Spontaneous otogenic pneumocephalus

Case report and review of the literature

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Pneumocephalus is commonly seen in clinical neurosurgical practice. Typical causes include trauma, tumor, and infection. Pneumocephalus may also occur iatrogenically at the time of intracranial surgery; it is not pathological and may be seen routinely on postoperative neuroimaging. Pneumocephalus is rarely encountered in the absence of the aforementioned entities. The authors report on an elderly woman in whom spontaneous intraventricular pneumocephalus occurred because of a congenital defect in the left tegmen tympani. Eustachian tube closure and middle ear exclusion were used to obliterate the fistulous connection. This case illustrates both an unusual cause and a unique treatment for spontaneous otogenic pneumocephalus.

Key Words • pneumocephalus • petrous bone • skull base surgery • middle ear

Case Report

History and Examination. This 78-year-old woman presented with a 2-week history of mild left-sided headaches, intermittent confusion, and forgetfulness. Two days before admission she developed increasing confusion and expressive aphasia. On admission she was lethargic and displayed a mild left-sided hemiparesis. The patient had undergone evacuation of an acute subdural hematoma via a right frontoparietal craniotomy 10 years before this latest admission. Following this subdural surgery the patient had been neurologically normal. Computerized tomography (CT) scans obtained 1 year before this present episode revealed only slight compensatory enlargement of the frontal horn of the right lateral ventricle secondary to very mild focal cerebral atrophy. However, a CT scan obtained on admission (Fig. 1 left) revealed marked asymmetric enlargement of the lateral ventricles. The right frontal horn was larger than the left, and both distended frontal horns were filled with air.

First Operation. The putative diagnosis of tension pneumocephalus mandated that the patient undergo an emergency ventriculostomy. Surprisingly, the expected rush of
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Fig. 1. Axial CT scans obtained in a 78-year-old woman with spontaneous pneumocephalus. Left: Admission CT scan revealing intraventricular pneumocephalus. Right: Postoperative CT scan revealing the entry site of intraparenchymal air. Circle shows the bone defect at the tegmen tympani.

Fig. 2. Drawing showing middle ear procedures. Bone wax was used to block the eustachian tube. Following recurrence of pneumocephalus, the middle ear air space was completely obliterated with saline and a fat graft was placed in the tympanic membrane defect.

escaping air was not detected. A CT scan obtained the following day showed slightly more rather than less intraventricular air. The most dramatic difference was the presence of a new air-filled tract extending from the apex of the well-pneumatized left petrous bone through the substance of the brain into the temporal horn (Fig. 1 right). Overall the ventricular size had not changed, but the sulcal markings appeared somewhat more prominent. Nevertheless, the patient’s neurological status improved. The hemiparesis only lessened, but her mental status seemed to clear completely. In response to persistent questioning she denied any symptom related to either otorrhea or rhinorrhea. However, she did report that for many weeks she had heard noise in her left ear and felt as though there was a “bubble” in her head. She had made several trips to an otorhinolaryngologist because of her concern.

A subtemporal, extradural approach provided excellent exposure of a 4 × 2–mm defect in the tegmen tympani of the left petrous bone. The dura was adherent, but no dural defect was found. Bone wax, temporalis fascia, and fibrin glue were used to achieve a tight closure. The ventriculostomy was removed. A follow-up CT scan obtained at 6 weeks showed only a tiny residuum of intraventricular air.

First Postoperative Course. Six months postoperatively the patient presented again with the recurrent sensation of noise in her left ear and a feeling of “air in my head.” Examination at this time revealed no neurological deficits. However, a repeated CT scan once again demonstrated extensive intraventricular air. In an attempt to reveal the fistulous connection, the technicians insufflated the eustachian tube with air in a manner similar to a technique described by Pitts, et al.15 However, the results of the repeated CT scan were unchanged. Nevertheless, another small communication with the middle ear was believed to be the likely location of the fistula. Because these congenital defects are often multiple and repeated surgery of the dominant temporal lobe was an unacceptable option, an approach via the middle ear was selected.

Second Operation. A technique was devised to reduce the transmission of air pressure into the middle ear via the eustachian tube (Fig. 2). The eustachian tube was obliterated with bone wax via an anterior myringotomy to shield the middle ear from the nasopharynx. This blockage of the normal drainage pathway would be expected to cause fluid accumulation in the middle ear. Therefore, to preserve air conductive hearing, a tympanostomy tube was placed. A postoperative CT scan revealed a subtle decrease in the amount of air. After follow-up scans were planned, the patient was sent home.

Second Postoperative Course. Two weeks later the patient returned, complaining of disorientation and left-sided weakness. A CT scan revealed an increase in the pneumocephalus. We concluded that definitive sealing of the fistula could only be accomplished by excluding all air from the middle ear.

Third Operation and Postoperative Course. Surgical obliteration of the middle ear cleft and mastoid would induce severe hearing loss. Therefore, the patient was treated by removing the tympanostomy tube and filling the middle ear with saline (Fig. 2). The tympanic perforation was obliterated using an autologous fat graft. The patient has remained asymptomatic for a period of 12 months since the final surgery. Follow-up CT scans have confirmed the complete resolution of all intracranial air pockets. Audiometric evaluation demonstrated the presence of conductive hearing, although the sensitivity was slightly less than on the uninvolved side.

Discussion

The topic of pneumocephalus was extensively reviewed in Markham’s paper.13 Of the 295 cases, only two (0.6%) were thought to be spontaneous in origin, one of which was originally described by Jelsma and Moore.9 However, the postcraniotomy patients represented just 3.7% of the total. This reflects a detection bias because only plain
radiographs were available for diagnostic purposes. With the advent of CT scanning, as little as 0.5 cm³ of air may be detected,¹⁶ which has clarified the fact that postcraniotomy prevalence of pneumocephalus is nearly 100%. Thus, the true incidence of spontaneous pneumocephalus would be far less than 0.6%.

A thorough review of the literature has yielded only five additional cases.⁷,¹¹,¹²,¹⁷,¹⁸ When combined with Markham’s two examples and with this case report, the total number of reported cases of spontaneous pneumocephalus is eight (Table 1). Factors shared among this group include a defect in the bony roof of the air cells in the petrous bone and a pressure differential between the middle ear and the intracranial space.

The air cells surrounding the middle ear develop in both the petrous and mastoid components of the temporal bone. However, the number and size of these air cells varies between individuals.⁴ A progressive increase in pneumatization occurs with age. Åhrén and Thulin¹ found in an autopsy series in which they evaluated the thickness of the tegmen tympani that there is a continuum among individuals. Some specimens had a solid bone covering, others had “transparent” thin bone, and 21% had a clear bone defect in the tegmen tympani. Impressively, 6% of the specimens had more than five defects. These findings demonstrate that there is a wide variation in the thickness of the protective barrier between the intracranial compartment and the air cells surrounding the middle ear. Thus, the threshold for rupture of this structure would also be expected to differ among individuals.

In five of the eight patients in this group there was a definite episode of elevated pressure in the middle ear. Such pressure fluctuations are well tolerated in most individuals. However, the presence of a bone defect creates the opportunity for the development of a fistula. In the other three patients no episode of elevated middle ear pressure was documented. It may be that the necessary pressure differential for air entrainment was created by abnormally low or even negative intracranial pressure (ICP). There are many well-documented incidents of nonspontaneous pneumocephalus associated with shunt placement, presumably caused by low or negative ICP.⁵,⁶,¹⁴ This can be added to the long list of possible complications of overshunting, along with subdural hematomas in adults and slit ventricle syndrome in children. The case reported by Pitts, et al.,¹⁵ clearly illustrates the possibility of developing air-filled ventricles after shunt placement. The pneumocephalus in that patient was arrested by occluding the shunt, but recurred when the shunt was open.

Low or negative ICP is commonly understood to occur as a rare complication of excessive cerebrospinal fluid (CSF) removal, either via a shunt or as the result of posttraumatic or postoperative CSF leakage. In the absence of these factors, several theories have been advanced to explain the phenomenon of spontaneous pneumocephalus. Horowitz⁸ described an “inverted soda bottle” mechanism based on the Monroe–Kelly doctrine. This doctrine states that the intracranial volume is an absolute constant and that variations in any of the three components (brain, spinal fluid, and blood) must be compensated by reciprocal variations in the others. Otherwise the pressure in the intracranial compartment will change. Horowitz postulat-

**TABLE 1**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Presentation</th>
<th>Location of Air</th>
<th>Mechanism</th>
<th>Location</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jelsma &amp; Moore, 1954†</td>
<td>39</td>
<td>M</td>
<td>hemiplegia, syncope</td>
<td>rt occipital, extracranial, subdural</td>
<td>spontaneous</td>
<td>mastoid</td>
<td>needle aspiration</td>
</tr>
<tr>
<td>Markham, 1967</td>
<td>64, F</td>
<td>headache, aphasia</td>
<td>lt parietal subdural, intraparenchymal cyst</td>
<td>lt occipital epidural</td>
<td>altitude change</td>
<td>petrous</td>
<td>dural repair</td>
</tr>
<tr>
<td>Madeira &amp; Summers, 1977</td>
<td>57, M</td>
<td>homonymous hemianopsia</td>
<td>lt occipital epidural</td>
<td>airplane, nose blowing</td>
<td>mastoid</td>
<td>dural repair</td>
<td></td>
</tr>
<tr>
<td>Goldmann, 1986</td>
<td>26, M</td>
<td>headache</td>
<td>lt parietal subdural</td>
<td>scuba diving</td>
<td>nose blowing</td>
<td>tegmen tympani</td>
<td>observation</td>
</tr>
<tr>
<td>Stavas, et al., 1987</td>
<td>64, M</td>
<td>expressive aphasia</td>
<td>lt parietal subdural</td>
<td>mastoid</td>
<td>dural repair</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spar, 1994</td>
<td>27, F</td>
<td>headache, vomiting, CSF otorrhea</td>
<td>subarachnoid, intraventricular</td>
<td>spontaneous repetitive Valsalva maneuver</td>
<td>tegmen tympani</td>
<td>petrous</td>
<td>mastoid</td>
</tr>
<tr>
<td>Maier, et al., 1996</td>
<td>24, M</td>
<td>headache, visual scotoma</td>
<td>rt parietooccipital; epidural</td>
<td>mastoid</td>
<td>mastoid</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Present study</td>
<td>78, F</td>
<td>hemiparesis, aphasia</td>
<td>rt–lt intraventricular, lt temporal cyst</td>
<td>spontaneous</td>
<td>tegmen tympani</td>
<td>dural repair, endoscopic closure</td>
<td></td>
</tr>
</tbody>
</table>

* In all cases the pneumocephalus resolved with treatment.
† Also described in Markham’s report.

Fig. 3. Schematic drawings showing low-pressure pneumocephalus. When the patient is in the horizontal position, the pressure is similar throughout the CSF system. However, a vertical posture creates a pressure gradient within the system, the spinal segment distends, and the ICP decreases. When a defect in the dural covering is present, air may be drawn into the head.
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ed that negative ICP results from excessive loss of CSF either through settling into the distensible spinal subarachnoid space or simply drainage via normal pathways with physiological activity such as respiration or Valsalva maneuver. However, in the presence of a fistulous connection across the dura to an aerated sinus, air may enter the intracranial space in response to the negative pressure gradient (Fig. 3). If the brain substance was tightly adherent to the dura at the fistula site, the air could bypass the extracerebral spaces and penetrate the brain directly in the path of least resistance.

Walker and Vern described a variant mechanism. In this hydrodynamic model, the CSF loss and air entry occur discontinuously. The CSF compartment is not viewed as a fixed compartment, but rather as firm but yielding, as in Horowitz’s model. With the escape of fluid when the head is in the upright position, the sides of the compartment buckle inward until an equilibrium is reached. Then, when a recumbent position is assumed and the effects of gravity on the fluid are removed, the elastic side walls of the compartment expand to their usual conformation, sucking air into the subarachnoid space.

The accumulation of intracranial air is explained by the “ball valve” mechanism as described by Dandy. Air enters through the fistula when the pressure is lower inside the cranium than outside. The dural and bone defects are then tamponaded by brain tissue when the pressure differential across the opening is reduced. This prevents egress of the entrained air or leakage of CSF. In the presence of arachnoidal adhesions the air may be forced into the brain parenchyma. An intracerebral pneumoecele may form or the air may simply enter into and fill the ventricular system.

Our case is similar in many ways to the case reported by Pitts, et al. Both involved a fistula to the middle ear through a defect in the tegmen tympani. In their case the onset of the pneumocephalus was initially related to the placement of a ventriculoperitoneal shunt, whereas in our case the pneumocephalus was truly spontaneous. Negative ICP was most likely the cause of the pressure differential across the hole in the petrous bone in both cases. That interpretation is supported by the two observed episodes of increases in the amount of intraventricular air in our case; the first was after the initial ventriculostomy placement, and the second was after the eustachian tube was obliterated, which eliminated the possibility of pressure transmission from the nasopharynx. At that juncture the air must have been sucked in through the tube in the tympanic membrane.

In both cases a subtemporal extradural repair was only temporarily successful, and in both the ultimate solution was to exclude air from the middle ear. Pitts, et al., resorted to a radical mastoidectomy because their patient had no hearing in that ear. We avoided complete destruction of the middle ear by using the technique presented here.

Conclusions

Spontaneous otogenic pneumocephalus requires both a defect in the bone covering the middle ear and a pressure gradient across the defect. In some cases that pressure gradient is supplied by high pressure transmitted to the middle ear via the eustachian tube from the nasopharynx or directly through rupture of the tympanic membrane as a result of severe barotrauma. In the case reported here the pressure gradient appears to have resulted from abnormally low ICP. Closure of defects in the tegmen tympani may be possible with an extradural, subtemporal approach. If that approach fails, obliteration of the eustachian tube and exclusion of the middle ear may be required. The technique described here allows for preservation of bone conduction hearing. The option of electronic amplification is thereby retained.

References

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