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Idiopathic CSF rhinorrhoea presenting with tension pneumocephalus and hemiparesis

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Abstract
A case of non-traumatic/non-iatrogenic CSF rhinorrhoea, presenting with tension pneumocephalus and hemiparesis is described. The possible pathological processes involved in this rare case are discussed. Cases in the literature of idiopathic CSF rhinorrhoea and also those of spontaneous pneumocephalus are reviewed.

Key words: Cerebrospinal fluid rhinorrhoea; Pneumocephalus

Introduction

Non-traumatic/non-iatrogenic CSF rhinorrhoea is occasionally seen in clinical practice. For the majority of these a causal pathology is identifiable. There are no previous reports of idiopathic CSF rhinorrhoea presenting with acute hemiparesis secondary to tension pneumocephalus. The possible pathological processes involved in the genesis of this fascinating condition are examined.

Case report

A previously fit 67-year-old right-handed Afro-Caribbean female presented to Queen Elizabeth Medical Centre, Department of Neurosurgery with a two-week history of proven CSF rhinorrhoea, progressive left hemiparesis and headache. There was no history of trauma, nasal symptoms, meningitis, endocrine or visual disturbance.

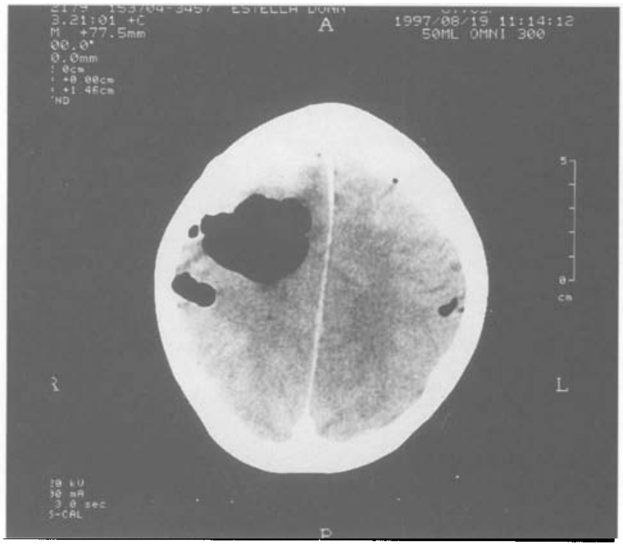

Fig. 1

Axial CT of the brain at presentation. Pneumocephalus is shown.

Neurological examination revealed minimal left upper motor neurone facial nerve paresis, grade 0 power in the left upper limb and grade 3 in the left lower limb. Otherwise, full neurological and full general physical examination was unremarkable. Cranial computed tomography (CT) and magnetic resonance imaging (MRI) revealed intracranial air whose distribution is seen in Figures 1 and 2, and also a liquid level in the right sphenoid sinus (Figure 3). There was no other intracranial abnormality. Emergency burr hole and aspiration of high-pressure air was undertaken. This produced a transient improvement in leg power and CT appearances but 48 hours post-operatively, the air collection had reformed and the limb

Fig. 2

Axial CT of the brain at presentation. Pneumocephalus is seen with air occupying a variety of sites. Hollow arrow indicates part of the large air pocket which is clearly seen in the Sylvian fissure in Figure 1. The long thin arrow indicates air outlining the 'T'-shape of the left Sylvian fissure. The long thick arrow indicates air in the fourth ventricle. The arrowhead indicates the lentiform pocket of air in the frontal region.

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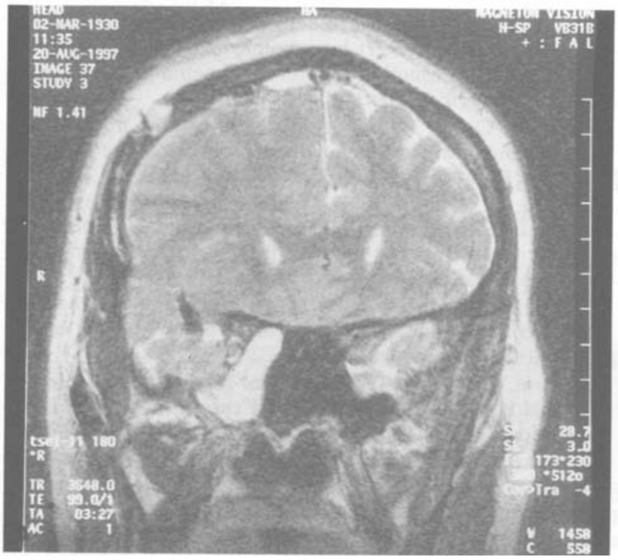

FIG. 3

Coronal MRI (T2 weighted) showing fluid in right sphenoid sinus.

deficit had reverted. Further burr-hole aspiration (using the same burr-hole) failed to improve symptoms so definitive repair was carried out. Exploration of the sphenoid was undertaken via the external ethmoidal approach by the senior ENT author. This exploration, using microscopy and endoscopy revealed a defect in the lateral wall of the right sphenoid sinus with obvious flow of CSF. The defect was closed by fascia lata, fat and tisseel.™ A second burr hole was drilled during the same anaesthetic and further high pressure air was released. The histological report of a mucosal biopsy taken from the site of CSF emergence showed no evidence of a malignant lesion. Chest X-ray, mammogram and abdominal ultrasound scan were normal and hence a skull-base metastasis was excluded. Post-operative CT scanning showed further resolution of the pneumocephalus (Plate 4). The operative closure of the CSF fistula resulted in total cessation of the

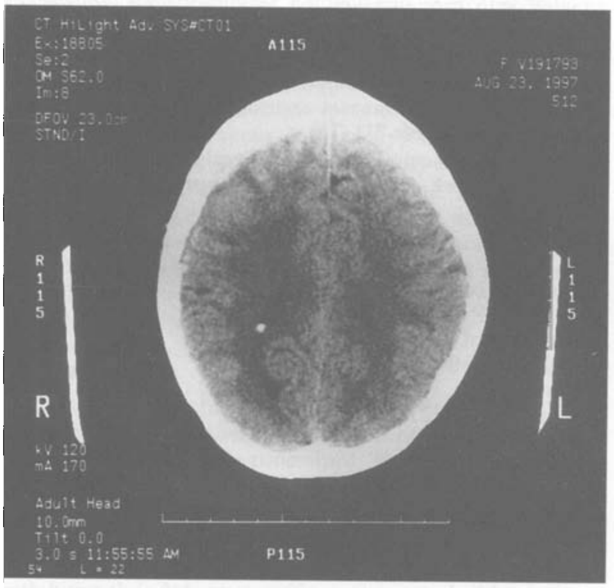

FIG. 4

Axial CT showing resolution of pneumocephalus following surgery.

leak and a gradual improvement in the patient's clinical condition. Six months after the definitive operation, the patient was able to walk and had grade 4 arm power.

Discussion

This case is an intriguing variation on the fascinating theme of idiopathic CSF fistula. Three key questions are addressed.

How did the CSF fistulas arise?

The aetiology of non-traumatic CSF rhinorrhoea has been previously classified (Ommaya *et al.*, 1968). In practice the majority of cases do have an identifiable cause e.g. tumour, congenital abnormality (such as encephalocoele), chronic hydrocephalus or focal infection. O'Connell first described the category of focal atrophy (O'Connell, 1964). He hypothesized that idiopathic atrophy of the olfactory bulb exposes the cribriform plate which is an area of inherent anatomical weakness. No author since has contested this hypothesis although the diagnosis is probably one of exclusion, rather than a discrete pathological entity.

Middle fossa sites of spontaneous leak mostly involve the sphenoid (McAllister and Parameswaran, 1981; Coiteiro *et al.*, 1995) and anterolateral floor of the middle fossa. The latter can be pneumatized in up to 17 per cent of cases, and may be continuous with the sphenoid sinus as a so-called lateral extension (Morley and Wotzmann, 1965). This region is important both after major trauma and as a site for congenital defects (Raaf, 1965). Posterior fossa defects can also arise (Ahren and Thulin, 1965), possibly congenitally and present as CSF rhinorrhoea via the Eustachian tube. Although essentially idiopathic, there has been no lack of speculation regarding the natural history of CSF fistulation at all of these sites. Prominent amongst these is that it is a combination of a skull base defect (either dehiscence (Ohnishi, 1981), congenital thinness, or presence of anatomical pit holes) and dural attrition. CSF pulsation in an arachnoid pouch (Ommaya, 1976) or basilar artery pulsation (Coiteiro *et al.*, 1995) may cause dural erosion in the same manner that arachnoid granulations cause depressions in the interior of the vault. The occurrence of arachnoid pouches has been confirmed using necropsy studies of normal pituitary anatomy (McLachlan and Williams, 1968). A theory of 'acquired meningocoeles or meningoencephalocoeles', has been proposed as an intermediate step to fistulation (Kauffman *et al.*, 1977). Maximal CSF pulsation pressures occur only in adults which might explain why the pathology is rarely seen below the age of 16 (Hubbard *et al.*, 1985). Such an ill-defined congenital or acquired defect probably exists in the case described here. Fortunately the focal abnormalities in the region of right sphenoid sinus obviated the need for further CSF fistula detection imaging.

By what mechanism did the air gain access to the interior of the skull?

In the absence of gas-forming microorganisms (Randall *et al.*, 1993), the acquisition of free intracranial air requires the direct communication of pneumatized bone or ambient air with the intracranial contents. Subdural air must additionally have traversed a dural defect. Spontaneous causes include numerous lesions of the paranasal sinuses (mucocoele (Farooki *et al.*, 1976), osteoma (Hardwidge and Varma, 1985), invasive ossifying fibroma (Tobey *et al.*, 1996), epidermoid (Kinsley and Dougherty, 1993), and carcinoma (Takahashi and Kanazawa, 1992)), otological lesions (Andrews and Canalis, 1986) and intradiploic

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epidermoid (Jakubowski *et al.*, 1997). The egress of CSF and ingress of air may simultaneously occur through a composite bone/dural defect by means of the 'inverted bottle effect' (Jooma and Grant, 1983), in which sudden displacements of a large volume of fluid is simultaneously replaced by an equivalent volume of air. Initiation of an active fistula usually requires a sudden marked change in pressure gradient across a previously occult defect. The tension of intracranial air will remain high if an anatomical ball-valve mechanism (Markham, 1967) is in operation and the magnitude of the tension must relate, at least initially, to the entry pressure. Sudden falls of intracranial pressure can occur during the medical intervention of either lumbar or ventricular CSF drainage. This may induce pneumocephalus *de novo* when an occult defect exists, or in cases of pre-existing CSF rhinorrhoea where the drainage is intended as therapy, air may also enter via the same fistula (Hubbard *et al.*, 1985). Conversely, a leak may be initiated by a sneeze (Chiari, 1884) or the use of therapeutic continuous positive airway pressure (CPAP) (Young and Nevin, 1994) (both sudden elevations of extracranial pressure locally).

Our patient undoubtedly experienced an episode of the inverted bottle effect which was probably associated with a forgotten sneeze.

What is the explanation for the observed distribution of intracranial air seen in this patient?

The remarkable feature of this case is the fact that spontaneous CSF rhinorrhoea should present with a focal neurological deficit. This was undoubtedly caused by the large pocket of air in the right frontoparietal region (Figures 1 and 2). Such a collection is probably in the right Sylvian fissure subarachnoid space but it is very unusual for air to split the adhesions of the Sylvian fissure. More commonly air is seen aggregating in smaller pockets as are seen outlining the 'T' shape of the left Sylvian fissure on axial imaging (Figure 2). Both the lentiform collection of air in the frontal region and the air in the fourth ventricle are more common findings in cases of pneumocephalus both spontaneous and post-traumatic (Figure 2).

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