Case report

Delayed spontaneous pneumocephalus in ventriculoperitoneal shunting: Two case reports and literature review

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ABSTRACT

Spontaneous pneumocephalus following cerebrospinal fluid shunt is a rare complication. In most cases, the air enters in the intracranial cavity via a skull base defect. We report 2 cases of delayed tension pneumocephalus, secondary to ventriculoperitoneal shunt, and review the etiopathogenesis, prevention and treatment of this condition.

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Neumoencéfalo espontáneo tardío posterior a la colocación de válvula ventriculoperitoneal: 2 casos clínicos y revisión de la literatura

RESUMEN

El neumoencéfalo a tensión es una rara complicación después de la colocación de sistemas de derivación de líquido cefalorraquídeo. En la mayoría de casos la etiopatogenia está relacionada con un defecto de la base del cráneo. Presentamos 2 casos de neumoencéfalo tardío a tensión después de la colocación de derivación ventriculoperitoneal y revisamos la literatura, analizando los mecanismos de etiopatogenia, así como las posibles formas de prevención y tratamiento.

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Introduction

Pneumocephalus after shunting may be immediate, occurring in the first few days after shunt placement, or delayed, appearing months or years following ventriculoperitoneal (VP) shunt insertion.\(^1,^2\) This second scenario is more unusual and, because of the lack of a temporal relationship between the VP shunt insertion and the pneumocephalus appearance, the causal diagnosis may be much more elusive.

We are reporting two new cases of delayed spontaneous pneumocephalus, which developed 1 and 5 years after VP shunt placement. We review the literature concerning the physiopathology and management, as well as possible approaches to prevention of this condition.

Case reports

Case 1

A 65-year-old man with a history of subarachnoid hemorrhage secondary to right paraophthalmic aneurysm that was treated endovascularly. The patient's condition was complicated by a communicating hydrocephalus that required a VP shunt. One year later the patient was readmitted complaining of headache, drowsiness, gait disturbance and progressive language impairment.

The cranial CT scan showed pneumocephalus in the frontal and ventricular regions (Figs. 1 and 2). Further assessment with high-resolution bone-window CT did not detect any defects at the skull base.

The shunt was clamped and the pneumocephalus was evacuated. Finally, a programmable anti-siphon valve was implanted, with the opening pressure set at 150 mmH\(_O\). Two weeks later the patient was discharged with residual hemiparesis but with complete resolution of the pneumocephalus (Fig. 3). There has been no recurrence during the following 18 months.

Fig. 3 – Follow-up CT scan with resolution of the pneumocephalus.

Case 2

A 64-year-old man, with a history of post-meningitis hydrocephalus treated with ventriculoperitoneal shunt 5 years previously (Fig. 4), was admitted to our department presenting with headache, nausea, vomiting and seizures. A right frontal basal pneumocephalus was observed in the CT scan (Fig. 5). The pneumocephalus was evacuated and the ventriculoperitoneal shunt was removed due to infection.

After these procedures, a CT scan reconstruction of the skull and cisternography were carried out. Although no cerebrospinal fluid fistula was detected, a probable skull defect at the level of the frontal sinus was observed in the skull CT reconstruction (Fig. 6). Therefore, the anterior cranial fossa was examined surgically and the skull base defect was confirmed and sealed.

Figs. 1 and 2 – Cranial X-ray and CT showing bifrontal tension pneumocephalus causing significant ventricular compression and midline shift.
A programmable anti-siphon valve was implanted, with the opening pressure set at 120 mmH₂O. The tension pneumocephalus resolved completely and the patient was discharged 3 weeks later with no new neurological deficit (Fig. 7).

Discussion

Spontaneous pneumocephalus related to a VP shunt has been attributed to the coexistence of two conditions: the siphon effect of a shunt diminishing intracranial pressure, and a skull base defect¹⁻⁷ that permits the passing of air into the cranial cavity. Specifically, the intracranial pressure becomes lower than atmospheric pressure, and this allows air to enter the intracranial cavity (ball-valve theory). In cases in which the pneumocephalus appears early after VP placement, this process is evident to the neurosurgeon but, when the complication develops months or years after the procedure, the cause may be not so easy to interpret.

Skull base defects may be congenital, but have most frequently been associated with meningeal-skull thinning and erosion, as a consequence of a long-lasting high intracranial pressure secondary to hydrocephalus. This condition has been most commonly observed at the middle cranial fossa, which is understandable given its relatively thin bone structure.

McCullough and Fox⁹ demonstrated excessive negative pressure in a high proportion of shunts performed. They measured the pressure at the level of the foramen of Monro in supine, sitting and standing positions, reporting mean changes ranging from −440 to −20 mmH₂O. These alterations in the pressure as a result of positional changes could be diminished using an anti-siphon device or high-pressure
shunt,\textsuperscript{3,6} avoiding the excess in negative pressure in vertical postures and increasing the safety margin. The mechanism of early or immediate pneumocephalus associated with CSF leaks is known as the “inverted bottle”; when CSF flows out, a negative pressure is created within the cranium and the space liberated fills with air.

Negative-pressure hydrocephalus could be the consequence of a pressure gradient between the cortical subarachnoid space (CSAS) compartment and atmospheric pressure because of a CFS leak that empties the cortical subarachnoid space, in turn, creating a transmantle pressure of greater than zero that leads to ventriculomegaly. Transmantine pressure is defined as the differential pressure equal to the ventricular pressure minus the CSAS pressure.\textsuperscript{10}

The diagnostic criteria for low pressure hydrocephalus (LPH) have been described by Pang and Alschuler.\textsuperscript{11} In hydrocephalic patients with progressive ventriculomegaly and neurologic deterioration after shunting, a LPH should be suspected. These authors also describe how to treat this entity; first and foremost, ventricular pressure must be substantially reduced with the objective of the decreasing the ventricular size. A less turgid and elastic brain with excessive compliance has been associated with this pathology.

The diagnosis of pneumocephalus can be made with a simple skull X-ray, but CT is the gold standard at present.\textsuperscript{12,13} Fine-cut CT scans with three-dimensional reconstruction is a valuable tool to locate skull base defects. However, the diagnostic efficacy of CT cisternography is limited in the absence of an active CSF leak that is attributable to the “ball-valve” effect: the air can get in but the contrast medium cannot get out, as occurred in our two cases.

Most authors agree that the first step in a tension pneumocephalus is the removal of intracranial air. In cases with a defect at the skull base, the treatment of choice is sealing of the defect by craniotomy or endoscopically. The presence of a porencephalic cyst has been considered useful to determine the region that requires surgical exploration when the defect is not evident.\textsuperscript{12,14} However, when the origin of the pneumocephalus is unknown, as in our first case, the approach was first to remove the intracranial air followed by temporary shunt ligation and subsequent insertion of programmable valve with anti-siphon device;\textsuperscript{15,16} to limit the CFS flow through the shunt during Valsalva’s manoeuvres or vasogenic IPC waves.

On the other hand, Filipidis et al.\textsuperscript{10} remind us that CSF leaks with pneumocephalus and low or negative pressure hydrocephalus may be refractory to standard surgical treatment. In such cases, Pang and Altschuler\textsuperscript{11} recommend treating with extended periods of external ventricular drainage; Rekate\textsuperscript{17} considers that external drainage of CSF for long periods of time with ICP maintained below the leak pressure level results in the healing of the leak. Another treatment option is neck wrapping, also proposed by Rekate,\textsuperscript{17} which serves to reverse the transmantle pressure and push more CSF through the ventricular drain, thereby decreasing ventricular size.

Conclusion

Delayed spontaneous pneumocephalus associated with VP shunt is a rare complication that can be attributed to the coexistence of a decrease in intracranial pressure generated by the shunt (siphon effect) and the presence of a cranial-dural defect. If a porencephalic cyst is observed in CT scans of patients with VP shunts and a neurologic deterioration, we should suspect the presence of and must search for a cranial-dural defect. Shunt devices with anti-siphon valves seem to be useful in the treatment of tension pneumocephalus.

LPH should be suspected in cases of CSF leaks in patients with ventriculomegaly and shunts malfunctions that do not improve with shunt with the regular pressure.

REFERENCES