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Temporal Lobe Encephalocoele Presenting with Seizures and Hearing Loss

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ABSTRACT

A case of a bilateral temporal lobe encephalocoele that presented as seizures and hearing loss for many years. Diagnosis was confirmed on CT and MR imaging, which showed deficiencies in the temporal bone. The patient subsequently underwent surgical repair and recovered from his presenting symptoms.

Keywords: brain hernia, encephalocoele, meningoencephalocoele, seizures, temporal lobe

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INTRODUCTION

Idiopathic brain herniation into the middle ear is a rare condition that can be a diagnostic challenge. Presenting symptoms and signs vary, and diagnosis is made with a high index of suspicion, with confirmation by both computed tomography (CT) and magnetic resonance (MR) imaging. We report a case of a 46-year-old man with bilateral temporal encephalocoele, who presented with a long-standing history of temporal lobe seizures and hearing loss. The encephalocoeles were missed in the initial radiologic studies and were found retrospectively after several years.

CASE REPORT

A 46-year-old Chinese man presented to the ENT department with a two weeks' history of right auricular pain and intermittent clear rhinorrhoea. The medical history was a long-standing bilateral hearing loss on hearing aid as well as temporal lobe seizures, which were well controlled with medication since 1985. Initial workup for his seizures, which included both CT and MR imaging of the brain, was reported to be normal many years ago. The patient did not have any past history of meningitis, head trauma or ear operation.

Otoscopy showed a right retracted tympanic membrane with a posterior mass in the attic region and pulsating clear fluid behind the left membrane. A preoperative audiogram indicated bilateral conducting hearing loss. Air conductions were 30 dB on the right and 45 dB on the left. Bone conductions were 10 dB on the right and 20 dB on the left. Tympanogram was Type B for both ears. The rest of the ENT examinations were normal. CT of the petrous temporal bone showed bony defects in both mastoid tegmens with associated soft tissue masses of the middle ear (Fig. 1). A review of the previous MR images taken in 1999 showed portions of the inferior aspect of both temporal lobes herniated into the middle ear cavity on coronal sequences (Fig. 2).

The patient subsequently underwent a repair of the left temporal encephalocoele via a combined transmastoid and middle cranial fossa approach. A repair on the right side was done several months later via the same approach.

At surgery, herniated brain tissues were found in both mastoid cavities and were amputated. A large 2 cm defect was found in the left mastoid tegmen. Homograft bone and temporalis fascia were harvested and used to cover the bony defect in a fascia-bone-fascia method. On the right, a large 5 cm defect in the mastoid tegmen was covered using similar method of fascia-bone-fascia. Pathology study of the amputated brain tissue showed inflamed neuroglial, connective and epithelium tissue and confirmed the diagnosis of encephalocoele.

Post-operatively, the patient's subjective hearing recovered fully and has not been using his hearing aid ever since. One year follow-up audiogram showed an improvement in air conduction from 30 to 25 dB and from 45 to 25 dB in his right and left ear respectively. Bone conductions were improved from 20 to 10 dB in the left and 10 to 5 dB in the right ear. He is also free from other symptoms of clear rhinorrhoea and otalgia. He is still being followed up and has so far remained seizure free.

DISCUSSION

Caboche first described the herniation of brain into the temporal bone in the French literature in 1902. Since then, a variety of such terms as brain hernia, brain fungus, brain prolapse, cerebral hernia, fungus cerebri, meningoencephalocoele or encephalocoele have evolved to describe this pathology[1].

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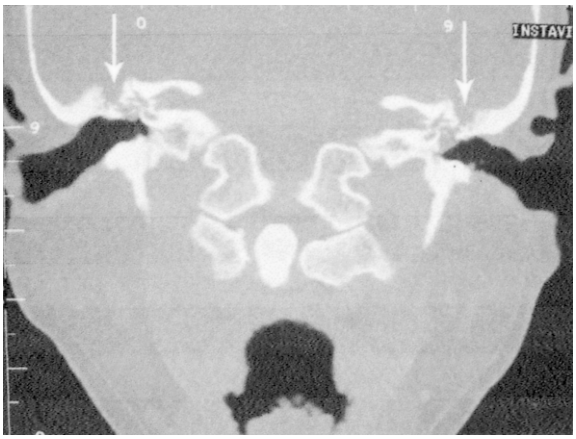

Fig. 1 Coronal CT image shows large defects in the tegmens (arrows).

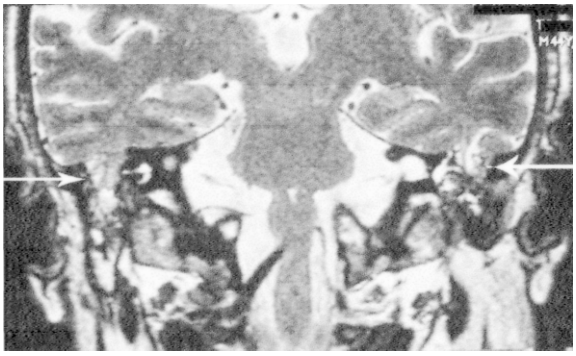

Fig. 2 Coronal T2-W MR image shows bilateral herniated temporal lobe into the middle ear cavity (arrows).

Meningoencephalocoele herniation into the middle ear is a rare condition that can develop in association with infection, previous surgery[2], head trauma or spontaneously. It occurs most frequently in association with previous ear surgery and chronic otitis media[1]. Spontaneous temporal encephalocoele may be further divided into congenital or idiopathic. The pathogenesis for the latter remains enigmatic. Post mortem and temporal bone studies have shown that the incidence of defects in the tegmen and petromastoid segment can be as high as 34% with 20% being bilateral[3]. Bony defects may be related to deficiencies in adult temporal bone remodeling, aging, increased intracranial pressure, low-grade inflammation[1] and erosive effects of aberrant arachnoid granulations[4]. However, a defect alone is not sufficient to promote herniation, as incidence of encephalocoele is far less than that of tegmen defects[1]. Normal dura is structurally strong, capable of supporting brain even over large defects. It is thought that a predisposing factor for brain tissue herniation into the middle ear can be the combination of a bony defect in the tegmen with dural deficiency on the same location[5]. Incidence of congenital encephalocoele is estimated at 1:3,000 to 10,000 live births, favouring females 2.3:1[6]. The majority occur in the posterior, occipital, or parietal regions, which account for some 75 to 80%, while the remainder were found in the skull base of which the most common involve the temporal bone and the middle fossa[7,8]. Congenital defects resulting from malformation and incomplete closure of fissures can result in communication between the middle ear space and middle cranial fossa. These defects can occur both in the mastoid portion of the temporal bone and the bony labyrinth[9]. Our patient fits well into the category of spontaneous encephalocoele, although whether his encephalocoele is congenital or idiopathic remains unknown.

Clinical presentation of a middle ear encephalocoele may include spontaneous cerebrospinal fluid (CSF) otorrhoea and/or rhinorrhoea, mass behind the tympanic membrane, hearing loss, meningitis, temporal lobe epilepsy[10], aphasia[10] and facial paresis[11]. The most common clinical picture, however, is that of a conductive or sensory hearing loss with a clear draining ear or a serous otitis media[1,2,12]. CSF may flow into the mastoid cavity or middle ear by various defects and find its way into the external ear canal through perforated tympanic membrane to present as otorrhoea or through the eustachian tube to present as rhinorrhoea[9]. The patient in discussion did have most of the reported symptoms and signs such as conductive hearing loss, seizures, mass and fluid in the middle ear and rhinorrhoea. Diagnosis may be obscure and is based on a high index of suspicion. Differential diagnoses include cholesteatoma, chronic otitis media, post surgical granulation tissue, cholestrol granuloma and serous mastoiditis.

Most series recommend CT and MR imaging as the confirmatory tests[12-14]. High resolution CT (HRCT) obtained at right angles to the tegmental plane, posterior and middle ear fossa and Eustachian tube can define the presence and size of the bony dehiscence. However, it is quite impossible on CT to differentiate the temporal herniation from cholesteatoma, granulation tissue and cholesterin granuloma. MR imaging is the method of choice to evaluate such lesions[13]. Sagittal MR imaging with pregadolinium and postgadolinium sequences would show the encephalocoele to be isointense, while cholesteatoma to be hypointense, and granulation tissue to be hyperintense, to the brain on the post contrast images.

Management of temporal encephalocoele is surgical. Three surgical approaches have been described: transmastoid (TM), middle cranial fossa (MCF), and combined transmastoid and middle fossa approach. Some series advocate the TM approach for small defects and the combined TM and MCF approach for large defects[1,2,12], as in the case of our patient. The fundamental principle of repair is to provide a compliant tissue seal for CSF leak and prevent

further herniation of brain. Numerous materials for repair have been described. Fascia (temporalis or fascia lata), muscle, cartilage, bone, perichondrium, proplast, fibrin glue have all been used either singly or in combination[1]. Herniated brain tissue that is strangulated is non functional and can be safety resected with no residual dysfunction[2,12]. Morton et al reported three patients with temporal encephalocoele presenting with medically intractable temporal lobe epilepsy recovered after operation[10]. The post-operative long-term prognosis of our patient's seizure requires further monitoring.

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