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Profound Pneumocephalus and Low-Pressure Hydrocephalus Triggered by Ventriculoperitoneal Shunt Placement after Resection, Fat Graft Reconstruction, and Radiotherapy for a Malignant Skull Base Schwannoma

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Abstract

Background Tension pneumocephalus is a rare postoperative complication, typically presenting with mental status changes or rapid neurological decline after craniotomy. We report a complex case of tension pneumocephalus triggered by graft retraction after ventriculoperitoneal (VP) shunt placement.

Case History A 39-year-old woman with a recurrent left trigeminal cavernous sinus schwannoma, status post one prior resection, two stereotactic radiosurgery treatments, and one course of fractionated radiotherapy, underwent radical resection with orbital exenteration and abdominal fat free graft reconstruction followed by adjuvant radiotherapy for malignant transformation. She developed subacute ventriculomegaly with altered mental status, prompting VP shunt placement. Three weeks later, she presented with profound pneumocephalus and intraventricular air originating from a large, left-sided sphenoid and maxillary defect, from which the fat graft had retracted. A right frontal external ventricular drain (EVD) was placed, resulting in immediate release of air under high pressure. Definitive treatment required skull base reconstruction with a latissimus dorsi free flap, contralateral nasoseptal flap, antibiotics, and VP shunt revision for treatment of combined cerebrospinal fluid (CSF) leak, pneumocephalus, ventriculitis, and low-pressure hydrocephalus. As of her last follow-up, she was restored to her initial postresection neurological baseline.

Conclusion Tension pneumocephalus is a rare and life-threatening emergency that requires immediate neurosurgical intervention. We report the index case of tension

Keywords
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- ventriculoperitoneal shunt
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pneumocephalus induced by graft retraction following radiotherapy and CSF diversion. Where observed, tension pneumocephalus resulting from a skull base CSF leak may be associated with low-pressure hydrocephalus, and successful long-term management demands balancing the need for CSF diversion against the integrity of the skull base reconstruction.

Introduction

Pneumocephalus is a relatively common clinical phenomenon, frequently observed after craniotomy or craniofacial trauma.[1,2] Trauma accounts for approximately 75% of the pneumocephalus incidence, in particular after skull base fracture,[3,4] although it may also be observed as a sequela of routine neurosurgical procedures such as craniotomy, ventriculoperitoneal (VP) shunt placement, or lumbar puncture, among others.[3,5,6]

Although most instances of pneumocephalus are low risk and self-limiting, atypical instances of tension pneumocephalus arise from a one-way trapping of intracranial air without a natural release, resulting in a rapid and potentially life-threatening increase in intracranial pressure (ICP).[7] Tension pneumocephalus may occur as an acute or delayed complication of intracranial surgery, but is exceedingly rare overall, with a case incidence rate of approximately 0.1 to 0.2% among all craniotomies.[8] Less common still, scattered case reports have described tension pneumocephalus after endoscopic skull base or sinus surgery, or even VP shunt placement.[3,9,10] While most patients present with typical signs of elevated ICP such as headache, nausea, and vomiting, more severe cases may involve altered mental status, seizure, focal neurological deficits, and death.

We report a unique case of a complex skull base patient presenting with delayed tension pneumocephalus several weeks after VP shunt placement in the setting of radiotherapy, complicated by meningitis/ventriculitis and ultimately leading to the development of low-pressure hydrocephalus.

Case History

A 39-year-old woman presented with headache, dizziness, and near-syncope, worsening over several months. She had a history of a left trigeminal nerve schwannoma, status post one prior open subtotal resection, two stereotactic radiosurgery (SRS) treatments, and one course of fractionated radiotherapy, all performed at an outside facility. She presented to our institution for a second opinion after new disease progression was observed, but she was told that no further treatment could be offered (►**Fig. 1**). Our multidisciplinary skull base team offered repeat resection via lateral supraorbital craniotomy with orbital exenteration. Intraoperatively the tumor was noted to be invasive through the skull base into the sphenoid sinus; a gross total resection (GTR) was achieved, and a complex multilayer closure was performed using allograft for dural reconstruction and an abdominal fat free graft to occlude the skull base defect (►**Fig. 2**). An early postoperative

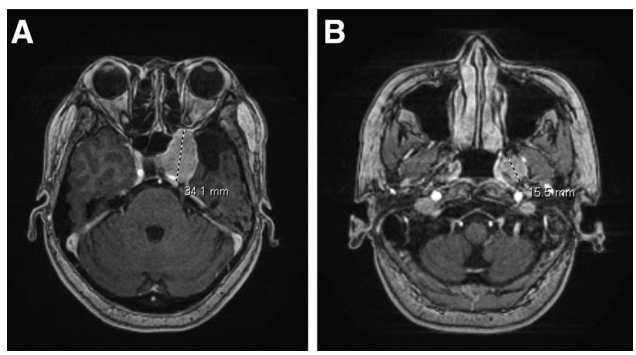

Fig. 1 Preoperative magnetic resonance imaging (MRI) demonstrating a fifth nerve schwannoma within the left middle cranial fossa centered at the cavernous sinus also involving Meckel's cave and left orbital apex. The lesion encases the left internal carotid artery (ICA) with mild to moderate narrowing of the petrous/cavernous segment. (**A**) The intracranial component measures up to 3.5 cm and (**B**) the infratemporal component measures approximately 1.5 cm. The lesion remodels the left sphenoid sinus and slightly displaces the pituitary gland. The left cisternal optic nerve course is along the margin of the tumor adjacent to the posterior communicating artery (PCOM).

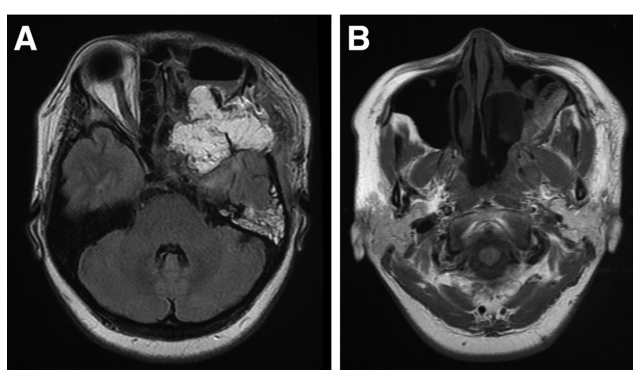

Fig. 2 (**A**, **B**) Postoperative magnetic resonance imaging (MRI) demonstrating postsurgical changes following left supraorbital craniotomy with left orbit exenteration and skull base lesion resection. Extensive fat grafting is seen along the left skull base and left orbital apex. There is layering fluid within the residual left orbit.

cerebrospinal fluid (CSF) leak was repaired via left nasoseptal flap, after which she recovered excellently without further issue for several months. Final pathology indicated transformation into a malignant peripheral nerve sheath tumor (MPNST), prompting adjuvant radiotherapy that was initiated at approximately 6 weeks after CSF leak repair.

While undergoing irradiation, the patient developed subtle but progressive mental status changes. An updated magnetic resonance imaging (MRI) was obtained, which demonstrated

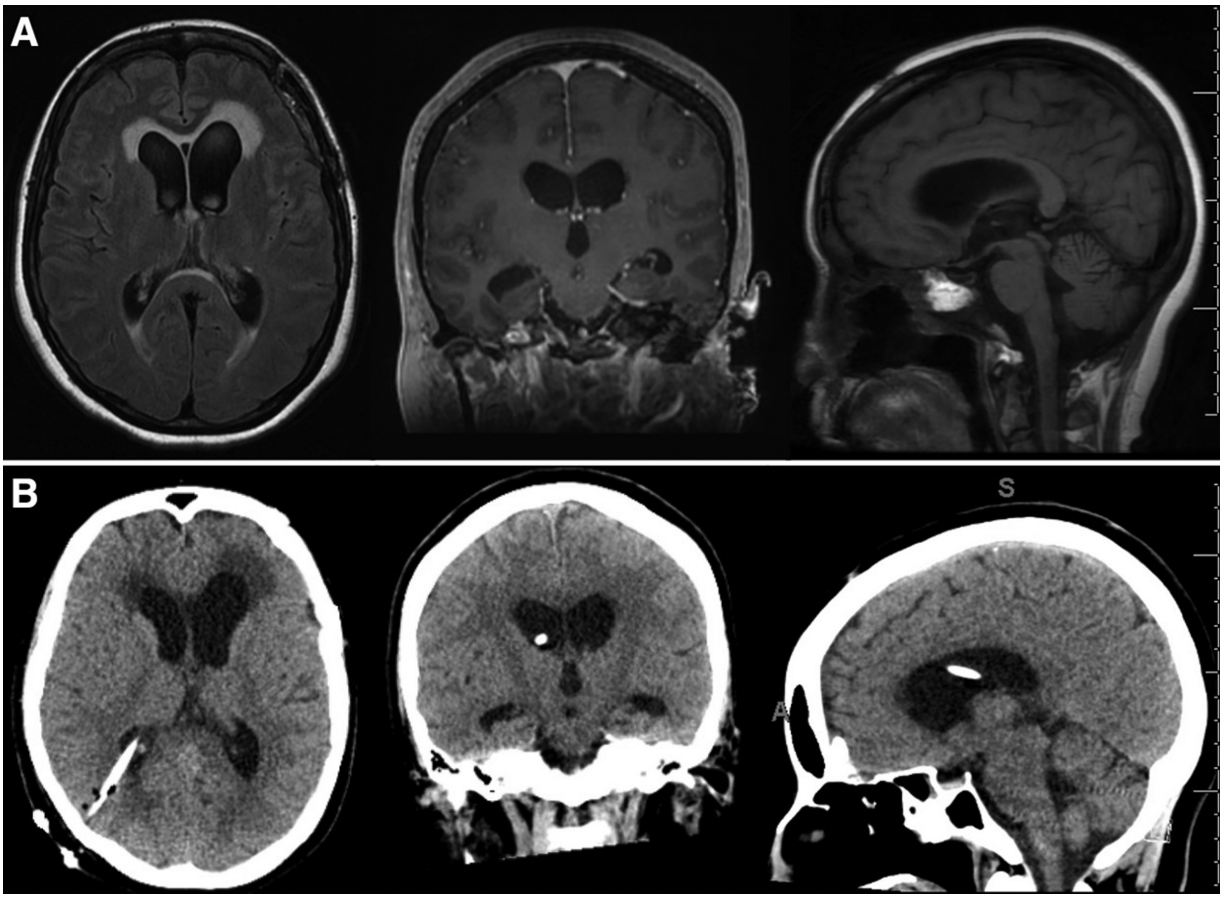

Fig. 3 (**A**) Magnetic resonance imaging (MRI) prior to ventriculoperitoneal shunt (VPS) placement demonstrating new triventricular hydrocephalus with periventricular edema. (**B**) Computed tomography (CT) status post VPS placement with unchanged lateral and third ventricle dilation.

marked ventriculomegaly, and placement of a right parietal ventriculoperitoneal (VP) shunt was recommended (►**Fig. 3**). Her mental status improved quickly after surgery and she was discharged home; however, she returned on postoperative day 10 with severely altered mental status, headache, imbalance, and persistent nasal drainage. Head computed tomography (CT) imaging revealed profound pneumocephalus and intraventricular air, as well as a large left-sided sphenoid and maxillary skull base defect from which the fat graft had retracted (►**Fig. 4**).

A right frontal external ventricular drain (EVD) was placed, resulting in immediate release of air under audibly high pressure. Subsequent CSF analysis confirmed ventriculitis, and the patient underwent a prolonged hospital stay that required long-term antibiotics and multiple interventions to balance the competing forces of low-pressure hydrocephalus and CSF leak.[11] Initial skull base repair with pedicled temporalis rotational graft retracted and failed when a new VP shunt was placed later that same admission; ultimately, the final reconstruction required a latissimus

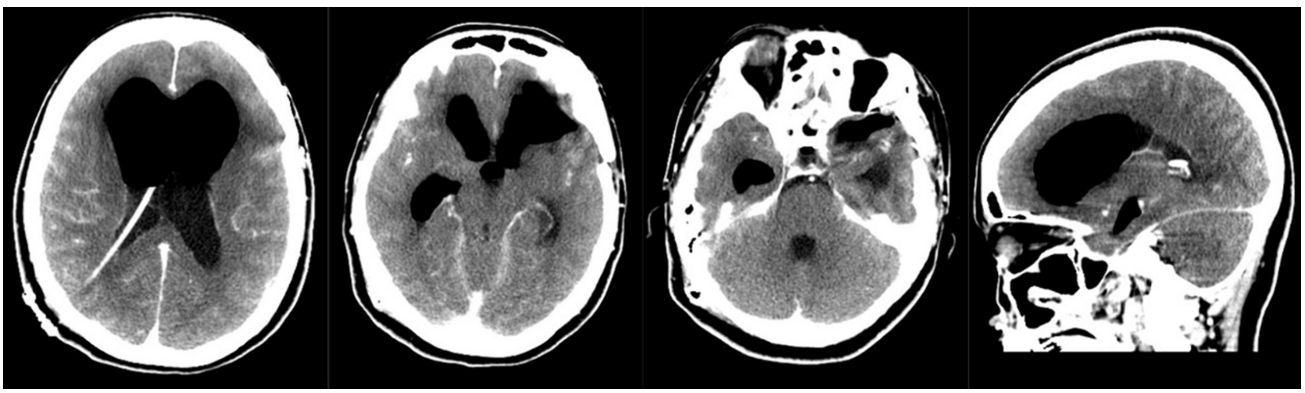

Fig. 4 Computed tomography (CT) with contrast obtained on postoperative day 10 demonstrating gas within the lateral ventricles laterally and the third ventricle, with ventricular dilation.

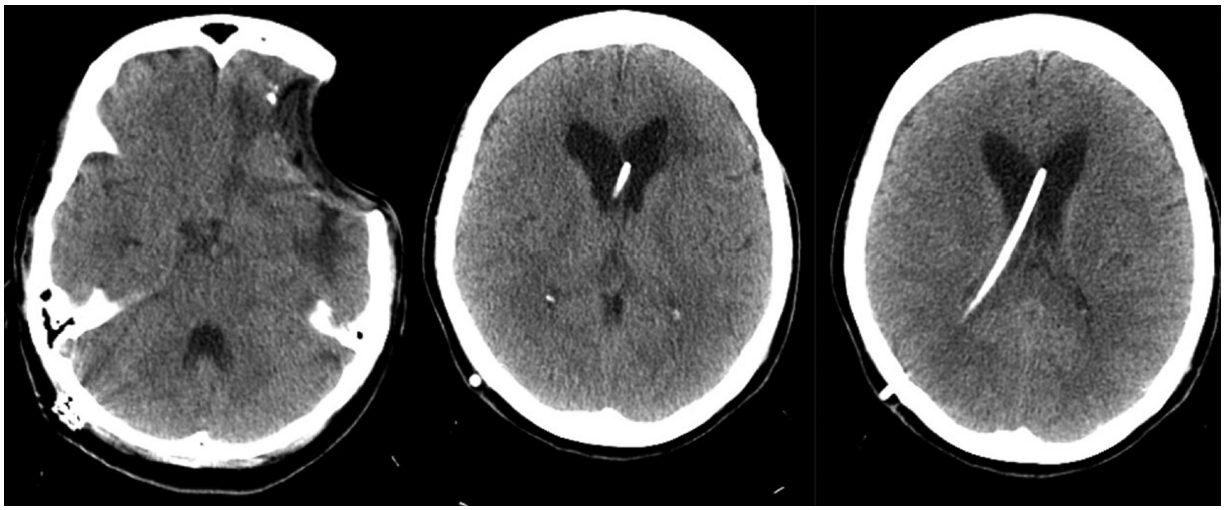

Fig. 5 Computed tomography (CT) scan demonstrating resolution of intraventricular air and stable size of shunted ventricles.

dorsi free flap for coverage of the nasal and orbital defects, inset via the craniotomy, pedicled to the superficial temporal artery and vein, and reinforced from below via contralateral nasoseptal flap. To minimize the risk of a third graft retraction and failure, the patient was maintained with bare-minimum CSF diversion via volume-titrated EVD management for 2 weeks prior to repeat VP shunt placement, which ultimately resulted in resolution of all intracranial air and restoration of normal neurological function.

As of last follow-up at approximately 6 months after surgery, the patient is at her neurological baseline, with preserved cognition, and sustained resolution of pneumocephalus without recurrent CSF leak or tumor recurrence (►**Fig. 5**). She successfully completed 31 of 35 radiation fractions prior to the onset of symptomatic pneumocephalus, and the decision was made to defer further treatment in that regard, unless new evidence of disease recurrence was noted on subsequent surveillance imaging.

Discussion

We report the first case of tension pneumocephalus secondary to skull base graft reconstruction after CSF diversion in the setting of radiotherapy. This unusual case required iterative multidisciplinary management to reach an ideal outcome for the patient, and showcases both an extreme instance of the CSF disorders that may arise after skull base surgery and the complex interventions that are potentially required to manage these patients.

Two major models have been proposed to conceptualize the pathophysiology of tension pneumocephalus. The first is called the "*inverted-soda-bottle effect of Horowitz and Lunsford*," in which excessive CSF loss from a postoperative CSF leak leads to a relatively negative ICP gradient, which in turn draws and traps air via vacuum effect through the craniotomy defect.[12] The second theory, known as the "*ball valve theory of Dandy*," posits that pneumocephalus results from the unidirectional movement of air from the outside environment into the cranial cavity, trapping it within like a ball valve mechanism.[13,14] While typical pneumocephalus is often an expected complication of craniotomy that eventually resolves, tension pneumocephalus is associated with significant morbidity and mortality if not rapidly treated.

A wide range of underlying etiologies have been described for tension pneumocephalus, including craniofacial trauma, skull base or sinus surgery, distant CSF leak, VP shunt placement, and even penetrating lumbar trauma.[5,6,15,16] Still other iatrogenic causes have been reported in scattered cases, including the positive pressure ventilation such as during continuous positive airway pressure therapy, administration of nitrous oxide, cranial infections such as osteomyelitis or meningitis, and hyperbaric oxygen therapy.[17–21] The severity of tension pneumocephalus varies. Mild symptoms may include headache, nausea, vomiting, and dizziness, while more severe cases yield restlessness, agitation, delirium, seizures, altered mental status, and ultimately comatose status.[16] Tension pneumocephalus impacting the posterior fossa is very rare but particularly dangerous, and may result in rapid respiratory failure, coma, and death.[22]

The gold standard for diagnosing pneumocephalus is a noncontrast head CT, given its very high specificity, rapidity, and ready access. Air has a Hounsfield coefficient of –1000; correspondingly, CT studies will detect as little as 0.55 mL of intracranial air, whereas a minimum of 2-mL air is needed to appreciate pneumocephalus on plain film radiographs—oftentimes significantly more. Key radiographic indicators of possible tension pneumocephalus were initially described by Ishiwata in 1988.[15] The geo-eponymous "Mt. Fuji sign" refers to intracranial air in the frontal region separating the tips of the frontal lobes, creating the silhouettelike appearance of Mount Fuji (►**Fig. 6A**).[23] This is very recognizable, and although not always clinically significant, should prompt a high index of suspicion if encountered on postoperative imaging—in particular for an altered patient. An important distinction from routine pneumocephalus is the marked prominence and separation of the bifrontal poles, which

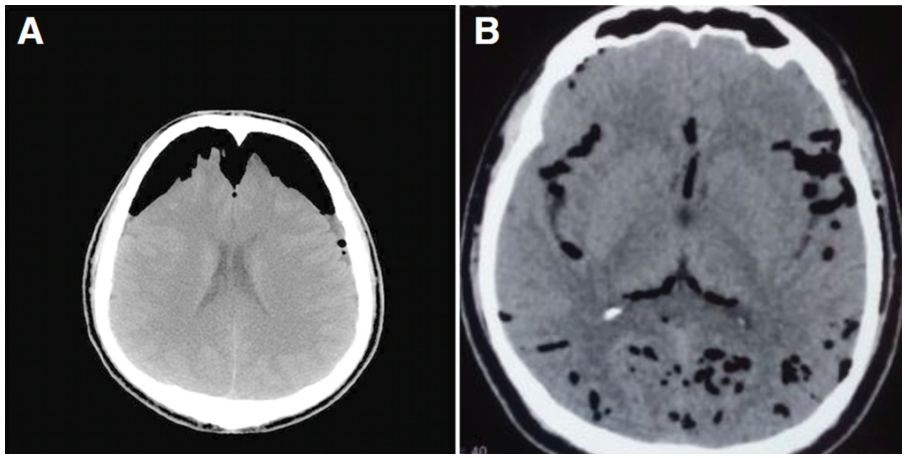

Fig. 6 Pathognomonic radiographic signs of pneumocephalus. (**A**) Mt. Fuji sign: frontal region air accumulation separating the tips of the frontal lobes, giving the appearance of the silhouette of Mt. Fuji. (**B**) Air bubble sign: air bubbles in various cisterns and subarachnoid space.

appear sharp and separated, rather than broadly compressed by a more nonpressurized frontal collection. The second is called the "air bubble sign," which describes widespread scattering of numerous air bubbles throughout the intracranial compartment, often involving essentially all lobes of the cerebrum diffusely (►**Fig. 6B**).

Once diagnosed, tension pneumocephalus requires emergent neurosurgical evaluation and intervention to prevent permanent neurological injury—more specifically, by releasing the air. Decompression options include needle puncture through an existing burr hole or craniotomy margin,[24] emergency creation of a new burr hole with a bedside drill, decompressive craniectomy,[25] ventriculostomy, or placement of a subdural evacuating port system.[26] Once the air has been released, attention can be turned to identifying the underlying etiology, and performing a definitive treatment in that regard.

Conclusion

We report a novel case of tension pneumocephalus in the setting of skull base graft reconstruction after CSF diversion in a patient undergoing skull base radiotherapy. Our patient presented with severe symptoms, requiring emergent treatment with EVD placement for release of pressurized intraventricular air, followed by a complicated, multistep, multidisciplinary treatment strategy that ultimately incorporated neurosurgery, rhinology, oculoplastics, and plastic surgery. Although tension pneumocephalus is quite rare, it may be provoked by a wide range of mechanisms, essentially all of which require urgent or emergent treatment. Given the rarity of the condition, it is challenging to generalize about preventing tension pneumocephalus; however, neurosurgeons should be aware of the entity and approach postoperative skull base patients with a healthy degree of suspicion, given the life-threatening nature of the disease, and the relatively straightforward and effective nature of the frontline interventions for immediate pressure relief and patient stabilization. Notwithstanding, definitive diagnosis and treatment of the underlying cause may be quite complex, in particular given the significant potential for secondary disease sequelae such as infection or low-pressure hydrocephalus, and definitive treatment may require the full suite of skull base strategies.

Conflict of Interest
None declared.

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