*The Journal of Laryngology & Otology (2008), **122**, 1168–1174.*

doi:10.1017/S0022215108001990

Main Article

Management of temporal bone meningo-encephalocoele

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Abstract

Meningo-encephalocoele of the temporal bone, also known as fungus cerebri, is a rare occurrence in clinical practice. We present a series of 13 patients with chronic otitis media who suffered brain herniation into the mastoid cavity. We also discuss the presentation and management of brain herniation with or without cerebrospinal fluid leak.

Study design: Retrospective.

Methods: Among 963 cases undergoing revision mastoid surgery, 13 patients suffered brain herniation. These cases were identified and analysed.

Results: All 13 patients' initial diagnosis was chronic suppurative otitis media with cholesteatoma, and all had undergone previous mastoid surgery resulting in a defect in the tegmen and weakening of the dura mater. The revision procedures performed included 10 (76.9 per cent) modified radical mastoidectomies without ossicular chain reconstruction and one (7.6 per cent) modified radical mastoidectomy with ossicular chain reconstruction; two (15.3 per cent) patients required a blind sac closure. Brain herniation and/or cerebrospinal fluid leak were repaired by a transmastoid ± minicraniotomy procedure.

Conclusions: Injury to the tegmen and dura should be avoided during surgery for chronic middle-ear disease. Cerebrospinal fluid leaks, if encountered, should be managed in the same surgical session. The transmastoid approach is helpful in repairing defects smaller than 1 cm in diameter, whereas the combined transmastoid-minicraniotomy approach provides good access when closing defects larger than 1 cm in diameter and also enables auto-calvarial grafting.

Key words: Meningo-Encephalocoele; Otologic Surgical Procedures; Mastoidectomy; Chronic Otitis Media

Introduction

Meningo-encephalocoele is defined as the herniation of brain tissue, with its meningeal covering, into the mastoid cavity and/or the middle ear. Caboche described the herniation of brain into the temporal bone as early as 1902 in the French literature.[1] Many terms have been used by various authors to describe the same pathology, such as brain hernia, brain fungus, brain prolapse, cerebral hernia, meningo-encephalocoele, encephalocoele and fungus cerebri.

Herniation of brain tissue into the mastoid cavity mainly occurs in cases of chronic otitis media, especially following surgery. Other aetiologies claimed to be predisposing factors include: chronic inflammation, trauma, neoplasia, congenital defects, irradiation and degenerative conditions.[2]

During the early part of the twentieth century, brain herniation was frequently related to otitis media and brain abscess. The management of brain abscess was by trephining through the infected mastoid and dura and draining the abscess. This resulted in delayed brain herniation. However, the incidence of this condition dropped significantly due to the introduction of exploratory trephination through a clean field, advocated by Dean and Rand.[3]

Although the incidence of brain herniation has decreased dramatically over the years, cases have been reported throughout the twentieth century. Table I lists cases reported in the literature, along with their predisposing factors.

Clinical features of meningo-encephalocoele

When dural defects occur as a result of mastoid surgery, or due to any other cause, the possible complications include cerebrospinal fluid (CSF) otorrhoea or otorhinorrhoea, seizures, meningitis, brain abscess, epidural abscess, and, rarely, death. Sometimes, brain herniation with or without CSF leak may be clinically silent, but may present with classical signs and symptoms of chronic middle-ear disease, such as hearing loss, chronic otorrhoea, tinnitus and vertigo.[5,7]

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Accepted for publication: 20 December 2007. First published online 17 March 2008.

TABLE I

REPORTED CASES OF BRAIN HERNIATION

Year	Author	Patients (*n*)	Remarks
1960	Schurr	3	PM
1963	Blatt	6	PM
1969	Stout *et al.*	1	PM
1969	Baron	3	PM
1970	Dedo & Sooy	41	PM
1971	Levy *et al.*	1	SH
1977	Fernandez *et al.*	4	PM
1978	Paperalla *et al.*	10	4 PM 6 COM
1979	Glasscock *et al.*	11	8 PM 3 COM
1995	Aristegui *et al.*[4]	27	PM
1996	Lundy *et al.*[5]	19	11 SH 6 COM 2 PT
1999	Escada *et al.*	3	PM
2001	Paleri & Watson	2	SH
2001	Fontes *et al.*	4	3 PM 1 COM
2002	Amoros *et al.*	2	SH
2002	Manolidis	29	28 COM 1 PM

PM = post-mastoidectomy; SH = spontaneous herniation; COM = chronic otitis media; PT = post-trauma

Diagnosis of meningo-encephalocoele requires a high index of suspicion. The usual findings are the presence of a previously operated upon mastoid cavity which is infected and moist, with granulations or cholesteatoma. There is usually a mass protruding into the middle ear or mastoid cavity and external auditory canal through a defect in the tegmen. Classically, this mass is pulsatile, smooth and surrounded by a watery discharge (CSF), and a pulsation may be elicited by the Valsalva manoeuvre.

The differential diagnosis of meningo-encephalocoele includes cholesteatoma, chronic otitis media, post-surgical granulation tissue, cholesterol granuloma and serous mastoiditis. Occasionally, the mass may herniate into the middle ear and the drum remain intact, with a CSF leak into the middle ear, presenting as otorhinorrhoea. This may be confused with serous otitis media. Reports have suggested that many such patients undergo repeated grommet insertions and experience recurrent meningitis.[8]

Management involves a thorough microscopic examination in order to ascertain the details of the mastoid cavity and middle ear. Audiological investigation may reveal either a conductive hearing loss (if pathology is limited to the middle ear, with CSF accumulation) or sensorineural hearing loss (if there has been involvement of the labyrinth during prior surgery, or due to the disease itself). The availability of computed tomography (CT) and magnetic resonance imaging (MRI) with gadolinium contrast has greatly assisted diagnosis of brain herniation. Computed tomography scanning provides details of bony anatomy, alerting the surgeon to tegmen defects and soft tissue herniation through the defect. Magnetic resonance imaging supplies improved soft tissue resolution, compared with CT, and helps to differentiate between recurrent disease in the mastoid and brain herniation. Together, CT and MRI are the 'gold standards' for diagnosis of meningo-encephalocoele. Magnetic resonance imaging also has a role in differentiating brain herniation from cholesteatoma, granulation tissue and cholesterol granuloma. An encephalocoele appears as isointense, a cholesteatoma as hypointense and granulation tissue as hyperintense, compared with brain tissue, on post-contrast images.[5,9,10] Although most instances of brain herniation have involved the temporal lobe (i.e. cerebrum), the senior author has previously reported one patient with cerebellar herniation.[11]

Materials and methods

A retrospective case review of 963 cases of revision of chronic ear disease, undertaken from 1990 to 2007, was conducted. Thirteen cases of brain herniation into the temporal bone were identified. These cases were reviewed, and data were extracted concerning the primary diagnoses, initial procedures, type and number of subsequent operations, follow up, and epidemiological data on age, sex and side (Table II).

The study group included six female and seven male patients ranging in age from seven to 56 years,

TABLE II

STUDY SUBJECTS: DEMOGRAPHIC AND CLINICAL INFORMATION

Pt no	Age	Sex	Side	Previous surgery? (*n*)	Time since last surgery	Otalgia	Facial pain	CSF leak	Giddiness or imbalance	Headache	Hearing loss type
1	7	F	L	2	4 y	+	−	−	+	+	SN
2	22	M	R	1	3 y	−	−	−	−		SN
3	26	M	R	1	2 y	−	−	−	−		M
4	29	F	L	1	7 y	+	−	−	+	−	SN
5	31	F	L	2	5 y	+	−	−	−	−	SN
6	34	F	L	1	10 mth	−	−	−	−	−	M
7	32	M	R	1	13 y	+	−	−	+	+	SN
8	40	M	L	1	2 y	−	−	−	−	−	C
9	43	F	R	1	8 y	−	−	−	−	−	M
10	41	M	R	1	3 mth	−	−	−	−	+	SN
11	44	M	L	1	3 y	−	−	+	+	−	SN
12	45	M	R	4	1 y	+	+	+	+	−	SN
13	56	F	L	2	5 y	+	+	−	−	−	SN

F = female; M = male; L = left; R = right; y = years; mth = months; SN = sensorineural; M = mixed; C = conductive

https://doi.org/10.1017/S0022215108001990 Published online by Cambridge University Press

with a mean of 34.6 years. Seven patients (53.8 per cent) had problems on the left side and six (46.2 per cent) on the right side. All patients had a history of previous mastoid surgery for management of chronic otitis media, performed elsewhere. Three of them had had two previous surgical procedures, and one patient had had four procedures. The most recent surgery had been undertaken three months previously, and the least recent 13 years previously. The mean time interval between previous surgery and the study date was 4.16 years (Table II).

Six of the patients had otalgia as the presenting complaint, and five had giddiness. Two patients had facial pain as an associated complaint, and three had CSF leak. All the patients had undergone a canal wall down procedure for previous cholesteatoma management.

Clinically, the most important sign in the diagnosis of brain herniation was the presence of a smooth-walled swelling or bulge in the mastoid cavity, surrounded by granulation, pus or clear fluid (Figure 1). The presence of pulsations, or pulsation induced by the Valsalva manoeuvre, was not demonstrable. Only one patient presented with a post-auricular swelling without acute symptoms, which mimicked a subperiosteal abscess but was in reality a herniating cerebellum and sigmoid sinus. The diagnosis was confirmed by CT and ultrasonography.

All patients underwent CT to identify the site and size of the bony defect. The smallest defect through which the brain had herniated was 7.0 mm in diameter and the largest was 3 cm. The mean defect size was 1.5 cm. Twelve patients had cerebral (temporal lobe) herniation through the tegmen tympani and one patient had cerebellar herniation.

Pre-operative audiometry showed only one (7.6 per cent) patient with conductive hearing loss, three (23 per cent) with mixed hearing loss (with mean bone conduction at 45 dB pure tone average) and nine (69.2 per cent) with profound sensorineural deafness.

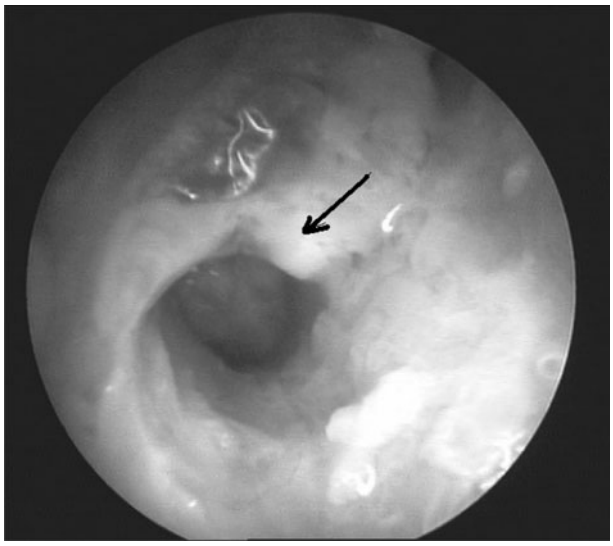

FIG. 1

Otoscopic photograph showing a smooth mass bulging from the roof of the mastoid cavity, partially obscuring the view of the tympanic membrane, with surrounding otorrhoea.

The surgical procedure performed was revision mastoid exploration. Five (38.4 per cent) patients underwent closure of the defect through the transmastoid route, seven (53.8 per cent) underwent closure of the defect through a combined transmastoid and middle cranial fossa approach through a minicraniotomy (two of these patients underwent blind sac closure) and one (7.6 per cent) underwent a posterior craniotomy and repair. The decision on the approach used for the repair was based on the site and size of the defect. Defects smaller than 1 cm in diameter were repaired using the transmastoid approach, and those larger than 1 cm underwent a combined transmastoid and minicraniotomy approach.

The transmastoid route involved a revision canal wall down procedure to expose the defect in the tegmen and the herniated brain tissue. The herniated tissue was then separated from the squamous epithelium by dissection and amputated using bipolar diathermy. The result was a very clearly delineated defect, which was subsequently repaired using conchal cartilage and a layer of temporalis fascia. This procedure was preferred when the defect was 1 cm or less in diameter.

Patients requiring a combined transmastoid and minicraniotomy procedure underwent the transmastoid procedure as described above; a rectangular window was then made in the squamous part of the temporal bone to give access into the middle cranial cavity and also to harvest bone, which was then utilised as a graft to repair the defect. The brain with its dural covering was retracted off the floor of the middle cranial fossa and the defect was repaired with a three-layer (fascia–bone–fascia) closure. That is, autologous, calvarial bone graft wrapped in lyophilized (cadaver) duramater which was placed over the defect in the floor of the middle cranial fossa through the window in the squamous part. The tegmen defect was then covered with a layer of temporalis fascia in the mastoid cavity; flaps were replaced and packed with bismuth iodoform paraffin paste (BIPP) soaked gauze (Fig 2–6).

Two patients required blind sac closure, the indication being multiple previous surgical procedures, profound sensorineural deafness and non-availability of healthy skin in the medial part of the cavity. In the authors' experience, it has been noticed that, whenever there is circumferential absence of skin in the medial part of the mastoid cavity, the cavity rarely heals completely and otorrhoea often persists. A canal wall down procedure was performed, the herniated part of the brain amputated, the mucosa cleared from the mastoid cavity and middle ear, the eustachian tube orifice sealed with cortical bone, the tegmen defect repaired, and the resultant cavity denuded of squamous epithelium and mucosa. Two lateral-based skin flaps were created, being elevated from the underlying cartilage in the remnant external auditory canal and then sutured to each other to form a blind sac. A flap of perichondrium on the medial

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FIG. 2
Illustration showing postaural exposure of the mastoid cavity with encephalocoele and site of minicraniotomy.

surface further reinforced this blind sac. The resultant cavity was filled with abdominal fat in one patient and with a periosteal, temporalis muscle flap in the other.

These patients then underwent routine mastoid bandaging after the wound was closed and the ear canal and mastoid cavity packed with bismuth iodoform paraffin paste, except for those patients who had undergone blind sac closure. All patients were discharged after one week, having received antibiotic cover (intravenous ceftriaxone and amikacin). Sutures were removed on the fifth post-operative day. The bismuth iodoform paraffin paste pack was removed on the 21st post-operative day in all patients except those who had undergone blind sac closure. The patients were followed up over the next two months to assess cavity healing. All patients had good healing at the end of three months. The patients

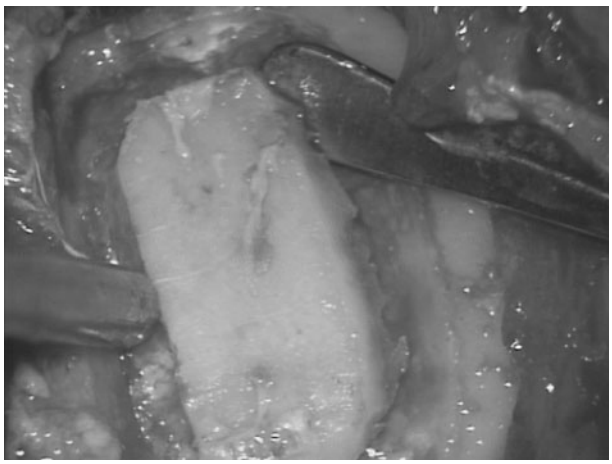

FIG. 3
Operative photograph showing harvesting of autocalvarial bone graft at minicraniotomy.

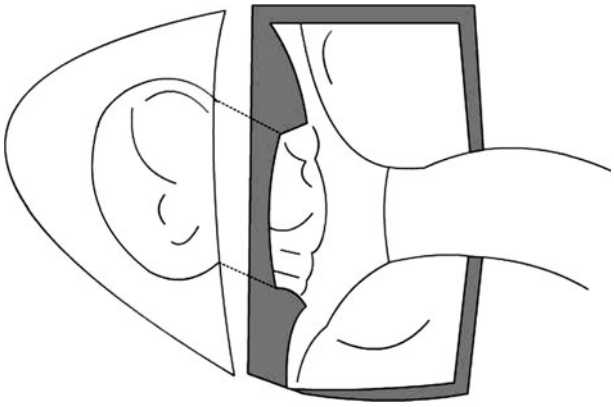

FIG. 4
Illustration showing retraction of the brain from the middle cranial fossa through the minicraniotomy, the defect in the tegmen and the encephalocoele.

were followed up for a minimum of one year and a maximum of four years.

Findings during surgery

One patient had a mobile stapes with no other ossicles; in this patient, a homo incus was used to transpose onto the stapes head as an ossiculoplasty. In all the other patients, there were no ossicles available; although the footplate was intact in nine (69.2 per cent) patients, no ossiculoplasty was performed as they had sensorineural deafness. The other common finding was the presence of a lateral semicircular fistula, in six (66.6 per cent) of the nine patients with sensorineural deafness. The dome of the lateral semicircular canal was the commonest site. The facial nerve was found to be exposed in the vertical segment in three (23 per cent) patients, although none had any pre-operative facial palsy. The sigmoid sinus was exposed in two (15.3 per cent) patients.

The patients had been followed up for a minimum of one year and a maximum of four years. During

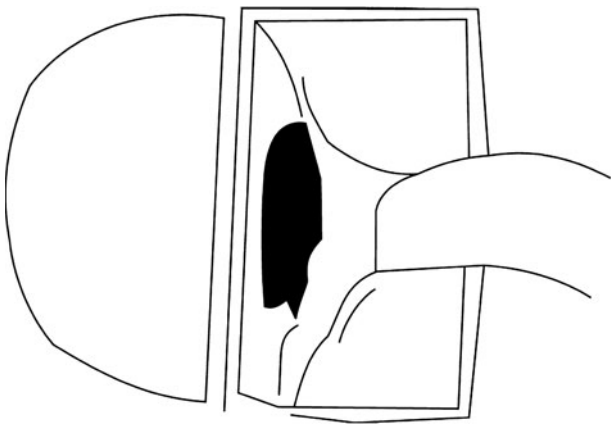

FIG. 5
Illustration showing the tegmen defect through the minicraniotomy window after amputation of the encephalocoele.

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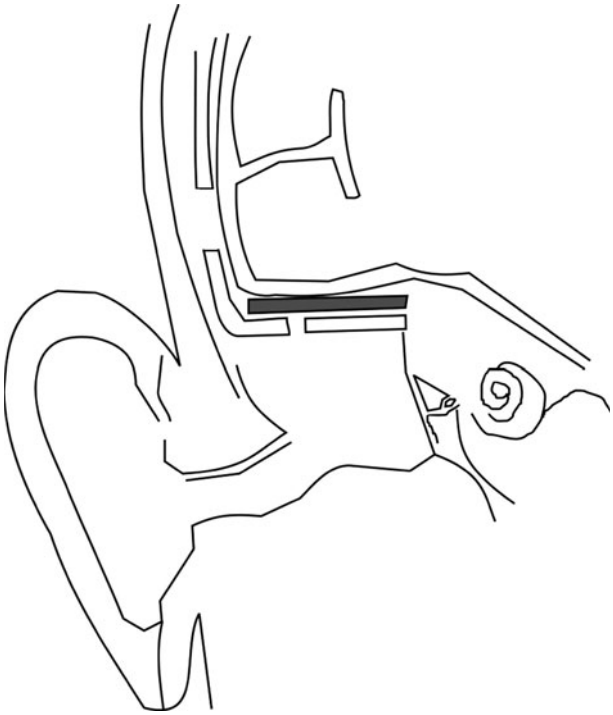

FIG. 6
Illustration showing the final appearance in the coronal plane, following repair of the tegmen defect.

follow up, they were examined clinically and investigated using CT (Figure 7). Twelve patients had no recurrence of herniation or CSF otorrhoea (Figure 8). The patient with cerebellar herniation had a recurrence within one year but was lost to follow up thereafter.

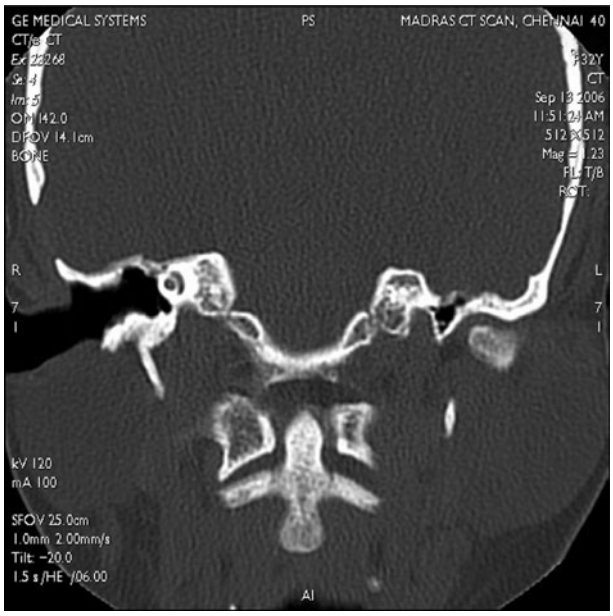

FIG. 7
Post-operative coronal computed tomography scan of a patient one year after repair using a combined transmastoid and minicraniotomy approach.

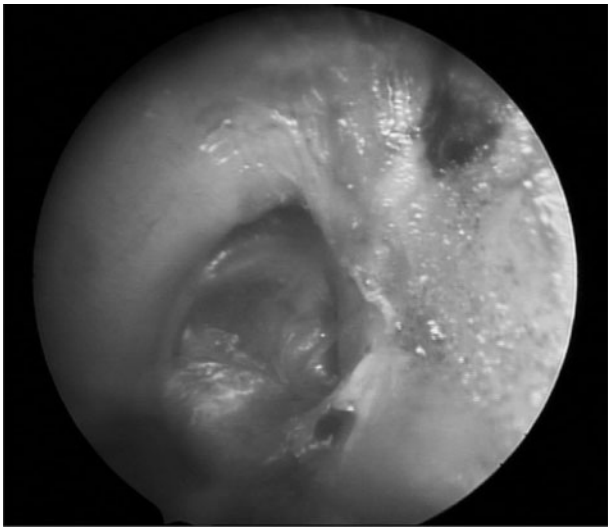

FIG. 8
Post-operative otoscopic photograph showing a well healing cavity with no signs of encephalocoele or otorrhoea.

Discussion

In the study by Manolidis, the incidence of dural herniation and encephalocoele in patients with chronic otitis media was found to be very high.[6]

The predisposing factors were: prior ear surgical procedures such as mastoidectomy (radical or modified radical), meningitis, cholesteatoma and active chronic otitis media. Iatrogenic injuries incurred during radical and modified radical mastoidectomy account for between 59 and 77 per cent of encephalocoele cases.[1,4] In our series, each of the 13 patients had undergone previous mastoid surgery. A dural defect may have various aetiologies, such as spontaneous, congenital, traumatic, iatrogenic, inflammatory or idiopathic. In our study, all dural defects appeared following surgical trauma. The most common site of dural defect is the tegmen plate.[12] Twelve of our 13 patients had a defect in the tegmen; only one patient had herniation of the posterior cranial fossa contents.

Diagnosis of meningo-encephalocoele is based on clinical evaluation and a high index of suspicion. High resolution CT and MRI can be used as preoperative evaluation modalities in order to confirm the diagnosis and assess the extent of the lesion. In certain cases in which differentiation between granulations, cholesteatoma and brain herniation is not easily made via CT, MRI with gadolinium contrast can be utilised.[5,9,10] Scintigraphy is very sensitive for CSF leak but is unhelpful in identifying the leak site. When combined with CT and MRI, radionuclide cisternography is useful, but it is not necessary for every patient. Jackson *et al.* state that as many as 89 per cent of encephalocoeles can be accurately diagnosed.[1]

The clinical presentation of meningo-encephalocoele may encompass spontaneous CSF leaks (otorrhoea and/or rhinorrhoea), a mass behind the tympanic membrane or in the mastoid cavity,

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hearing loss, meningitis, temporal lobe epilepsy, aphasia or facial paresis.[13]

- **This paper reports clinical experience in the management of herniation of brain tissue, with its meningeal covering, into the mastoid cavity and/or middle ear**
- **This condition usually occurs in patients with chronic otitis media, commonly following surgery, although other, rare aetiologies have been described**
- **Diagnosis is mainly clinical and requires a high index of suspicion; it can be confirmed radiologically via computed tomography and magnetic resonance imaging with gadolinium contrast**
- **Different surgical approaches have been used to repair such defects, e.g. transmastoid, middle cranial fossa and minicraniotomy. Two patients required surgical blind sac closure; this has not been previously reported in the management of this condition**
- **In this study, cerebrospinal fluid leak was not a common accompanying symptom, in contrast with other reports**

Most of our patients presented because of ear discharge and hearing loss, despite previous surgery. A clear, watery ear discharge was an additional symptom in only two patients. No patient had a history suggestive of meningitis or secretory otitis media, in contrast with other authors' reported experiences.

A high degree of suspicion is needed when considering a diagnosis of meningo-encephalocoele. Suspicion should especially be aroused by a smooth bulge or swelling arising from either the roof or posterior wall of the mastoid cavity.

Surgery is the primary modality of treatment, and different techniques and approaches have been described for the repair of the dural defect. These include the middle fossa craniotomy approach, the transmastoid approach, and the combined transmastoid and middle fossa approach.[5] A combined transmastoid and middle cranial fossa approach provides the best exposure of the defect in the tegmen plate.[8] Adkins and Osguthorpe have advocated the minicraniotomy procedure,[14] while May *et al.* recommend a keyhole craniotomy.[15] The disadvantage of keyhole craniotomy is that any defect in the anterior attic will not be accessible.

Kuhweide and Casselman have described the surgical management of encephalocoele by five-layer closure, using a transmastoid approach with minicraniotomy.[16] They concluded that this procedure offers a relatively simple and reliable method for repair, without the inherent risks of middle fossa craniotomy. The herniated brain tissue is non-functional and is thus usually removed after shrinkage with diathermy.[4,5] Various materials can be used to seal the defect, in different combinations, such as temporalis fascia, conchal cartilage, titanium mesh, cadaveric dura, muscle, bone, Proplast® and fibrin glue.[1]

Twelve of the 13 patients in our series had an uneventful recovery following either a transmastoid or a combined transmastoid and minicraniotomy approach. We advocate a flexible approach in the management of such patients; some may require only a transmastoid approach, while others may require a minicraniotomy also. The emphasis lies on achieving complete, secure, firm closure of the defect, in layers. We have had good results with both temporalis fascia, cartilage combination in defects less than 1 cm and a sandwich of lyophilised dura, auto-calvarial bone graft and temporalis fascia for defects bigger than 1 cm.

Conclusion

Brain herniation into the mastoid cavity following previous mastoid surgery has been documented for over a century. With developments in science, technology and surgical techniques, the incidence of this condition has reduced. Nevertheless, one unfortunately still comes across this pathology within a busy otology practice. Most often, it is due to an unattended dural injury sustained during mastoidectomy. These measures will go quite a way towards preventing development of meningoencephalocoeles. If it does occur, the surgeon should desist from continued drilling (with a cutting or diamond burr) or cautery over the exposed dura mater, as these will cause a loss of dura mater toughness. As much as is possible, these defects should be covered in the same surgical sitting, using autologous cartilage grafts obtained from the concha or the tragus. These measures will go quite a way towards preventing dural injury from occurring. Regarding the surgical management, most publications have reported good results with various materials; the authors prefer the procedure detailed above.

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Professor K K Ramalingam takes responsibility for the integrity of the content of the paper.
Competing interests: None declared