Unusual and Rare Pneumocephalus Presentations in a Tertiary Care Center: Management Strategies and Review of Literature

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Pneumocephalus has various forms of presentations and a diverse etiology ranging from trauma to iatrogenic craniotomies and spinal surgeries. Most cases are benign and uncomplicated and resolve with minimal or no intervention. Few cases of pneumocephalus or pneumoventricles are either persistent despite treatment or present in various unique forms leading to considerable difficulty in the diagnosis and management of these patients. In this study, we analyze six cases of pneumocephalus with unusual and rare form of presentations and discuss their management strategies along with a review of relevant literature.

Abstract

Keywords

► pneumocephalus
► pneumoventricles
► management

Introduction

Pneumocephalus is a common neurosurgical entity that may either occur per se or associated with other cranial and intracranial pathologies. The etiologies include trauma, being the most common,1,3 as well as other nontraumatic conditions, including iatrogenic causes, seen commonly after neurosurgical interventions like craniotomies. Most pneumocephalus are benign, needing minimal intervention and resolve without any major complications. In few cases, they may increase in size and become relentless producing considerable symptoms and signs requiring treatment varying in spectrum from simple nasal oxygenation to emergency surgical decompression. Rarely, pneumocephalus may also assume strange bizarre forms and proportions providing a considerable management dilemma and a therapeutic challenge to the treating neurosurgeon. Despite contributing significantly to the morbidity, the problem of pneumocephalus tends to get severely underrated. Current neurosurgical literature regarding pneumocephalus is mainly limited to case reports and few small case series and lacks any comprehensive or exhaustive literature reviews focusing on the disease burden, presentation, and management options. We have highlighted six unusual and rare presentations of pneumocephalus in our institution, each with a unique mode of presentation and provided unique management challenges. We also review the relevant literature regarding optimal management strategies for pneumocephalus.

Materials and Methods

We have illustrated six cases of pneumocephalus with unusual presentations and discuss their management strategies. All these patients were admitted in our institution, a tertiary care center, between June 2018 and February 2019.

Case Reports

Case 1

A 55-year-old man was admitted with road traffic accident (RTA) while riding a two-wheeler. He had nasal bleed and Glasgow Coma Score (GCS) 15/15. Computed tomography (CT) brain (►Fig. 1A) showed frontoorbital fracture with frontal contusion. Nine hours later (►Fig. 1B), the patient had worsening headache with altered sensorium. Repeat CT brain showed a delayed new-onset bifrontal tension pneumocephalus. He was managed conservatively with intermittent 100% nasal O2 at 6 L/min and head end of bed elevated to 30 degrees. He symptomatically improved over the next 4 days and follow-up CT brain showed resolving pneumocephalus (►Fig. 1C).
Case 2
A 30-year-old man had RTA and was admitted with multiple craniofacial injuries and cerebrospinal fluid (CSF) rhinorrhea. CT brain showed bilateral frontoorbital and nasoethmoid complex fractures with small frontal pneumocephalus. He was managed conservatively and discharged after symptomatic improvement and resolution of pneumocephalus. Two months later, he presented with progressive left hemiparesis, headache, and vomiting. CT brain showed a large right frontal pneumocephalus with mass effect and midline shift >5 mm toward left side (►Fig. 2A). He underwent craniotomy and evacuation of pneumocephalus followed by anterior cranial fossa repair. Postoperatively, there was complete symptomatic resolution and scan showing no residual pneumocephalus (►Fig. 2B).

Case 3
A 20-year-old man with RTA was admitted elsewhere with craniofacial injuries. A diagnosis of bifrontal pneumocephalus with bilateral frontal horn pneumoventricles (►Fig. 3A) was made and he was conservatively managed for 20 days. His headache worsened and he was referred to our institution. CT brain on admission showed resolved pneumocephalus but the pneumoventricles persisted. He was continued on conservative management but on the day 7, his headache worsened and had altered sensorium. CT brain revealed large bifrontal tension pneumoventricles (►Fig. 3B). He underwent emergency burr hole and release of pneumoventricles with an under-water sealed external ventricular drain (EVD). EVD was removed on the postoperative day 7 and follow-up serial CT scans showed gradual resolution of pneumoventricles (►Fig. 3C, D). The patient had a protracted postoperative course with symptomatic resolution over a period of 2 weeks.

Case 4
A 40-year-old male presented with gait disturbance and altered sensorium. He was previously diagnosed with colloid cyst third ventricle and underwent endoscopic excision 1 month before. CT brain on admission showed bifrontal tension pneumocephalus with bifrontal pneumoventricles (►Fig. 4A). Initially conservative management was attempted, but since there was no neurological improvement, he underwent burr hole and evacuation of tension pneumocephalus. Postoperatively, he was administered nasal oxygen and nursed in a head-up position. Follow-up scans (►Fig. 4B) showed gradual resolution of pneumoventricles also. Patient had good recovery postoperatively and was discharged after 10 days.

Case 5
A 2-year-old female child, who was a diagnosed case of spinal dysraphism with tethered cord syndrome and underwent...
Lumbar (L 1 and 2) laminectomy and detethering of the cord, had an episode of tonic seizure lasting a few seconds on the first postoperative day. CT brain showed the presence of pneumocephalus in the high-parietal interhemispheric region (Fig. 4C). The child was started on antiepileptic medications and had no further seizures. Follow-up CT scan showed complete resolution of the pneumocephalus (Fig. 4D).

Case 6
A 26-year-old male who had undergone C1–2 fixation for atlantoaxial dislocation, presented 2 weeks later with worsening headache, one episode of seizure, and CSF leak from the operative wound site. CT brain showed a bizarre pattern of pneumocephalus with diffuse, bilateral multiple air pockets involving both cerebral hemispheres and all the ventricles (Fig. 5A–C). Pneumocephalus was managed conservatively and gradually resolved over a period of 2 weeks, though he later underwent surgical repair for persistent CSF leak.

Results
In our series, cases 1, 2, and 3 presented with a delayed-onset pneumocephalus, each with a differing time of onset. The initial CT brain of case 1 did not show any evidence of air but subsequent CT after 8 hours showed a tension pneumocephalus. Case 2 presented with a massive pneumocephalus with hemiparesis 2 months after the initial trauma, and needed surgical intervention. Case 3 had persistent bifrontal pneumoventricles despite conservative management for 4 weeks and then proceeded to become tension pneumoventricles, requiring emergency surgery. Case 4 developed pneumocephalus following endoscopic surgery, which progressively increased in size producing altered sensorium which required surgical intervention. Cases 5 and 6 are rare complications that occurred following spinal surgeries, with the former presenting with seizures. Both cases were managed conservatively for pneumocephalus, with the latter eventually requiring surgery for persistent CSF leak. All the above patients improved following treatment and were
Discussion

The above examples illustrate the vagaries of pneumocephalus presentations. By definition, pneumocephalus is “air within the cranial cavity.” It varies from simple isolated air pockets to life-threatening subdural, subarachnoid, intraparenchymal, or intraventricular air. Such cases are termed “tension pneumocephalus” or “tension pneumoventricles” depending on the location of air. It is commonly due to one-way entry of air causing progressive accumulation within the cranium and producing mass effect. Even though trauma is the most common cause of pneumocephalus, various other etiologies include surgical procedures like craniotomies and, rarely, spinal surgeries. Also, the presentation can also be acute or delayed.

The most common route of air entry into the cranial cavity is via a breach in the dura and communication with the external environment, as in the cases of many traumatic basal skull fractures. Air enters the subdural space and gets collected most commonly in the frontal region compared with other locations. This may be due to two reasons: first, the patient’s supine position with the frontal region being the highest point may contribute to this; and second, the dura is comparatively thinner and more attached to the bone in the frontal region. When there is also a breach in the arachnoid layer in addition to the dura with or without CSF fistulas, the air gets trapped in the subarachnoid spaces and ventricles, producing pneumoventricles. Penetrating brain injuries and gas-forming organism infections like clostridium may produce intraparenchymal pneumocephalus.

Two mechanisms have been proposed to explain the pathogenesis of tension pneumocephalus or tension pneumoventricles: Walter Dandy, in his pioneering work on
pneumocephalus in 1926 described the so-called “Ball-valve mechanism.” According to Dandy, during maneuvers that cause rapid increase in intracranial pressure like coughing, sneezing, etc., there occurs one-way entry of air inside the cranium through a communicating dural breach, causing air to get trapped within cranium. Based on the observations of Horowitz in 1964, Lunsford et al in 1979 proposed the “Inverted soda bottle effect,” like in cases of CSF fistula, where continuous CSF loss creates a negative pressure that sucks in air, producing progressive air accumulation. The latter may also account for the development of pneumocephalus in some patients, after surgery in a sitting position. In rare instances, pneumocephalus can also occur following spinal surgeries when the dura or arachnoid is breached. Such patients may present with various symptoms like headache, seizures, and confusional states.

The clinical manifestations of pneumocephalus are extremely variable but the most common symptom is headache followed by altered sensorium and seizures. Neurological deficits like motor weakness or cranial nerve palsies are rare. Meningitis may occur, particularly when associated with CSF leak. The single sign that is unique to pneumocephalus is “Bruit Hydro-aérique,” similar to the succussion splash that is seen in the abdomen. It is due to the movement of fluid in an air-filled cavity on postural change and can be heard by both patient, as well as the examiner. According to Dandy, air is an irritant and induces inflammation and edema in the surrounding brain parenchyma, thereby resulting in seizures.

The investigative modality of choice is CT, as it can detect even 0.5 cc of air. The hypodensity (Hounsfield’s unit of 1,000 for air) seen in CT scan is unmistakable and aids in its identification. Though it is radiologically difficult to interpret whether the pneumocephalus is subarachnoid or subdural, the former usually produces a “diffuse” pattern of pneumocephalus, whereas the latter is more “localized” and presents more commonly in the frontal region. Also, the presence of pneumoventricles favors a subarachnoid location of air. An important sign, “Mount Fuji Sign” or “twin-peak sign,” described by Ishiwata et al is classical of tension pneumocephalus, where air under tension compresses both the frontal lobes and also causes separation of both lobes by accumulating in the interhemispheric region, creating the characteristic silhouette of two peaks. This should be differentiated from the less severe “Peaking sign,” where there is no separation of the frontal lobes.
Most pneumocephalus, either subdural or subarachnoid, get absorbed spontaneously. Specific treatment is required only when the volume of intracranial air is >25 cc or if the patient is symptomatic. General measures consist of bed rest, nasal oxygen, avoidance of nose blowing, or coughing and analgesics.\(^7\) The optimal head position is still debatable at present with many authors favoring a flat lie and some even advocating a Trendelenburg’s position. But the consensus is generally to keep the head end elevated to approximately 30 degrees above horizontal. The next definitive management is the administration of nasal oxygen. Inhalation of oxygen decreases the concentration of nitrogen in the blood thereby creating a negative gradient and causes nitrogen (major constituent of intracranial air) in the pneumocephalus to rapidly diffuse into the bloodstream and get exhaled out. Again, there is no consensus regarding the volume, time, duration, and mode of oxygen therapy. Dexter and Reasoner in 1996, in a theoretical study concluded that, the use of normobaric 100% oxygen therapy at FiO\(_2\) 40% for 1 week considerably decreased the pneumocephalus and higher volumes of pneumocephalus required oxygen at higher inspiratory pressures.\(^8\) Gore et al advocated the use of normobaric 100% oxygen therapy at higher FiO\(_2\) at 68% for approximately 24 hours and also concluded that the use of oxygen mask was superior to the nasal cannula while treating patients with pneumocephalus.\(^9\) Some authors recommend the use of hyperbaric oxygen instead of normobaric oxygen,\(^10\) while some others question the validity of using hyperbaric oxygen therapy in the treatment of pneumocephalus,\(^11\) though a consensus is yet to be reached in this regard.

Even though there are rare reports of large asymptomatic massive pneumocephalus resolving with conservative management, surgical intervention should be considered for the following:

1. Pneumocephalus that does not reduce in size after 2 weeks of conservative management.
2. Tension pneumocephalus or tension pneumoventricles.
3. Worsening of existing symptoms or new onset neurological deficits or altered sensorium.
4. CSF fistula with active leak.

Surgical options include burr hole or craniotomy and evacuation of the air and filling the cavity with saline. An under-water external drain is preferable following burr hole evacuation of air. Site of CSF leak should be identified and dural defects should be closed in a meticulous water-tight fashion using fascia or other dural substitutes. Exposed air sinuses should be completely exteriorized, mucosa excised, and sinus cavity filled with muscle patch or covered with vascularised pericranial flap. Specific meningitic prophylaxis may be considered in the presence of active CSF leak.

**Conclusion**

Pneumocephalus can have myriad presentations ranging from trivial air pockets to massive air collections that may be life-threatening. They may not show up immediately but may present weeks to months after an initial trauma or surgery, as shown in our examples. Also, bizarre and unusual radiological patterns of pneumocephalus are not uncommon and can be therapeutically challenging for the neurosurgeon. This may lead to a potential delay in decision making regarding the optimal management strategy in these patients. Understanding the cause and mechanism of pneumocephalus helps a long way in making a correct decision in treating such patients, either medically or surgically. Prompt timing of surgical intervention when indicated, plays a major role in the recovery of these patients.

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**Conflict of Interest**

None declared.

**References**