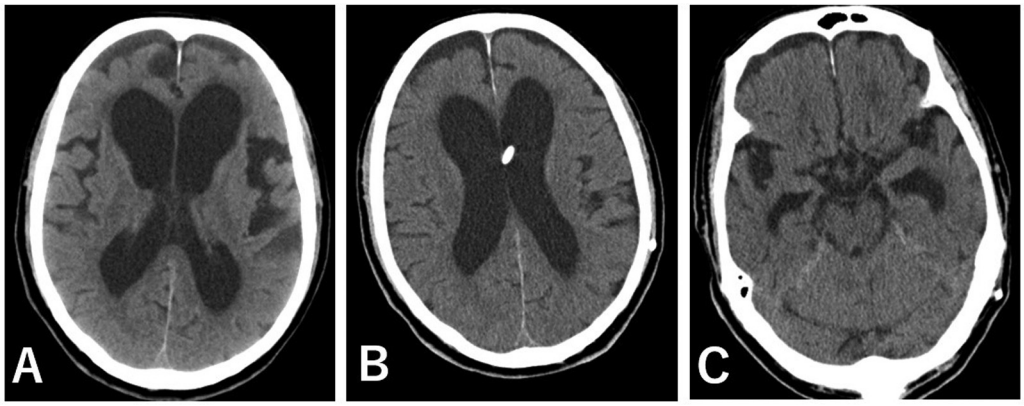

Fig. 4 (**A**) Axial CT scan 2 months after the repair surgery for the second CSF leakage shows less air and ventricular dilatation. (**B**) Axial CT scan after VPS (three and a half months after the second repair surgery) shows improvement in ventricular dilatation. (**C**) Axial CT scan after VPS shows shrinking of the RCC. CSF, cerebrospinal fluid; CT, computed tomography; RCC, Rathke's cleft cyst; VPS, ventriculoperitoneal shunt.

Discussion

iNPH is a CSF circulation disorder that causes gait and cognitive dysfunction in the elderly, and it is treated by CSF shunting with good recovery of these functions.[1] LPS placement is now widely performed for probable iNPH patients, especially in Japan, and the number of surgical cases is increasing with the aging of the population and the spread of the disease concept.[2] Incidental coexistence of RCC in such patients is not frequent, but quite possible. In the present patient, since the RCC enlarged and caused progressive visual impairment, it needed to be treated.

An RCC is a benign cystic lesion, believed to be the embryological remnants of Rathke's pouch, which is located within the sellar and suprasellar region.[3] It is most commonly found incidentally, but it may become sufficiently large to compress the optic apparatus and pituitary gland, resulting in symptoms such as headache, visual disturbance, and endocrine dysfunction. In patients with clinical symptoms and radiographical progression, treatment is indicated. The gold-standard treatment remains TSS, and several surgical strategies have been described, ranging from simple cyst fenestration to complete resection.[4] To prevent recurrence, wide cyst fenestration without sellar reconstruction is recommended to promote continuous drainage of the cyst, unless an intraoperative CSF leak is found.[5] In the present case, because enlargement of the cyst was confirmed and visual impairment was progressing, surgery seemed appropriate, and simple fenestration without sellar floor reconstruction was selected.

One of the most common postoperative complications following TSS for sellar lesions is CSF leakage, which has been reported to occur in up to 15% of cases (range: 0.8–15%), with most authors citing an incidence rate of 1 to 4%.[6] Lobatto et al reported that cystic lesions, for example, craniopharyngioma and RCC, appear to harbor the highest risks, but few studies have looked at the relationship between various forms of pathology and CSF leakage.[7] In the present case, CSF leakage and subsequent pneumocephalus occurred in a delayed fashion that appeared about 1 month after TSS. Recently, Strickland et al reported that delayed CSF leakage developed in patients in whom there was neither an intraoperative leak nor sellar floor repair, but they reported that there were no cases of postoperative rhinorrhea in patients who underwent sellar floor repair without intraoperative CSF leakage.[6] Marcus et al reported a relatively higher incidence of CSF leakage in patients with RCC in their series than in the other reported series.[8] They suggested that this was because, if no obvious CSF leak is identified intraoperatively, they often attempted to establish free drainage of the cyst with the sphenoid sinus, and in such cases, where there is a laceration in the arachnoid, either unrecognized intraoperatively or later postoperatively, this could result in a CSF fistula.

In the present case, only opening of the dura and minimal splitting of the pituitary gland in front of the sellar floor and drainage of the cyst were performed, and CSF leakage was not observed intraoperatively. Moreover, no air collection in the intracranial subarachnoid space was detected on CT 1 week after the operation, and apparent CSF rhinorrhea was not observed. Therefore, intraoperative laceration of the arachnoid membrane was not presumed to have occurred. Intracranial pressure is usually higher than the pressure in the cyst, so the fenestrated cyst shrinks as it is compressed. In the present case, however, an LPS had been placed, which caused the intracranial pressure to decrease. Therefore, the intracranial pressure was lower than the intracapsular pressure, which might have caused the cyst to enlarge, resulting in rupture of the cyst wall and leading to pneumocephalus. Krishnan et al reported one case of the late presentation of delayed tension pneumocephalus in a patient who underwent endoscopic transnasal repair of a CSF fistula followed by LPS and noted that the condition is potentially lethal and requires prompt recognition and surgical treatment.[9]

One reason for the recurrence of CSF leakage and subsequent pneumocephalus after the initial CSF leak closure was thought to be the reversal of shunt suspension too early after the repair surgery. Therefore, it was temporarily removed after the second repair of the CSF leak. The patient was finally cured by performing a VPS. Although RCCs are known to grow and shrink spontaneously,[10] enlargement of the cyst after shunting suggests that intracranial pressure may have been involved in the spontaneous change in cyst size. If TSS is chosen for RCC after CSF shunting, it may be desirable to set

the shunt to virtual off in advance during the perioperative period or to perform sellar floor reconstruction after drainage. Therefore, a CERTAS plus valve with a virtual mode should be used as the shunt system.

Conclusion

A rare case of repeated CSF leakage after transsphenoidal simple drainage for RCC that coexisted with iNPH treated by LPS was reported. The decrease in intracranial pressure due to continuous CSF shunting likely disturbed the collapse of the cyst, resulting in repeated episodes of pneumocephalus. When performing simple drainage without sellar reconstruction for RCCs in patients with CSF shunt insertion, it is desirable to stop the flow of the shunt for a sufficient period.

Ethical Approval and Informed Consent

This study was approved by the Ethics Committee of Kansai Medical University (No. 2020055). Need for written patient consent was waived by the Ethics Committee because data were deidentified.

Conflicts of Interest

None declared.

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