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Radiology in Focus

Chronic cerebrospinal fluid leak into skull base causing intramedullary osteolysis

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Abstract
We present the rare case of a 74-year-old woman with extensive intramedullary osteolysis of the petroclival skull base straddling both sides of the basi-occiput and basi-sphenoid. She presented with clinical features of recurrent spontaneous cerebrospinal fluid (CSF) otorrhoea despite three previous attempts at repair of the CSF leak. Recent advances in imaging techniques enable accurate radiological diagnosis of skull base lesions. We performed T1- and T2-weighted magnetic resonance (MR) images, a fluid attenuated inversion recovery (FLAIR) sequence MR, and a diffusion scan to study the characteristics of the skull base pathology. This revealed extensive osteolysis, with cystic spaces within the clivus and the petrous pyramid extending also to the basi-occiput. The leak was sealed using the technique of subtotal petrosectomy with obliteration of the eustachian tube and blind pit closure of the ear canal. The patient has been followed up for six months with no recurrence of symptoms.

Key words: Cerebrospinal Fluid Otorrhoea; Osteolysis; X-ray, Diagnostic

Introduction

Cerebrospinal fluid (CSF) leaks secondary to trauma or surgery are usually clinically obvious and present few diagnostic problems. However, spontaneous CSF leaks may be overlooked due to their subtle and intermittent presentations.[1] Spontaneous CSF otorrhoea may occur along a preformed bony pathway around or through the labyrinth, which can present early in life. Alternatively, it can occur later in life (over the age of 50 years) as the congenital structures carrying CSF i.e. arachnoid villi and granulations enlarge with increasing age and physical activity with intermittent increases in subarachnoid pressure.[2] The bone erosion that occurs with this pulsatile pressure may occur close to a pneumatized bone such as the paranasal sinuses and the middle-ear cleft. This rupture could then lead to CSF rhinorrhoea or otorrhoea.[2]

Cystic lesions affecting the skull base include congenital epidermoid cysts, arachnoid cysts and cholesterol granulomas.[3–5] These lesions can produce CSF otorrhoea or rhinorrhoea either with bone erosion or after trivial trauma.

The object of the present case report is to highlight the dilemma that the neurotology and neuroradiology teams faced in the diagnosis and management of this rare case of extensive intramedullary osteolysis of the petroclival regions filled with CSF. The report also presents a rare complication of chronic CSF leakage into the skull base i.e., osteolysis. Also highlighted are the improved techniques of imaging that enable conclusive diagnosis of intracranial and skull base lesions.

Case report

A 74-year-old lady presented with recurrent spontaneous left-sided CSF otorrhoea. Past surgical history included attempted repair of defects in the left middle fossa via a craniotomy 14 years ago and a transmastoid approach 12 years ago.

High resolution computerized tomographic (CT) imaging of the head revealed an extensive lesion of the skull base occupying the petroclival regions on both sides (Figure 1). The symmetrical areas of abnormality about the basi-occiput and basi-sphenoid were hypodense on CT and the appearances were those of patchy bony destruction sparing the cortex. No obvious abnormalities in the roof of the petrous bones were noted. A T1-weighted MR revealed the lesion to be of a low intensity with no contrast enhancement (Figure 2). T2-weighted images showed a hyperintense signal from the lesion in the petroclivus (Figures 3 and 4). The lesion presented a low signal on a FLAIR sequence MR scan (Figure 5) and a large area of low signal intensity interspersed with patchy high signals (bone marrow remnants) on diffusion scanning (Figure 6) confirming the radiological diagnosis of intramedullary cystic spaces filled with CSF.

With the above radiological features in mind, a left subtotal petrosectomy and obliteration of the petrous cavity with abdominal fat and muscle was undertaken with blind sac closure of the external ear canal and obliteration of the eustachian tube. An attempt at biopsy of the lesion via a sphenoidotomy for a histopathological diagnosis

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FIG. 1

Axial view CT scan showing extensive, multiple lytic lesions within bones of the petroclival regions (black arrowheads).

FIG. 3

T2-weighted sagittal view MR scan with a hyperintense signal from the lesion in the petroclival lesion (black arrowhead).

resulted in a small CSF leak, that was immediately repaired with fat and muscle. It has been six months since surgery and the patient remains asymptomatic.

Discussion

There are many cases reported in the literature that allude to chronic spontaneous CSF leak secondary to arachnoid granulations and arachnoid cysts.[1,2,4] Intracranial cysts show certain characteristic radiological features depending on their contents that could range from CSF-containing arachnoid cysts, fat-containing lipomas, cholesterol granulomas and epidermoid cysts.[4,5] Epidermoid cysts are hypodense on CT, hypointense on T1 MR and hyperintense on T2 MR scans.[3,5] However, on FLAIR sequence, epidermoid cysts present a high signal that remains bright on diffusion scan.[6–9] In cases of fluid containing cystic spaces, such as arachnoid cysts and CSF filled osteolytic lesions, the signal intensity is low both on FLAIR and diffusion MR.[6–9]

The dilemma in this case was whether or not this represented just CSF in cystic spaces or CSF with some underlying pathology. With the imaging characteristics described above and in the illustrations, CSF alone was suspected. However, in order to definitively exclude an

FIG. 2

T1-weighted sagittal view MR scan with gadolinium contrast showing a low intensity space-occupying lesion in the clivus with no enhancement (black arrow); an incidental enhancing lesion is seen in the floor of the fourth ventricle (possibly a choroid plexus papilloma; black arrowhead).

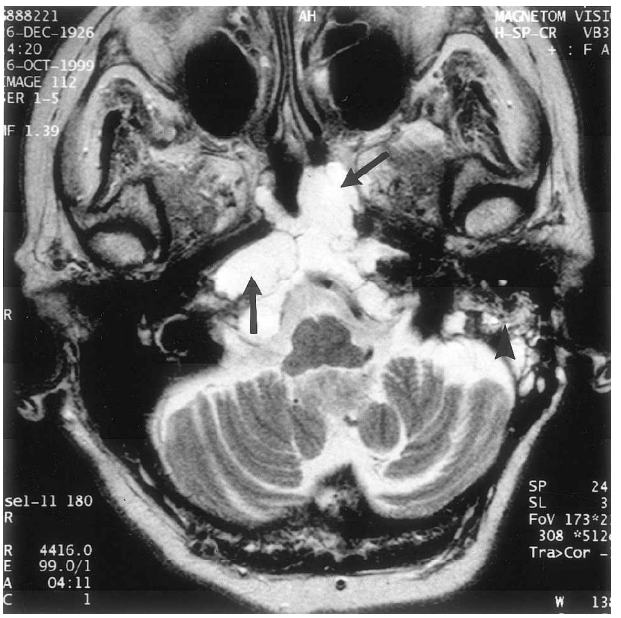

FIG. 4

T2-weighted axial MR scan depicting an extensive high intensity abnormality involving the basi-occiput and the clivus (black arrows); incidental inflammatory changes in the left mastoid can be seen (black arrowhead).

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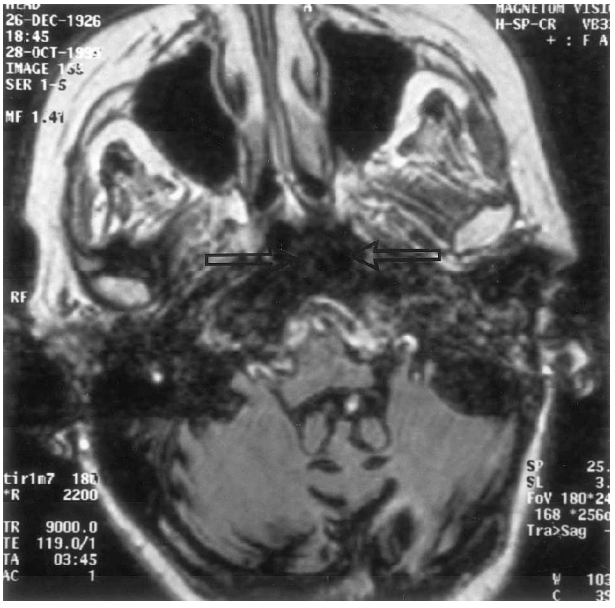

FIG. 5

A FLAIR sequence axial MR scan shows a low signal with the petroclival lesion confirming the pressure of fluid (outlined arrows).

FIG. 6

An axial view diffusion scan shows low signal in the petroclival areas (outlined arrows) with very little high signal from marrow remnants (black arrowhead).

underlying epidermoid, a sphenoidotomy was carried out with opening of the basi-sphenoid. This confirmed that the pathology was a spontaneous CSF leak with no underlying lesion. The opening in the basi-sphenoid was immediately sealed with soft tissue and fibrin glue. The follow-up plan for this patient includes MR imaging once a year to study the characteristics of the petroclival lesion.

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