Meningo-encephalocoele of temporal lobe-management by blind SAC closure

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Abstract Brain herniation into the middle ear and mastoid is rare but is a described complication of chronic ear disease. The diagnosis is mainly clinical and requires a high index of suspicion. This can be confirmed by imaging studies. Different surgical modalities have been described in managing this condition. We present a case managed by combined trans-mastoid mini-craniotomy approach and blind sac closure.

Keywords Meningo-encephalocoele · Mini-craniotomy · Blind sac closure

Introduction

Fungus cerebri is herniation of meninges and/or brain into the middle ear and mastoid cavity through a bony defect in skull. Usually the defect is in the tegmen. Caboche was the first to describe the herniation of brain into the temporal bone in French literature in 1902 [1]. Since then, a variety of terms such as brain hernia, brain fungus, brain prolapse, cerebral hernia, fungus cerebri, meningo-encephalocoele or encephalocoele have been used to describe this condition¹. The etiology of brain herniation has been attributed to temporal bone trauma, iatrogenic injuries, cholesteatoma, chronic otitis media, congenital cranial base defect, arachnoid granulations, neoplasia, irradiation and idiopathic causes [2–6]. Of these, injuries to tegmen and dura by cutting burr during mastoid surgery are probably the commonest cause. It has been encountered most frequently following previous ear surgery for chronic otitis media [7]. The most common site of temporal lobe herniation is through a defect in the tegmen tympani and tegmen antri [7]. It is found in less than 1% of revision otologic surgeries [8]. The modality of treatment for these patients is mainly surgical. The approaches include the middle cranial fossa craniotomy approach, transmastoid approach and combined approach [9]. The defect in the tegmen can be closed using different materials such as autografts – cartilage, calvarial bone and temporalis fascia and fascia lata etc. Homografts such as cadaver duramater or allografts such as proplast, fibrin glue etc have also been used [1].

We present a patient who was managed using a combined trans-mastoid, mini-craniotomy approach and the defect closed using sandwich graft of Lyophilised duramater, autograft calvarial bone graft and temporalis fascia. The external auditory meatus was closed as a blind sac.

Materials and methods

A 56-year-old female presented with history of left ear discharge and intermittent pain for the past 15 years. She also

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had loss of hearing and occasional giddiness. The discharge was watery and at times, mucopurulent or blood stained and occasionally the ear was dry. The patient had three mastoid surgeries elsewhere for her aural complaints 19 years; 16 years and 14 years back respectively. No details of the surgeries were available. There was no history suggestive of any other extra cranial or intracranial complications. She was known to be suffering from diabetes and hypothyroidism and was on medication with good control. General condition of the patient was good. On examination of the left ear- there was multiple post-aural scar suggesting previous surgeries, the meatus was wider than the other side and soggy due to discharge. The external auditory canal was wider than the opposite side and a soft tissue mass was seen arising from posterior and superior part of external auditory canal. A large mastoid cavity was noticed which was filled with the mass, clear discharge and debris. On probing, the mass was insensitive to touch and pressure, wide based and arising from the roof of the cavity. The medial part of external auditory canal and tympanic membrane were not seen because the mass was obscuring the view. Examination under microscope of ear was performed and the above findings were confirmed. Tuning fork tests showed severe nerve deafness on left side and normal hearing on the right side. Fistula sign was negative; there was no facial palsy or neck rigidity. Pure-tone audiometry confirmed severe nerve deafness on the left side while the hearing in right ear was normal. MRI of temporal bone was done which showed mastoiditis on the left side with soft tissue lesion occupying the epitympanum, mesotympanum and mastoid cavity. The mass was found to be continuous with the temporal lobe on the left side. The tegmen defect shown in Computed Tomography scan measured 1.5×2 cm in dimension (Fig. 1). No ossicles were seen.

Fig. 1 Preoperative CT scan of temporal bones, coronal section showing a defect in the left tegmen through which the brain tissue has herniated into the external auditory canal (mastoid cavity). [White arrow]

Patient was informed about the huge mastoid cavity and profound sensorineural deafness in the diseased ear and need for revision mastoidectomy and repair of tegmen defect. In view of three previous unsuccessful mastoid surgeries the patient was also told about the possibility of blind sac closure, under general anaesthesia. Informed consent was taken and patient was prepared for surgery.

Procedure The patient was operated under oro-tracheal intubation and general anaesthesia. The patient was in supine position with the head turned to the opposite side. The operative part was painted with povidone iodine and draped. A Wilde's post-aural incision was made through the scar. The periosteum was elevated off the mastoid and an irregular mastoid cavity was exposed. A smooth surfaced mass with surrounding debris was seen filling the mastoid cavity. On clearing the debris the mass was found to be arising from the roof of the cavity with clear water like fluid discharging from around it. The mastoid cavity was well-saucerised and facial ridge adequately lowered. On elevation of the unhealthy skin over the mass, it was found that the mass was protruding from the cranial cavity through a bony defect in the tegmen plate, which was measuring approximately 1.5×2 cm. There was CSF (Cerebro-spinal fluid) leak from around the mass. The rest of the mastoid cavity was devoid of any squamous epithelium and filled with granulation tissue and mucosa. There were no ossicles in the middle ear. All the granulations and mucosa were removed carefully using a diamond burr. The herniated part of the brain was amputated at its base using bipolar diathermy. A defect of 2×1.5 cm was seen near posterior end of tegmen plate. Temporalis fascia graft was harvested. Temporalis muscle was elevated off the squamous part of the temporal bone. A mini-craniotomy was done and a 2.5×1.5 cm size of temporal bone graft (auto-calvarial graft) was harvested (Fig. 2 with illustrations). Temporal lobe of the brain with its meningeal covering was gently retracted off the floor of the middle cranial fossa through the mini-craniotomy itself and the defect in the tegmen plate delineated. The auto-calvarial bone graft was then wrapped with lyophilized homograft duramater and inserted into the floor of middle cranial fossa through the mini- craniotomy so as to cover the defect in tegmen plate. The defect in the mastoid cavity side was covered with temporalis fascia. As there was no skin found in the remaining part of external auditory canal, it was decided to perform blind sac closure of the external auditory canal. All remnant skin and mucosa in the middle ear and mastoid cavity was carefully removed. Eustachian tube opening was plugged with cortical bone and soft tissue. Sino dural angle was obliterated with conchal cartilage and soft tissue. An inferiorly based musculo-periosteal flap was used to obliterate the mastoid cavity. Haemostasis was obtained. Wound was closed in 2 layers. Postoperative antibiotics and anticonvulsant was given for a period of 1 week. Patient was followed for 3 years and she had no evidence of discharge or recurrence as confirmed by CT Scan [Figs. 3, 4]

Minicraniotomy in squamous part of temporal bone.

Retraction of brain through the minicraniotomy to expose defect and herniated brain *.

Fig. 2 (with illustrations). Operative photograph showing harvesting of autocalvarial bone graft from the squamous temporal bone

Blind sac closure

Exteriorizing the lateral part of external auditory canal as a cul-de sac with or without obliteration of mastoid cavity.

Procedure The lateral based flap of skin from the external auditory canal is elevated all round i.e., 360 degrees.

It is separated from the underlying cartilage. The skin is then everted into the external auditory meatus. External auditory canal was closed using interrupted sutures using 3-0 Vicryl taking care to keep the margins everted (Figure 5). This creates a blind sac of the external auditory canal. The medially based tympanomeatal flap along with epithelium

and tympanic membrane is removed in toto. The undersurface of the blind sac is reinforced by a periosteal flap which is folded over the closed meatus and sutured to the cartilage of external auditory meatus with 2-0 chromic catgut.

Discussion

An encephalocoele is herniation of brain tissue through a skull defect, because the evolution of such a herniation will almost always entail extrusion of meninges, it is often referred to as meningo-encephalocoele. Dural defect can occur due to various causes such as, spontaneous – congenital/ idiopathic, traumatic, iatrogenic and inflammatory [10]. However a defect alone is not sufficient to promote herniation, as incidence of encephalocoele is far less common than that of tegmen defects. 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/mastoid cavity can be the combination of a bony defect in the tegmen with dural deficiency on the same location, which usually occurs due to drilling over the dura or using diathermy over the dura [1]. In this patient also, there was history of multiple previous ear surgeries and CSF leak was seen during the surgery which explains the above defect and dural deficiency. Wooten, Christopher T et al. (2005) in their data reviewing 1130 cases found the incidence of temporal bone encephalocoele to be approximately 1% [20]. They also demonstrated that patient at risk for developing brain herniation and or CSF leak have long standing history of multiple operations. Clinical presentations of meningo-encephalocoele include spontaneous CSF leak (otorrhoea and rhinorrhoea), mass in the mastoid cavity or behind the intact tympanic membrane, hearing loss, men-

Fig. 3 Post operative CT scan of temporal bones, coronal section showing a completely obliterated left mastoid cavity with the defect in the tegmen covered by the autocalvarial graft (white arrow) and the external auditory meatus ending in a blind sac (red arrow).

Fig. 4 Post operative CT scan of temporal bones in axial section showing complete obliteration of the left mastoid cavity and external auditory canal with soft tissue – i.e., blind sac closure (white arrow)

Fig. 5 Operative photograph showing the external auditory canal having been closed in a blind sac

ingitis, temporal lobe epilepsy [12], aphasia [12] and facial paresis [13]. The patient in discussion did not have most of the reported signs and symptoms such as meningitis, facial paralysis but presented with chronic ear discharge, hearing loss and occasional pain. Diagnosis may be obscured and is based on high degree of suspicion. Though different authors have described the presence of pulsations in the herniated brain mass, we did not find such pulsations in our patient. Differential diagnosis includes cholesteatoma, chronic otitis media, cholesterol granuloma etc.

High resolution computed tomography (HRCT) & MRI scans with or without contrast are the most important investigations to localize the defect and to know the extent of defect and to plan surgery. Jackson et al. (1997) have concluded that as many as 89% of encephalocele can be diagnosed accurately with HRCT/MRI.

Surgery is the primary modality of treatment and different techniques and approaches have been described to repair such herniation. They include the middle cranial fossa craniotomy approach, transmastoid approach and combined approach [9]. Some series advocate transmastoid approach for small defect (less than 1 cm) and combined for large defect (greater than 1 cm) [1].

In this patient a combined trans mastoid with mini-craniotomy was performed to repair the defect and blind sac closure was performed as there was no canal skin, patient had no serviceable hearing and the defect was 2 cm in its greatest dimension. The fundamental principle of repair is to provide a compliant tissue seal for CSF leak and prevent further herniation of brain. Numerous methods for repair have been described. Fascia (temporalis or fascia lata) muscles, cartilage, bone, perichondrium, proplast, fibrin glue have all been used either simply or in combination [1]. Herniated brain tissue that is strangulated is nonfunctional and can be safely resected with no residual dysfunction [9, 14]. Kuchweide R et al. (1999) have described the surgical management of encephalocele by a 5-layer closure using a transmastoid approach with minicraniotomy. They concluded that this procedure offers a relatively simple and reliable method for repair without the inherent risks of a middle fossa craniotomy [5].

Adkins and Osguthorpe (1983) have advocated the mini-craniotomy procedure [15], while May et al. (1995) recommend a keyhole craniotomy [16].

Blind sac closures have been traditionally performed for patients with previous history of multiple temporal bone surgeries with cavity problem and non-serviceable hearing loss. El-kashlan et al. (2003) has described this procedure with cochlear implantation in cases of chronic suppurative otitis media with profound nerve deafness [17]. Two traditional techniques have been described in the literatures. Coker et al. (1986) has done this technique for lateral approach to skull base through temporal bone. He has suggested Subtotal petrosectomy with tympanomastoid obliteration with blind sac closure provides a relatively safe and secure closure of the surgical defect in the temporal

bone and eliminates the problems associated with an open mastoid cavity [18]. We, in our case, have followed the above technique. Another method described by Meyerhoff et al. (1988) is modified Rambo technique [19].

Conclusion

In the author's opinion, the combined transmastoid with minicraniotomy approach with blind sac closure provides the best exposure of the defect for repair and the best means to prevent cavity problems in a completely dead ear.

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