Pneumocephalus in patients with CSF shunts

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The authors present two cases of pneumocephalus occurring in patients with permanent shunts and review nine previously reported cases. Mental status changes and headache are the most common presenting symptoms. Six of the 11 cases of pneumocephalus occurred in patients with shunt placement for hydrocephalus secondary to aqueductal stenosis. In these patients, thinned cerebrospinal fluid barriers secondary to long-standing increased intracranial pressure may predispose them to pneumocephalus. Temporary extraventricular drainage is an effective method of treatment in this group of patients. Two other etiologies are identified with significance to treatment, and the role of craniotomy is discussed.

KEY WORDS • pneumocephalus • cerebrospinal fluid shunt • hydrocephalus • extraventricular drainage

Pneumocephalus was first described in 1884 by Chiari as an autopsy finding in a patient who died of ethmoiditis. Luckett was the first to make the roentgenographic diagnosis of pneumocephalus in 1913. Since then, more than 370 such cases have been described. Between 1914 and 1918, there were frequent reports of pneumocephalus secondary to war injuries. Indeed, the majority of cases of pneumocephalus are caused by trauma. In 1962, pneumocephalus was reported by Kessler and Stern as a complication of cerebrospinal fluid (CSF) shunting in a patient with a ventriculopleural (VP) shunt. The first case of pneumocephalus in a patient with a ventriculoperitoneal (VP) shunt was reported in 1975 by Pitts, et al. Since then, seven isolated case reports have appeared in the English literature, and we are adding two more. In this paper we briefly review these 11 cases with particular attention to the management of pneumocephalus in patients with CSF shunts.

Case Reports

Case 1

This 28-year-old man was admitted to Northwestern Memorial Hospital in Chicago in August, 1984, complaining of amnesia. He initially came to medical attention at the age of 19 years, when an enlarged sella turcica was noted on skull films taken after a minor skiing accident. Endocrine studies at that time were normal. At 25 years of age, he presented for evaluation of a generalized complex seizure. Investigation revealed hydrocephalus secondary to aqueductal stenosis, and a VP shunt was placed. He did well and led a very active life as an insurance claims adjuster. Six days before admission he was water-skiing and fell several times without sequelae. Two days prior to admission he awoke with a headache and complained of malaise. He attributed this to influenza, and spent 2 days in bed. On the morning of admission he felt much better and denied headache. His mother, however, identified obvious memory deficits and brought him to the emergency room.

Examination. Upon questioning, the patient denied headache and felt well but had been amnestic for the prior 2 days. He was alert and oriented. His short-term memory was poor. He was unable to recite the months backwards but was able to give his home address and telephone number. The remainder of his physical and neurological examination was normal. Abdominal roentgenograms were normal. Computerized tomography (CT) revealed a large collection of air in the right inferior frontal region and the anterior portion of the lateral ventricles. Coronal sections demonstrated a communication through the roof of the ethmoid bone anterior to, and to the right of, the crista galli. Enlarged ventricles were noted, and the patient was taken to surgery.
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Operations. The VP shunt was removed, and an extraventricular drain was placed. The cephalic portion of the shunt was found to be occluded, and the distal tip was patent. Shunt tip cultures were negative. The patient did well until 11 days postoperatively, when he became confused and drowsy. He had limited upward gaze. A CT scan revealed an increased ventricular size; air was seen, contained as a pneumocoele and apparently not in communication with the ventricles or cranial base. The ventriculostomy was revised. He improved dramatically, and the difficulty with upward gaze resolved. Six days later metrizamide was instilled through the ventriculostomy into the ventricles, which were found not to communicate with the resolving intracerebral air pocket. The shunt was internalized 21 days after initial removal.

Postoperative Course. The patient was discharged 13 days later and has done well. Prophylactic antibiotic agents were given during the time of extraventricular drainage. Throughout his course, the extent of pneumocephalus was followed with skull roentgenography using a portable system. These films demonstrated a gradual absorption of the air.

Case 2

In 1973, this previously healthy 7-year-old boy sustained a serious brain contusion in an automobile accident. He underwent a right subtemporal decompression. A VP shunt was inserted after he developed posttraumatic hydrocephalus. Within weeks the shunt became infected and was removed. He was transferred to Children's Memorial Hospital in Chicago, where he was found to have hydrocephalus and a posterior fossa arachnoid cyst. The cyst was resected, and a VP shunt was placed. He underwent six shunt revisions. He presented for the seventh shunt revision with a low-grade fever, anorexia, and lethargy. Head circumference was 53 cm, and there was no papilledema. Cultures of CSF, blood, and urine were negative. Three days after admission he remained lethargic and the craniectomy site was bulging.

Examination. The patient was an alert but significantly impaired child who could only follow simple commands. Head circumference was 53 cm, and there was no papilledema. Cultures of CSF, blood, and urine were negative. Three days after admission he remained lethargic and the craniectomy site was bulging.

Operations. His shunt was revised, and was found to be obstructed at the peritoneal end. A Raimondi spring catheter was connected to the ventricular portion of the unishunt system, and the abdominal portion was placed through a trocar inserted through the abdominal musculature into the peritoneal cavity. Three days postoperatively the patient developed spiking fevers. Multiple cultures during this time were negative and shunt-track and abdominal examinations remained normal. The patient continued to spike fevers; CSF was obtained by cisternal puncture for culture and was found to be sterile. The child's fever defervesced and he was released fifteen days after surgery.

He was readmitted 1 week later after an episode of vomiting. He reportedly had remained listless and tended to bend his neck to the left side (opposite to the shunt tube). His abdominal examination revealed no tenderness or peritoneal signs. Skull roentgenograms showed air in the right ventricle. He underwent shunt revision. The abdominal incision was opened and the shunt withdrawn. It was noted to be covered with feculent material. Bile was seen draining through the incision. Limited exploratory laparotomy revealed dense vascular adhesions to the anterior abdominal mass and a small colon perforation at the hepatic flexure. This was repaired, and an extraventricular drain was placed.

Postoperative Course. The patient was treated with antibiotic agents. Serial daily cultures of CSF revealed Escherichia coli for 4 days, which then cleared. The shunt catheter tips grew out multiple organisms on culture. The shunt was internalized 16 days later, when it was clear that the infection was controlled. Since that time, the patient has made slow progress intellectually, but has not required a subsequent shunt revision.

Summary of Cases

Presentation

Table 1 provides a summary of the reported cases of pneumocephalus associated with CSF shunts. Mental status changes ranging from mild memory deficits to coma with decerebrate posturing were present in five patients. In four patients, mild to severe headache was the primary complaint. Focal neurological findings were present in three patients: two had a mild spastic hemiparesis, and one had "intermittent" quadripareisis. The two patients with pleural complications had a persistent cough. Fever was the initial sign in two cases. Only one patient gave a history of a "sloshing" noise in his head — the classic "bruit hydroaerique" thought to be the only pathognomonic symptom of pneumocephalus.

Etiology

The diagnosis of pneumocephalus was confirmed in six cases by skull roentgenograms, in four cases by CT scanning, and in one case by angiography. In most of these cases there was difficulty in localizing the air communication preoperatively. Of the seven patients with VP shunts, five had fistulous communication with the skull base (two with the ethmoid sinus, two with the frontal sinus, and one through both tegmina tympani), in one patient the colon was perforated by the shunt catheter, and in one patient no fistula could be demonstrated. Of the three cases of ventriculoatrial shunts, two had fistulous communication with the skull base (one with the frontal sinus, and one with the ethmoid sinus), and one had catheter erosion into the
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**TABLE 1**

*Reported cases of shunt-associated pneumocephalus*

<table>
<thead>
<tr>
<th>Authors, Year</th>
<th>Age (yrs), Sex</th>
<th>Shunt Type</th>
<th>Cause of Hydrocephalus</th>
<th>Period of Shunting</th>
<th>Symptoms</th>
<th>Treatment</th>
<th>Etiology</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kessler &amp; Stern, 1962</td>
<td>29, M</td>
<td>pleural</td>
<td>arachnoiditis</td>
<td>6 wks</td>
<td>severe headache</td>
<td>reinsertion into opposite pleural cavity</td>
<td>erosion of bronchiole by pleural end of catheter fistulas of both tegmina tympani</td>
<td>good, 1 yr</td>
</tr>
<tr>
<td>Pitts, et al., 1975</td>
<td>21, ?</td>
<td>peritoneal</td>
<td>irradiation for cerebellar astrocytoma</td>
<td>1 yr</td>
<td>dementia, intermittent quadripareisis</td>
<td>lt: subtemporal duraplasty; rt: simple mastoidectomy</td>
<td>good, ? follow-up period</td>
<td></td>
</tr>
<tr>
<td>Little &amp; MacCarty, 1976</td>
<td>22, M</td>
<td>peritoneal, med-press Hakim valve</td>
<td>aqueductal stenosis</td>
<td>8 wks</td>
<td>rt leg weakness, rt Babinski, gait ataxia</td>
<td>lt frontal craniotomy, duraplasty</td>
<td>frontal sinus fistula</td>
<td>good, 1 yr</td>
</tr>
<tr>
<td>Muizelaar &amp; Walder, 1977</td>
<td>16, M</td>
<td>atrial, med-press Holter valve</td>
<td>posttraumatic</td>
<td>10 days</td>
<td>headache, emesis</td>
<td>frontal craniotomy, duraplasty</td>
<td>lamina cribrosa fracture</td>
<td>good, 1 mo</td>
</tr>
<tr>
<td>Stuntz &amp; Shuman, 1977</td>
<td>3½, F</td>
<td>atrial, Holter valve</td>
<td>posttraumatic</td>
<td>1 yr</td>
<td>tachypnea, cough, fevers, hypoxemia weakness Lt side</td>
<td>planned EVD, ligation of shunt</td>
<td>catheter erosion of bronchiole during ventilation</td>
<td>died preop</td>
</tr>
<tr>
<td>Ikeda, et al., 1978</td>
<td>22, F</td>
<td>peritoneal, med-press Pudenz valve</td>
<td>aqueductal stenosis</td>
<td>12 wks</td>
<td>sudden headache, comatose, decerebrate</td>
<td>lt frontal craniotomy</td>
<td>frontal sinus fistula</td>
<td>good, 24 hrs</td>
</tr>
<tr>
<td>Steinberger, et al., 1979</td>
<td>29, F</td>
<td>atrial, med-press Hakim valve</td>
<td>aqueductal stenosis</td>
<td>1 wk</td>
<td>sudden headache, comatose, decerebrate</td>
<td>bifrontal craniotomy</td>
<td>frontal sinus fistula</td>
<td>good, 5 mos</td>
</tr>
<tr>
<td>Findler, et al., 1980</td>
<td>18, M</td>
<td>peritoneal</td>
<td>aqueductal stenosis</td>
<td>8 yrs</td>
<td>2 wks post craniofacial op, incontinence, confusion</td>
<td>orbital roof twist-drill hole, aspiration of air, Trendelenburg op, antiisophon device</td>
<td>unknown</td>
<td>good, 1 wk</td>
</tr>
<tr>
<td>Jooma &amp; Grant, 1983</td>
<td>12, M</td>
<td>peritoneal, on-off valve, Heyer-Shulte shunt</td>
<td>aqueductal stenosis</td>
<td>9 mos</td>
<td>headache, CSF rhinorrhea</td>
<td>lt frontal craniotomy, duraplasty</td>
<td>ethmoid fistula</td>
<td>good, ? follow-up period</td>
</tr>
<tr>
<td>Ruge, et al., 1985</td>
<td>28, M</td>
<td>peritoneal, med-press unishunt shunt</td>
<td>aqueductal stenosis</td>
<td>3 yrs</td>
<td>memory deficit</td>
<td>temporary EVD</td>
<td>ethmoid fistula</td>
<td>good, 5 mos</td>
</tr>
<tr>
<td></td>
<td>7, M</td>
<td>peritoneal, med-press unishunt</td>
<td>posttraumatic</td>
<td>3 wks</td>
<td>emesis, nystagmus, fever</td>
<td>temporary EVD</td>
<td>perforated colon</td>
<td>good, 11 yrs</td>
</tr>
</tbody>
</table>

* Med-press = medium-pressure; EVD = extraventricular drainage; CSF = cerebrospinal fluid.

The only case of ventriculopleural shunt-associated pneumocephalus was secondary to catheter erosion into a bronchiole. Of the six patients shunted for aqueductal stenosis, five patients had fistulous communication with the cranial base irrespective of the terminus of the shunt.

Attempts to document the fistula preoperatively were successful in one case where polytomography revealed a suspicious area on the posterior wall of the frontal sinus; this was confirmed at craniotomy. In our Case 1 the fistula was seen on coronal CT cuts through the ethmoid bone but was never directly visualized. In seven cases, the pathology was confirmed at surgery either by direct visualization of a fistula or by hearing air blown through an external defect. The fistula was indirectly detected in a patient with a ventriculopleural shunt as CSF was seen freely flowing out of the endotracheal tube. In another patient, postmortem instillation of contrast material through the ventriculointeral shunt showed opacification of the right superior lobe of the lung, confirming a bronchial fistula.

**Discussion**

Pneumocephalus occurs most commonly secondary to trauma. In a series of 295 cases, Markham reported that trauma accounted for 73.9% and neoplasm for 12.9%. Intra- and postoperative pneumocephalus is well documented. Surgery in the sitting position, nitrous oxide anesthesia, and CSF draining intraoperatively all have been implicated. Infection...
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can also cause pneumocephalus either because of intracranial extension, as in sinusitis and otitis media, or by a primary gas containing brain abscess.\(^1\) Pneumocephalus has occurred iatrogenically when air was forced into the cranium using continuous positive airway pressure with a mask or by a pneumatic otoscope.\(^7\) We have reviewed 11 cases where pneumocephalus occurred in association with permanent CSF shunting. This phenomenon occurs when air is forced through the shunt or enters through the cranial base because of iatrogenic postsurgical communication, congenital fistula, trauma, or thinning of the cranial base.

The catheter can be a direct route for introduction of air into the cranium. Three cases are presented where the tube eroded either through a bronchiol or through the colon. The treatment of choice is shunt removal, closure of the communication if necessary, and replacement of the shunt at a different site, perhaps after a period of temporary extraventricular drainage. To our knowledge, pneumocephalus caused by colon perforation of a VP catheter has not been reported previously.\(^29\)

Continuity of the cranial floor is important in preventing pneumocephalus in the shunted patient. It is clear that large negative intracranial pressures can be developed by the siphon effect of shunts. Normal CSF pressure in the sitting position ranges from \(-30\) to \(-155\) mm H\(_2\)O. It has been shown that, in patients with shunts, pressures drop to as low as \(-440\) mm H\(_2\)O at the foramen of Monro.\(^19\) The CSF is displaced either through the shunt or through the cranial floor. The negative pressure and the displaced volume of CSF allow for the ingress of air to fill the vacuum. Air is then trapped by brain plugs by a ball-valve phenomenon. The “inverted bottle” theory postulates no requirement for a valve.\(^11\) Crucial to therapy is the etiology of the cranial base communication.

Six of these 11 known cases of pneumocephalus associated with shunted hydrocephalus occurred in patients with a history of aqueductal stenosis. Thinning of the cranial floor and dorsum sellae has been clearly demonstrated in patients with aqueductal stenosis.\(^5\) The bone erosion is apparently caused by chronically elevated ICP. Patients with a history of aqueductal stenosis requiring a CSF shunt seem particularly at risk for developing pneumocephalus. Is this because a prospective fistula becomes symptomatic or because thinned CSF barriers are easily traumatized by as little as a sneeze or a water-skiing fall, as possibly occurred in our Case 1? Craniotomy and direct repair of the communication might be too aggressive, and perhaps only a temporary solution as one might expect another area of thinning or potential fistula to be awaiting pressure alterations or mild trauma. Temporary extraventricular drainage against a pressure gradient resulted in sealing of the communication and resolution of the pneumocephalus in our Case 1. Perhaps these patients with thinned CSF barriers would benefit from high-pressure shunt valves or antisyphon devices as prophylaxis against pneumocephalus. These might reduce the high negative pressures and increase the margin of safety.

The patient with an isolated fistula through the cranial floor and no evidence of cranial floor thinning should be cured by craniotomy and direct fistula repair as there would be no reason to expect fistula recurrence. The role of temporary extraventricular drainage in these patients, mildly symptomatic from pneumocephalus, has yet to be determined.

Conclusions

Eleven cases of pneumocephalus in the permanently shunted patient are briefly reviewed as to presentation, etiology, and management. Mental status changes and headache were the most common presenting symptoms. Three categories of etiology were identified with significance to treatment. Direct shunt perforation of an air-containing organ is best managed with shunt removal, perhaps with temporary ventricular drainage and then replacement of the shunt at a different site. Isolated cranial base fistulas without evidence of basal thinning are managed best with craniotomy and direct fistula repair. Patients shunted for aqueductal stenosis appear to be particularly at risk for developing pneumocephalus. Temporary extraventricular drainage against a pressure gradient is an effective method of treatment in these patients. These therapy recommendations might serve as general guidelines until further experience is gathered with this uncommon complication of CSF shunting.

References

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