Pneumocephalus Due to Invasive Fungal Sinusitis

Pneumocephalus is an accumulation of intracranial air that occurs when there is a connection between the intracranial space and the extracranial space. Rarely, tension pneumocephalus develops—an accumulation of air through a one-way valve that causes dramatic neurological deterioration. Two proposed mechanisms are a ball-valve effect—where air is pulled into the intracranial space during coughing, swallowing, or straining—and an inverted-bottle effect—where excessive leakage of CSF leads to negative intracranial pressure and replacement of fluid with air [1]. We present a unique case of pneumocephalus secondary to a fungal infection.

A 77-year-old man with a history of steroid-dependent autoimmune hemolytic anemia and myelodysplastic syndrome was admitted to the hospital with a 1-day history of altered mental status. He was somnolent and withdrew only from painful stimuli. His vital signs were stable except for a respiratory
Figure 1. A. Head CT scan of a patient with pneumocephalus secondary to invasive fungal infection at time of admission. Marked pneumocephalus surrounds frontal lobes anteriorly. B. Smear of purulent material from patient’s frontal sinus that shows fungal hyphae (Paragon stain; original magnification, ×1000).
rate of 33. Physical examination demonstrated CSF rhinorrhea. There were no signs of head trauma.

The WBC count was 22,400/µL, hematocrit was 15.6%, and platelet count was 26,000/µL. A head CT scan revealed marked pneumocephalus surrounding the frontal lobes (figure 1A). There was no intracranial hemorrhage, mass lesion, or fracture.

The patient’s neurological status deteriorated, and he developed aspiration pneumonia. The family requested comfort care, and the patient subsequently died.

An autopsy confirmed massive pneumocephalus with compressive narrowing of the anterior cerebrum. Destruction of the inner wall of the right frontal sinus by extensive fungal osteomyelitis allowed air to enter the cranium. The fungus was not grown in culture because of an inadequate specimen, but microscopic examination of purulent material from the frontal sinus showed septate hyphae of uniform width with dichotomous branches at 45° angles (figure 1B), findings consistent with Aspergillus species.

In 1967 Markham [1] evaluated 295 cases of pneumocephalus that dated back to 1884. The most common cause of pneumocephalus was trauma (74% of cases), followed by neoplasm (13%). Infection accounted for 9% of cases, and surgery accounted only for 4%; however, invasive neurosurgery is more common today. Three of the infectious cases resulted from sinusitis, whereas most of the infectious cases were due to chronic otitis media. Further review of the literature revealed several other cases related to otitis media [2] and a few due to meningitis caused by gas-forming bacteria [3]. To our knowledge, no cases of pneumocephalus secondary to invasive fungal infections have been reported.

Aspergillus is the most common cause of fungal sinusitis [4]. Other etiologies include Fusarium species, Bipolaris species, Curvularia lunata, Pseudallescheria boydii, Rhizopus arrhizus, Cunninghamamella bertholletiae, and several other sporadically identified fungi [5]. Invasive sinus aspergillosis occurs predominantly in immunocompromised patients but can occur in immunocompetent hosts [6]. Invasion into the brain is a well-documented complication of fungal sinusitis [7]. Although not previously documented, this complication may create a bone defect that sets the stage for the development of pneumocephalus.

The diagnosis of invasive fungal disease is difficult and requires histopathologic examination that demonstrates the characteristic hyphae. Invasive aspergillosis is very difficult to treat, even with amphotericin B and surgical intervention. The mortality rate associated with this condition is >50%, and is even higher when there is intracranial involvement [4, 7].

In conclusion, pneumocephalus is uncommon and is usually seen after head trauma or surgery. We present a unique case in which a frontal sinus infection, probably due to Aspergillus species, led to symptomatic or tension pneumocephalus. Although uncommon, infectious etiologies including fungal disease should be considered when pneumocephalus is discovered, especially in immunocompromised patients.

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References