Spontaneous pneumocephalus related to CSF shunting and pneumosinus dilatans in a patient with a large cavernoma

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Spontaneous pneumocephalus is a rare entity that neurosurgeons may encounter in patients with idiopathic cerebrospinal fluid (CSF) rhinorrhoea.[1] Very rarely, it can be caused by shunts, a complication that has been reported in 37 cases in the literature up to 2001.[1] The association between spontaneous pneumocephalus and pneumosinus dilatans (PSD) is extremely rare and has been reported in the literature once only in a patient with a meningioma.[2] We report a unique case of a patient who developed spontaneous pneumocephalus most likely due to the combined effect of PSD and CSF shunting. The case is also unusual in that the PSD was probably related to the raised intracranial pressure (ICP) caused by a large cavernoma.

A 28-year-old female patient was admitted as an emergency to our hospital. She gave a 2 months history of headache, vomiting, and a slowly progressing left sided weakness. Few days before admission, she became obtunded and drowsy. A brain CT scan revealed a 7 x 5 cm non-contrast enhancing space-occupying lesion in the right temporo-parietal region with obstructive hydrocephalus. In view of her relatively depressed conscious level and the presence of acute hydrocephalus, we elected in the first instance, to perform an urgent left sided ventriculo-peritoneal (VP) shunt using a medium pressure valve. Five days later, she underwent a craniotomy and total excision of the lesion, which was well capsulated, multiseptated, avascular, and containing dark brown fluid. Histologically, the lesion proved to be a large cavernoma. She made a good postoperative recovery, and the post-operative brain CT scan showed complete excision of the lesion with reduction in the size of ventricles. She remained stable at regular follow up visits to the outpatient clinic. Three years later, she was readmitted as an emergency with a 2-week history of progressive headache and rapidly becoming slow and obtunded. There was no history of trauma, convulsions, vomiting, or CSF rhinorrhea. Clinical examination revealed that she was opening her eyes spontaneously, moving her limbs without weakness, but was aphasic and unresponsive to verbal stimuli. Brain CT scan (**Figure 1**) showed intraparenchymal pneumocephalus. There was also evidence of abnormal dilatation of all the paranasal sinuses, suggestive of PSD, although the patient had no obvious facial deformity clinically. A suspicious defect in the inner wall of the left frontal sinus was also noted. Isotope cisternography was carried out to confirm the site of the fistula but proved negative. On reviewing the CT scans that were carried out during the initial presentation, there was evidence of PSD. She underwent a left frontal craniotomy. The frontal lobe was adherent to the back of the left frontal sinus and once the adhesions were released, the intraparenchymal entrapped air escaped and the tensed frontal lobe deflated. The frontal sinus was packed with muscle, bone dust, and fibrin glue, and the fistula was repaired using a fascia lata graft. The patient made a good postoperative recovery. She became alert and oriented with no neurological deficits and was discharged from the hospital a few days later. The issue of removal of shunt, insertion of an antisiphon device or replacing the valve with a high-pressure one was discussed, and she elected to wait. She remained well at one-year follow up.

Pneumosinus dilatans is a radiological diagnosis that was first reported by Meyers in 1898 and classified by Benjamins in1918.[3] It refers to abnormally enlarged, air filled paranasal sinuses without evidence of localized bone destruction, hyperostosis, or mucous membrane thickening.[2] It differs from 'sinus pneumocele' by the lack of bony wall defect and the lack of air outside the bony cavity or in the soft tissue.[3] Our patient had evidence of a small defect in the posterior wall of the frontal sinus, which may imply by definition that the intracranial air was a 'sinus pneumocele'. However, she had generalized paranasal sinuses dilatation, which was observed at the time of her initial admission, 3 years before the development the pneumocephalus. We therefore feel that PSD describes her sinuses abnormalities more accurately. Pneumosinus dilatans is either idiopathic

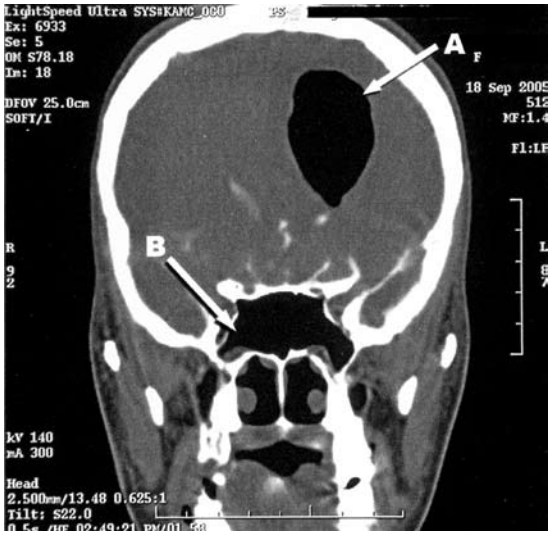

Figure 1 - Brain CT, bone windows coronal cuts with intrathecal contrast showing evidence of pneumocephalus (A) and evidence of pneumosinus dilatans in the sphenoid sinus (B).

or secondary to the presence of chronic intracranial or intra-orbital pathologies such as meningioma.[2,3] The PSD can be restricted to the sinus close to the lesion,[2] or affecting other sinuses distant from the lesion, such as in our case.[3] In addition, the association between PSD and several congenital disorders such as fibrous osseous dysplasia[3] would suggest a possible genetic predisposition for the sinus remodeling process that explains why some patients with chronic ICP elevation develop PSD and not others. It is likely that the chronic rise in ICP associated with the large cavernoma and the obstructive hydrocephalus in our case, had led to thinning of the base of the skull bones. The presumed remodeling process that followed probably resulted in the development of PSD. Pneumosinus dilatans is often found accidentally in asymptomatic individuals, but can clinically present with a variety of signs and symptoms ranging from facial swelling to headaches, vertigo, visual impairment, proptosis, and endocrine disturbance.[2,3] It has been postulated that the intracranial symptoms in patients with PSD are due to sinus pressure disequilibrium. It is likely that through a valve mechanism, air gets entrapped in the sinus leading to sinus pressure elevation, which may cause a problem when the ambient pressure is reduced.[2,3] Hence, it is accepted that the creation of a wide communication between the sinus and the nasal cavity can be curative in symptomatic patients with PSD.[3] At times, optic nerve decompression or cosmetic resection of the enlarged sinus may be necessary.[2,3] Pneumocephalus indicates a free connection between the cranial cavity and atmosphere. Air may be located in the extradural, subdural, subarachnoid, intraventricular, or as in our case intraparenchyma.[4] Pneumocephalus occurring as a complication of CSF shunts is a rare entity. It has been reported in patients aged 2 months to 77 years and diagnosed between 8 hours to 9 years after the shunt placement.[1] Such patients may present with headache, altered consciousness, motor deficits, vomiting, urinary incontinence, and hypotonia. In addition, they may complain of slashing sounds in the head.[1] Most shunted patients that develop pneumocephalus have a cranial floor defect with or without CSF leak.[5] The defect may be congenital or acquired. In our case, it is likely that the combination of the chronic raised ICP and the PSD had led to thinning of the cranial floor to a degree that the bone and dura became easily traumatized by unreported minor accidents or events such as sneezing.[4] The presence of the shunt allows the CSF to be displaced out of the cranium causing a negative ICP and allowing air to fill the vacuum. The situation was probably made worse by the potentially elevated sinus pressure due to the PSD. Intracranial air is then trapped, and it expands as a result of a ball-valve phenomenon produced by a brain plug at the level of the defect.[1,4,5] Based on the pneumocephalus etiology and air entry site location, the patients can be treated by duraplasty with or without shunt modification, shunt modification only, air aspiration, externalization, and repositioning and skin defect closure.[1] In addition to the craniotomy, some authors suggest that such patients would benefit from a high-pressure shunt valve or an antisiphon device to reduce the high negative pressure.[1] Our patient, who elected to wait before shunt removal or modification, has been informed that in the presence of a shunt and PSD, the craniotomy and repair of the fistula may have been a temporary solution as another area of thinning or potential fistula might occur.

In summary, it is hoped that this case will serve to remind neurosurgeons to look for PSD in their patients with chronic ICP elevation related to a lesion. Shunting of such patients will require careful follow up as the combined effect of shunting and PSD will make them particularly at risk of developing spontaneous pneumocephalus.

Received 27th November 2007. Accepted 25th March 2008.

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