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Case Report

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A Rare Case of Temporal Bone Pneumocephalus Tracking through the Internal Auditory Canal

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Keywords
Cranial base · Otology · Middle ear · Radiology

Abstract
A 39-year-old male with chronic hydrocephalus requiring biventricular shunts presented with progressive pneumocephalus over several years. He showed no improvement following ventriculoperitoneal (VP) shunt revision and anterior skull base repair for a sphenoid dehiscence. Imaging continued to show worsening pneumocephalus with air tracking along the right facial nerve from the geniculate ganglion to the internal auditory canal (IAC). The patient then underwent tympanomastoidectomy and skull base reconstruction. Based on a search of published literature, this appears to be the first reported case of temporal bone pneumocephalus coursing through the IAC, unlike most cases associated with tegmen defects and middle fossa pneumocephalus.

Introduction

Pneumocephalus is the accumulation of air in the epidural, subdural, subarachnoid, intraventricular or intraparenchymal space [1]. Common causes include postcraniotomy and post-traumatic skull defects, skull-base neoplasms and intracranial infection with gas-producing organisms [2].

Spontaneous otogenic pneumocephalus (SOP) requires a defect in the temporal bone leading to air inside the skull. Unlike other forms of pneumocephalus, SOP is not associated with neurosurgical procedures, trauma, or skull base neoplasms. Less than 30 cases have been reported in the literature [3]. Presentation varies from headache and gradual decline in neurologic status to otalgia and motor aphasia [4].

Case Presentation

A 39-year-old male with a history of intellectual disability, optic nerve fibrillary astrocytoma treated with pterional craniotomy, and chronic hydrocephalus treated with VP shunts presented with recurrent headaches and altered mental status. Computed tomography (CT) scan of the head showed pneumocephalus of the basilar cisterns and lateral ventricles (Fig. 1). His presentation was attributed to VP shunt malfunction, which was revised with return to baseline mental status.

He presented several years later with recurrent headaches, nausea and vomiting. Repeat head CT showed progressive pneumocephalus of the basilar cisterns and lateral ventricles with a clear dehiscence of the roof of the right sphenoid sinus (Fig. 2). Additionally, there was communication from the dehiscent wall of the

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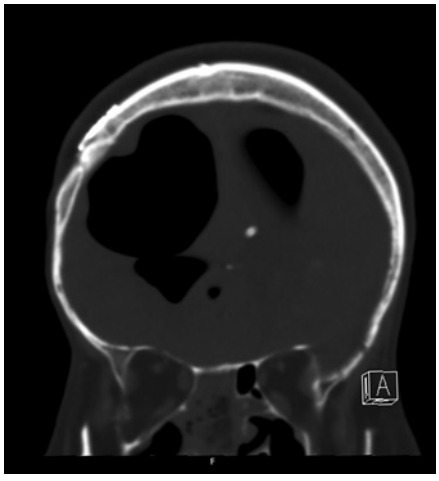

Fig. 1. Preoperative computed tomography (CT) image showing diffuse pneumocephalus of basilar cisterns and lateral ventricles.

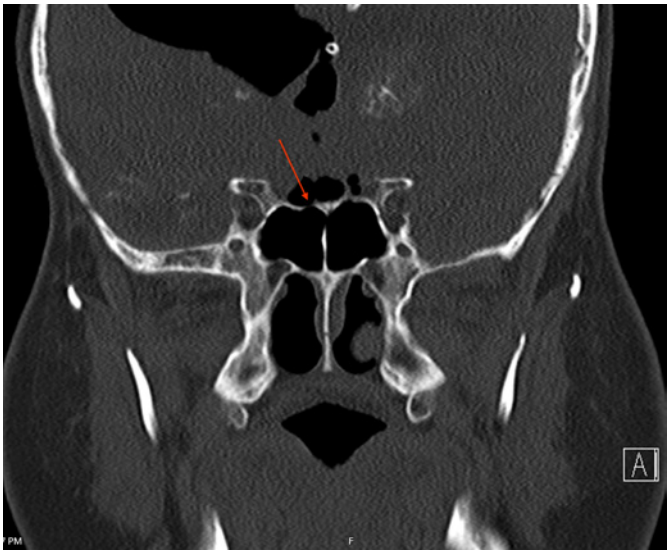

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Fig. 2. Preoperative CT showing right sphenoid sinus dehiscence (marked by a red arrow) and pneumocephalus of lateral ventricles and basilar cisterns.

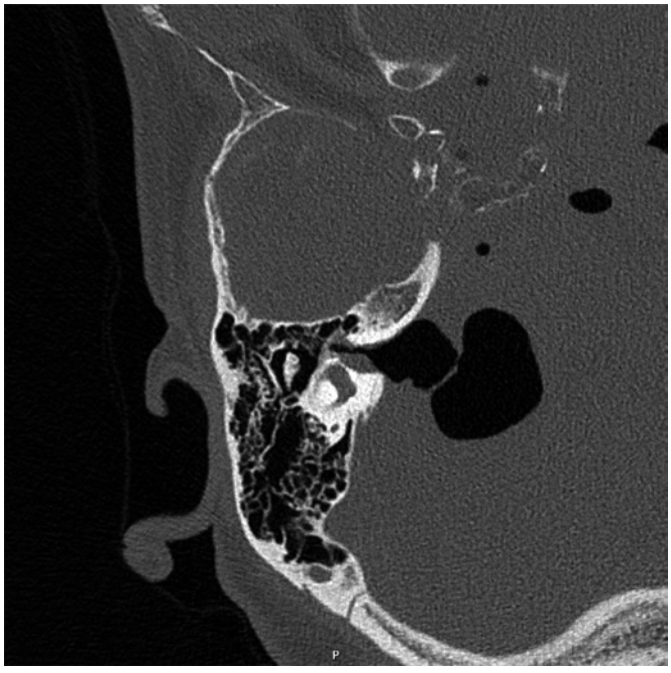


Fig. 3. Preoperative CT showing communicating pneumocephalus tracking from anterior genu of the right facial nerve canal into the internal auditory canal.

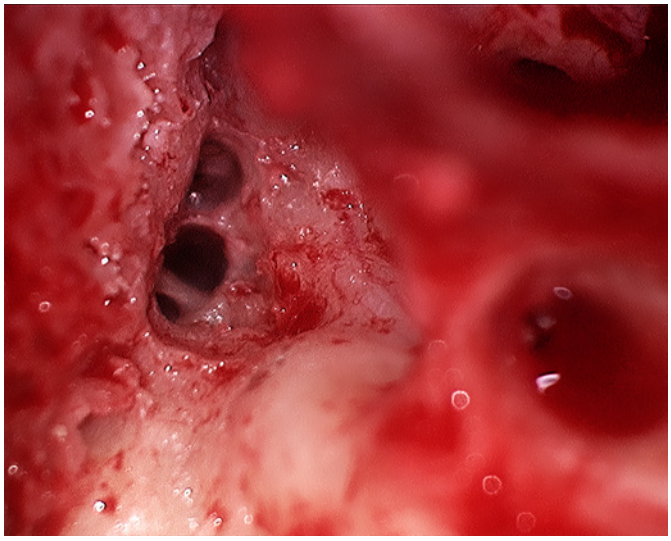
right facial nerve canal to the IAC and into the cranial vault. Because of the larger volume of air in the anterior skull base, this defect was repaired with a combination of a nasoseptal flap and fibrin glue.

A few weeks later, the patient presented with recurrent headache and vomiting. Head CT at this time showed further worsening of pneumocephalus with air seen tracking from the region of the anterior genu of the right facial nerve canal into the internal auditory canal and right cerebellar pontine angle cistern (Fig. 3). The patient was taken to the OR for right tympanomastoidectomy, skull base reconstruction, and obliteration of the right middle ear cavity, with the tympanic membrane removed. Intraoperatively, the middle ear and mastoid were found to be highly pneumatized with a large air cell tracking superiorly to the geniculate ganglion extending towards the petrous apex (Fig. 4). There was no evidence of an active cerebrospinal fluid (CSF) leak. The concerning air cell was packed with bone wax, the eustachian tube was packed with temporalis muscle, and the tegmen, anterior epitympanum, and eustachian tube were covered with bone cement followed by abdominal fat graft placement in the middle ear, epitympanum, and mastoid, and closure of the ear. The patient recovered well from the surgery and postoperative imaging showed the start of the resolution of the pneumocephalus (Fig. 5).

Discussion/Conclusion

SOP is caused by a fistula between the pneumatized temporal bone and the intracranial compartment without associated predisposing conditions [3, 4]. About half of previously reported cases have noted SOP triggered by increases in middle ear pressure, but many cases appear without an obvious trigger [5–9].

While typically asymptomatic, patients with pneumocephalus have been reported to present with headache,



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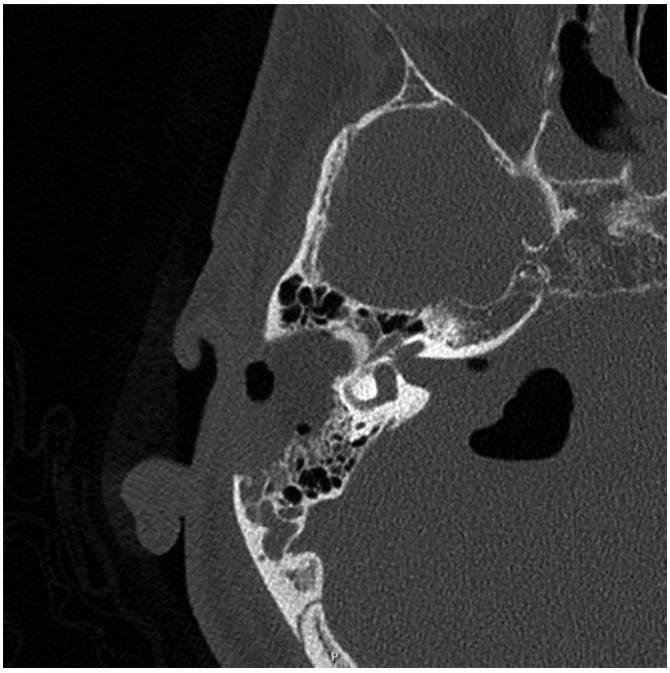

Fig. 4. Intraoperative photograph of the right ear showing air cell tracking superiorly to the geniculate ganglion extending toward the petrous apex.

Fig. 5. Postoperative CT imaging showing resolving pneumocephalus.

CSF rhinorrhea, postoperative lethargy, nausea, vomiting, seizures, altered mental status, hemiparesis and abducens nerve palsy [8]. Patients with otogenic pneumocephalus have also presented with otalgia, tinnitus and palpable retroauricular emphysema [4].

Surgical evacuation of intracranial air is the standard treatment for tension pneumocephalus causing mass effect. Dural repair is the most effective treatment for simple pneumocephalus with visible dural defects, but, given the rarity of this condition, there is no standard approach to surgical management. Since many patients with simple pneumocephalus are asymptomatic, conservative management and observation is also an acceptable option [10]. Shunt-associated pneumocephalus also raises the question of proper shunt management in addition to treatment of the dural defect. Half of previously reported cases of otogenic pneumocephalus were associated with VP shunt removal or ligation, while the rest chose to adjust settings or leave the shunt in place [11, 12].

Only 15 previous reports describe otogenic pneumocephalus associated with VP shunting [11, 12]. In all previously reported cases of otogenic pneumocephalus following shunt placement, the osteodural defect was located in the tegmen tympani or the petrous portion of the temporal bone [11, 12].

In this case, our patient had a remote history of optic nerve resection complicated by chronic hydrocephalus requiring biventricular shunt catheters, both factors that could potentially predispose him to pneumocephalus. The optic nerve resection may have left small subclinical skull base defects that were exacerbated by the subsequent hydrocephalus. Through these fistulous connections, air could have accumulated intracranially through the ball valve mechanism, consistent with his sphenoid sinus roof dehiscence. Following repair of his anterior skull base defects, however, his pneumocephalus did not resolve, raising the possibility of a superimposed SOP through the IAC, without an apparent inciting event. Drainage of CSF from the biventricular shunts could have led to negative intracranial pressure and worsening of the pneumocephalus due to the air trapping by the vacuum effect.

Previous reports of SOP have described skull base defects mainly in the tegmen tympani and posterior fossa [4, 11]. The location of this patient's pneumocephalus, with air tracking from the anterior genu of the facial nerve canal into the internal auditory canal and cerebellar pontine angle cistern, is highly unusual and, to the best of our

knowledge, has not been previously reported in the literature. Because this patient presented with simple, rather than tension, pneumocephalus, he was first managed conservatively with shunt adjustment, then, without resolution of symptoms, repair of the anterior skull base defects. Due to the extensive osteodural defects and residual pneumocephalus following anterior skull base repair, we chose to close the fistulous connections with bone wax, packing the eustachian tube with temporalis muscle, and covering the tegmen, anterior epitympanum, and eustachian tube with bone cement followed by abdominal fat graft placement in the middle ear, epitympanum, and mastoid.

Conclusion

This is a unique case of temporal bone pneumocephalus as a possible delayed complication of shunting for chronic hydrocephalus following post-surgical skull base defects. While rare, otogenic pneumocephalus should be kept in mind as a potential etiology of otherwise unexplained declining mental status in patients with history of VP shunt and skull base surgery.

Statement of Ethics

The study was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Written informed consent to publish this case report and accompanying images was obtained from the legal guardian of this patient.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

W.W. and E.M. performed the literature review and drafted the paper. A.K. and S.J.E. provided images, clinical history and critical revision.

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