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Radiology in Focus

Hypercellularity of the mastoid as a cause of spontaneous pneumocephalus

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Abstract
In this paper two cases are reported in which spontaneous entry of air into the head appears to have occurred through a hypercellular mastoid air cell system. In both these cases forceful sneezing and nose blowing were considered contributory factors. They underwent surgical repair of the bony defects which, combined with less vigorous nose blowing, has affected a successful repair. The aetiology of pneumocephalus is discussed and a review of the pertinent literature is also presented.

Key words: Pneumocephalus; Mastoid, Surgical Procedures, Operative

Introduction

Intracranial pneumatocele or aerocele is a rare condition in which air or gas is present within the skull. It usually results from entry of air into the skull from the atmosphere and its presence implies that there must be a breach in the skull through which air can enter. Such a defect can result from skull fracture, neoplasm, infection or surgery. Gas has also been reported in the skull as a result of intracranial infection with gas-producing organisms such as *Clostridium welchii.* Air in the cranial cavity may cause symptoms ranging from mild headache to raised intracranial pressure and may lead to death especially if there is a sudden increase in pressure after sneezing, for example. The entrance of air may be associated with the escape of cerebrospinal fluid, and with the entry of organisms leading to life-threatening intracranial infection. In this paper the authors report two cases in which spontaneous entry of air into the head appears to have occurred through a hypercellular mastoid air cell system.

Case reports

Case 1

A 17-year-old man presented to the Accident and Emergency Department with a five-week history of a soft painless swelling above and behind the right ear. Attempted aspiration was unsuccessful. Two weeks later he developed right-sided otalgia and blood-stained discharge and was seen in the Department of Otolaryngology. He reported that the swelling behind the ear increased in size when he blew his nose. He complained of feeling generally unwell but had no more specific symptoms. In particular there was no complaint of headache or of visual disturbance, and no CSF otorrhoea or rhinorrhoea. There was no previous history of significance and in particular no history of head injury. On examination there was a 5 cm soft painless swelling behind the right ear. There was mild otitis externa on both sides, and the tympanic membranes were intact. Neuro-otological examination, and audiometry were both normal.

A computed tomography (CT) scan showed that the swelling was due to a collection of subcutaneous air that was continuous with a hyperpneumatized mastoid air cell system through a defect in the cortex. In addition there was a large defect in the bone of Trautmann's triangle, extending from the sigmoid sinus to the otic capsule and there was an extradural collection of air present in the posterior cranial fossa (Figures 1 and 2).

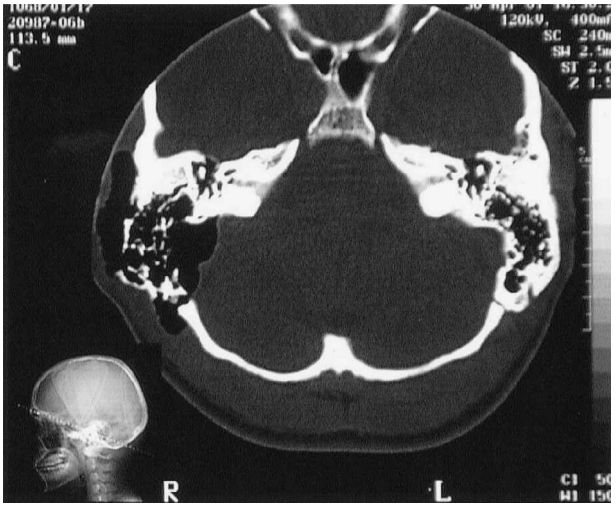

Fig. 1

Case 1 showing a hyperpneumatized mastoid air cell system with a large defect in Trautmann's triangle and overlying subcutaneous air.

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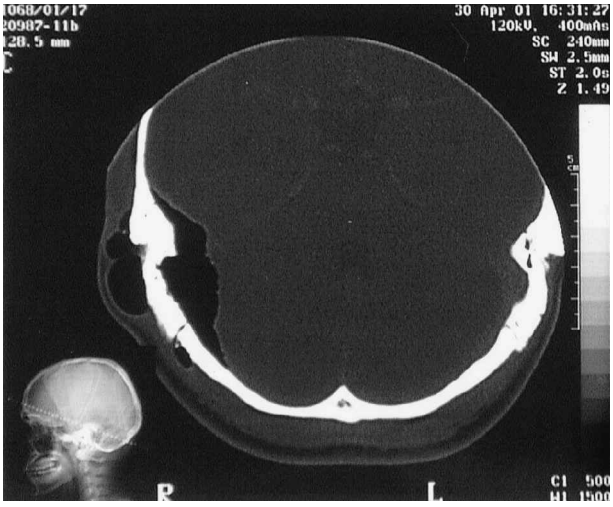

FIG. 2

Case 1 showing subcutaneous and subdural air collections.

He was advised to desist from nose blowing, and the swelling decreased in size. He was admitted for repair, and at surgery the cortical defect was immediately identified. A cortical mastoidectomy was performed and the large bony defect of the posterior wall of the petrous bone was identified. It extended from the otic capsule to the retrosigmoid air cell system and from the sinodural angle towards the jugular bulb. The dura was intact, there was no CSF fistula confirming that the aerocele was entirely extradural. The bony defect was repaired using a paté of autogenous bone dust and fibrin glue, and the cortical mastoidectomy was lined with fascia and obliterated with abdominal fat (Figure 3).

The post-operative course was uncomplicated and there has been no recurrence of the aerocele after six-months' follow-up.

Case 2

This 50-year-old female presented in February 2000 with a two-month history of burning pain and paraesthesia over the left cheek and lower jaw. Her symptoms were markedly worse if she coughed or sneezed. She also complained of pain in the vertex on waking in the morning. There was no history of ear disease nor of head injury. There was no CSF otorrhoea or rhinorrhoea. She did, however, have a history of allergic rhinitis and nasal polyps, and blew her nose frequently and with vigour. Treatment of her facial pain with carbamazepine, amitryptilline, and codeine was ineffective. On examination the tympanic membranes were normal. There was depression of sensation in the second and third divisions of the left trigeminal nerve. The other cranial nerves were normal, as were the optic fundi. There were no long tract or cerebellar signs. She had almost complete nasal obstruction from polyps. A pure tone audiogram revealed a mild bilateral high frequency hearing loss.

A CT scan showed the presence of intracranial extradural air in the region of the left petrous apex (Figure 4). Both petrous bones were highly pneumatized with air cells extending into the clivus and into the occipital bones. These appearances were more marked on the left side. In addition there was a small bony defect in the inner cortex of the left petrous bone at the junction of the transverse and sigmoid sinuses.

Surgical exploration and repair was recommended but before she could be admitted she suffered a pulmonary embolus, which took some months to treat. She was advised to stop blowing her nose or sneezing if possible. When she was eventually seen for review prior to the postponed surgery, she stated that her symptoms had gone completely. A further CT scan was therefore performed and this failed to reveal the presence of air in the head. Nevertheless it was felt advisable to explore the ear in order to prevent recurrence. At surgery the lateral venous sinus was found to be dehiscent over an area of 5×5 mm adjacent to the posterior cranial fossa dura. The exposed surface of the sinus was granular and there was evidence of old haemorrhage in the surrounding air cells. The dura itself was intact. Behind the lateral sinus the air cell system was noted to extend widely into the occipital bone. This air cell system was obliterated using a paté made of bone dust and fibrin glue and the mastoid was filled with abdominal fat. Initially she remained well but at two weeks after the surgery she returned with a subcutaneous collection of air

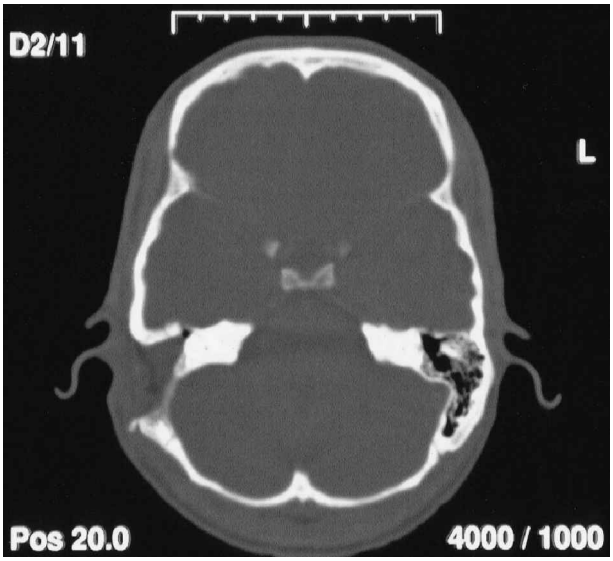

FIG. 3

Post-operative view of *Case 1* following repair of the bony defect.

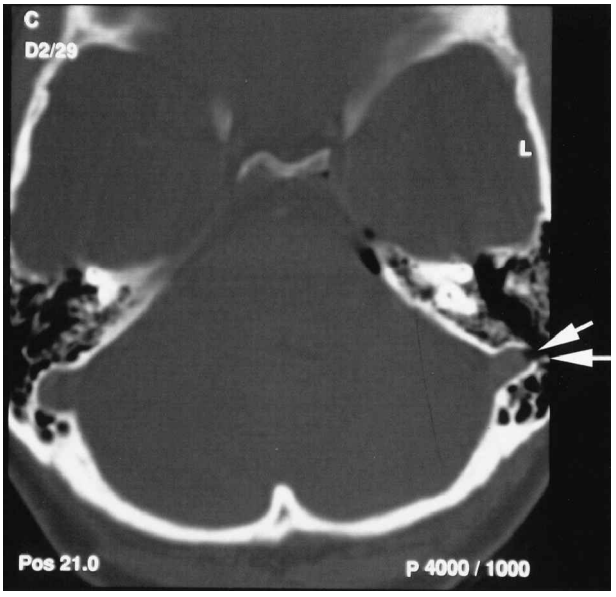

FIG. 4

Case 2 showing highly pneumatized petrous bones with a bony defect at the junction of the transverse and sigmoid sinuses.

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behind and above the left ear and extending into the cheek. Although she said she had not been blowing her nose, it was felt appropriate to deal with the nasal polyps rather than re-explore the ear. This proved successful and she has had no further problems for six months at the time of writing.

- **Two cases of spontaneous pneumocephalus which appear to have occurred through a hypercellular mastoid air cell system**
- **Both underwent surgical repair of bony defects and were advised to blow their noses less vigorously**

Discussion

The presence of air in the head nearly always indicates the presence of a defect in the skull though which air enters from the atmosphere. Occasionally gas-forming organisms, introduced at the time of a penetrating injury, such as *Clostridium welchii* may be responsible but this is rare.[1] Injury or disease in the paranasal sinuses or middle-ear cleft may be responsible for causing the breach in the skull. Skull base fracture and surgical trauma are probably the commonest aetiological factors, but skull base tumours and cholesteatoma may also be responsible. Spontaneous pneumocephalus is rare, amounting to 0.6 per cent of the series of 295 cases described by Markham.[2] According to Jelsma and Moore[3] the first description of pneumocephalus was by Lecat in 1741, although Chiari in 1884[4] described it at post mortem and Luckett in 1913[5] was the first to demonstrate intracranial air radiologically. Dandy[6] originally classified pneumocephalus as subarachnoid, subdural, intracerebral or intraventricular. Jelsma and Moore[3] recognized that extradural air collections might also occur. When there is an associated meningeal defect CSF leakage may occur. If there is a compensatory flow of CSF out of the cranium as air enters, the intracranial pressure may not rise. In some circumstances, however, there may be a valve-like effect that allows air in but prevents air or CSF from escaping.[7] This can result in a sudden and potentially lethal increase in intracranial pressure in the event of sneezing or coughing.

In the cases described here there was no clearly defined aetiological factor. Both individuals had hyperpneumatized temporal bones, however, and it is possible that there were congenital dehiscences of the posterior fossa dura, and in *Case 1* dehiscence of the mastoid cortex. Forceful sneezing and nose blowing were contributory factors in both cases, and it seems possible that this could have caused the dura to strip off the petrous bone, causing an extradural collection of air. In neither case was there a CSF fistula, a fact that lends support to the clinical and radiological impression that the meninges had not been breached and that the pneumatoceles were extradural. Defects in the temporal bone are of course common with an incidence of as high as 47 per cent.[8] There are few other cases of spontaneous pneumocephalus associated with hypercellularity of the temporal bone,[9–14] and it is perhaps remarkable that condition is not more common.

As far as the authors can determine there are no recorded cases where the responsible bone defect has been in the posterior fossa as opposed to the middle fossa. The role of nose blowing, coughing, or similar acute pressure alterations is important. Ahren and Thulin[8] recorded the case of a man who developed lethal intracranial complications following paracentesis and inflation with a Politzer balloon in the external meatus. The cause of death was acute extradural aerocele followed by extradural haematoma formation, and at postmortem bony defects were found in the tegmen. In the case described by Stavas *et al.*[14] coughing exacerbated the symptoms. Nyrop *et al.*[13] described the case of a man with hyperpneumatization of the skull base who habitually performed Valsalva's manoeuvre, and postulate that the pneumatization was the result of this habit. They were able to demonstrate a reduction in the pneumatization of the skull base, with re-ossification of the occipital and temporal bones some seven months after persuading their patient to desist from this practice. Interestingly their patient's symptoms were influenced by altitude, either flying or skiing. Both of our patients reported a worsening in their symptoms on nose blowing and were advised to desist from this practice. In *Case 2* this lead to the resorption of air and resolution of symptoms.

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