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Radiology in Focus

Idiopathic diffuse erosion of the skull base presenting as cerebrospinal fluid rhinorrhoea

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Abstract
We present an unusual case of generalized erosion of the skull base. We have not found a similar case reported in the world literature. The presenting symptom was spontaneous cerebrospinal fluid (CSF) rhinorrhoea which arose from a bony defect associated with herniation of the right temporal lobe into the sphenoid sinus. We discuss the management of such a case including imaging of the skull base and the endoscopic repair of the bony defect.

Key words: Skull; Encephalocoele; Paranasal sinuses; Cerebrospinal rhinorrhoea

Introduction

Most cases of primary spontaneous CSF rhinorrhoea are associated with a localized defect in the skull base. The majority can be defined by high resolution cranial computerized tomography (CT) scans (Hughes *et al.*, 1997) although a minority are detected by a variety of means including CT with contrast, magnetic resonance imaging (MRI), and fluorescein lumbar puncture. Much more unusual is the case of generalized thinning of the skull base leading not only to CSF rhinorrhoea but also herniation of the brain substance through several defects.

Case report

A 71-year-old male was referred to the ENT department with a five-week history of intermittent right-sided clear rhinorrhoea. The initial episode had occurred spontaneously whilst stooping forwards. He was otherwise fit and well. There was no past history of head injury or cranial surgery. On examination clear right-sided rhinorrhoea was present.

The nasal discharge was confirmed to be cerebrospinal fluid by immunofixation of ß-transferrin. A CT scan showed widespread patchy thinning of the skull base floor (Figure 1) together with herniation of the right temporal lobe into the sphenoid sinus. An MR scan of the petrous bone showed the defect in the basal part of the right sphenoid sinus (Figure 2) and confirmed several areas of skull base thinning bilaterally. The full blood count, urea and electrolytes were all within the normal range. The C-reactive protein was 19 (normal at 71 years). The autoimmune screen identified a strongly positive thyroid cytoplasm antibody, however, other autoantibodies were negative. The CSF opening pressure was found to be normal at 180 mm (reference range <210 mm CSF). He had no evidence of hyperparathyroidism or bony erosion elsewhere.

The patient underwent transnasal endoscopic surgery. The antero-inferior defect in the right sphenoid sinus was identified (Figure 3) and closed with a fascia lata graft supported by fat. The surrounding mucosa was noted to be

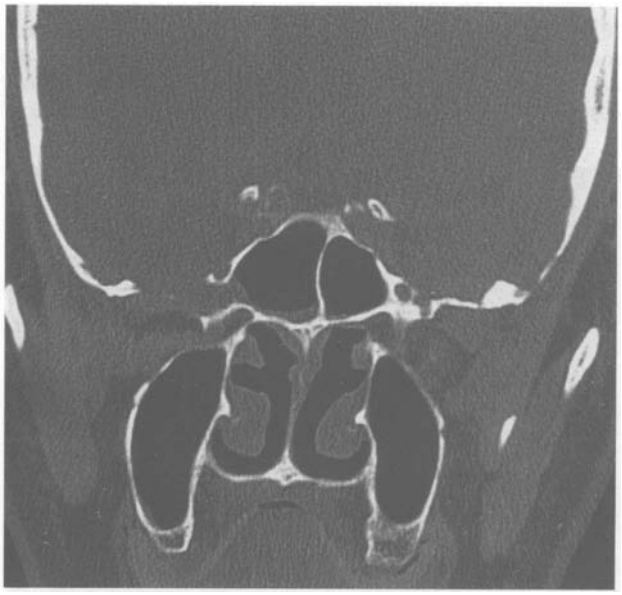

Fig. 1
Coronal CT scan showing widespread bilateral erosion of the skull base with soft tissue entering the sphenoid sinus inferolaterally.

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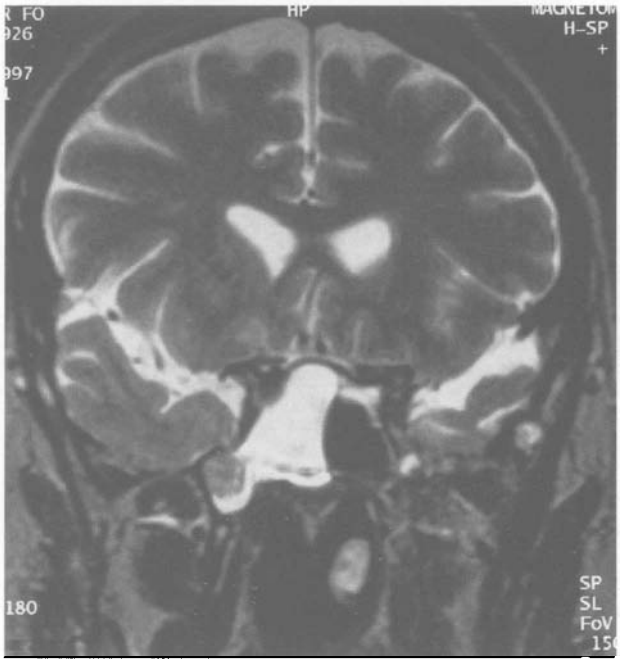

FIG. 2

Coronal T2 weighted MRI scan showing herniation of the temporal lobe as well as CSF in the sphenoid sinus.

healthy and no biopsy was taken because of the proximity of the carotid artery and optic nerve. As the CT scan showed bilateral thinning and erosion on the left side, this was also prophylactically grafted to reduce the potential of a further CSF leak developing. The patient remains well and there has been no recurrence of the CSF leak four months post-operatively.

Discussion

Primary CSF rhinorrhoea may be the first presentation of a previously occult malformation of the skull base. Such malformations can also present with meningitis (Pappas-Jr *et al.*, 1993; Schick *et al.*, 1997). The central skull base ossifies from cartilage with at least 25 separate ossification centres involved in this complex process (Madeline and Elster, 1995). Congenital abnormalities arise either from incomplete fusion or premature fusion of the normal bony elements of the skull base. With incomplete fusion a herniation of cranial contents (cephalocoele) can arise. In the case described a meningocephalocoele (involving herniated temporal lobe) was present. Neoplastic causes of CSF rhinorrhoea can occur secondary to lesions invading the nasopharynx and paranasal sinuses such as metastatic tumours from lung, breast or prostate; and much less commonly intrinsic lesions (eg pituitary tumours, gliomas, neuromas, meningiomas, craniopharyngiomas and rhabdomyosarcomas) (Laine *et al.*, 1990; Abrahams and Eklund, 1995). Other conditions that can give rise to CSF rhinorrhoea include suppurative sinus disease and inflammatory conditions (McCoy, 1963).

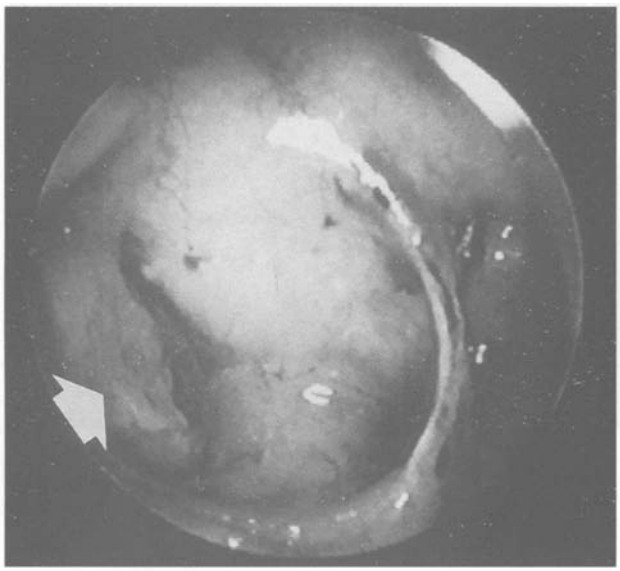

FIG. 3

Endoscopic view of the lateral defect in the right sphenoid sinus.

Radiological investigation has a key role in identifying the site of the skull base defect as well as providing information as to the underlying cause. CT scanning gives good bone detail whilst MR imaging techniques provide information about the soft tissues (Hamlin and Hasso, 1994; Lustrin *et al.*, 1994). Thus both forms of investigation taken together provide complementary information to help in diagnosis and surgery.

In the case presented there was generalized thinning of the skull base on CT. Massive osteolysis of the skull base has been reported in the literature (Frankel *et al.*, 1997). More localized defects have also been reported (Tolley *et al.*, 1991; Schick *et al.*, 1997).

Surgery to stop CSF rhinorrhoea is primarily performed to prevent meningitis which in follow-up studies has been shown to be a not uncommon as well as serious complication (MacGee *et al.*, 1970; Klastersky *et al.*, 1976). The surgical management of CSF rhinorrhoea has traditionally been by a craniotomy approach as described by Dohlman (Dohlman, 1948). More recently, transnasal endoscopic surgical techniques have been used to close CSF leaks. The first description of an endoscopic closure was by Wigand (Wigand, 1981). In North America Dodson *et al.*, (1994) reviewed 29 patients who underwent endoscopic repair of CSF rhinorrhoea and more recently Lanza *et al.* (1996) presented 36 patients who underwent surgical repair by endoscopic surgery. In the UK Hughes *et al.* (1997) described 17 cases of endoscopic closure for CSF rhinorrhoea. In this case, the diagnosis of CSF rhinorrhoea was confirmed both by laboratory testing (immunofixation of ß-transferrin) and radiological investigation. Whilst the aetiology for the extensive erosion of the skull base in this gentleman is unknown, his CSF leak has been dealt with endoscopically with minimum morbidity.

References

Abrahams, J. J., Eklund, J. A. (1995) Diagnostic radiology of the cranial base. *Clinics in Plastic Surgery* **22 (3):** 373–405.

Dodson, E. E., Gross, C. W., Swerdloff, J. L., Gustafson, L. M. (1994) Transnasal endoscopic repair of cerebrospinal fluid rhinorrhea and skull base defects: a review of twenty-nine cases. *Otolaryngology – Head and Neck Surgery* **111:** 600–605.

Dohlman, G. (1948) Spontaneous cerebrospinal rhinorrhoea. *Acta Otolaryngologica* **67 (Supp):** 20–23.

Frankel, D. G., Lewin, J. S., Cohen, B. (1997) Massive osteolysis of the skull base. *Pediatric Radiology* **27:** 265–267.

Hamlin, J. A., Hasso, A. N. (1994) Magnetic resonance imaging of the skull base. *Topics in Magnetic Resonance Imaging* **6(3):** 183–201.

Hughes, R. G. M., Jones, N. S., Robertson, I. J. A. (1997) The endoscopic treatment of cerebrospinal fluid rhinorrhoea: the Nottingham experience. *Journal of Laryngology and Otology* **111:** 125–128.

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Klastersky, J., Sadeghi, M., Brihaye, J. (1976) Antimicrobial prophylaxis in patients with rhinorrhea or otorrhea: a double-blind study. *Surgical Neurology* **6:** 111–114.

Laine, F. J., Nadel, L., Braun, I. F. (1990) CT and MR imaging of the central skull base. *Radiographics* **10(5):** 797–821.

Lanza, D. C., O'Brien, D. A., Kennedy, D. W. (1996) Endoscopic repair of cerebrospinal fistulae and encephaloceles. *Laryngoscope* **106:** 1119–1125.

Lustrin, E. S., Robertson, R. L., Tilak, S. (1994) Normal anatomy of the skull base. *Neuroimaging Clinics of North America* **4 (3):** 465–478.

MacGee, E. E., Cauthen, J. C., Brackett, C. E. (1970) Meningitis following acute traumatic cerebrospinal fluid fistula. *Journal of Neurosurgery* **33:** 312–316.

Madeline, L. E., Elster, A. D. (1995) Postnatal development of the central skull base: Normal variants. *Radiology* **196(3):** 757–763.

McCoy, G. (1963) Cerebrospinal rhinorrhoea: a comprehensive review and a definition of the responsibility of the rhinologist in diagnosis and treatment. *Laryngoscope* **73:** 1125–1157.

Pappas, D. G. Jr., Hammerschlag, P. E., Hammerschlag, M. (1993) Cerebrospinal fluid rhinorrhea and recurrent meningitis. *Clinical Infectious Diseases* **17:** 364–368.

Schick, B., Draf, W., Kahle, G., Weber, R., Wallenfang, T. (1997) Occult malformations of the skull base. *Archives of Otolaryngology–Head and Neck Surgery* **123(1):** 77–80.

Tolley, N. S., Lloyd, G. A. S., Williams, H. O. L. (1991) Radiological study of primary spontaneous CSF rhinorrhoea. *Journal of Laryngology and Otology* **105:** 274–277.

Wigand, M. E. (1981) Transnasal ethmoidectomy under endoscopical control. *Rhinology* **19:** 7–15.

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