MENINGOENCEPHALOCELE OF THE MIDDLE EAR CLEFT

Three tier management

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Abstract: Meningoencephalic herniation into the middle ear is a rare condition that can be life threatening for the patient due to eventual infective intracranial complications. It is characterized by a bony and dural defect localized in the tegmen through which meninges and encephalic tissue can herniate. Meningoencephalic hernia of the middle ear can appear as an insidious complication of Chronic mastoiditis (infective) and its surgery, trauma or spontaneous. High resolution computed tomography (CT scan) and Magnetic resonance imaging (MRI) must be done to evaluate the extent and the contents of the disease. Early and effective surgical repair, the only appropriate treatment, is necessary to avoid complication and recurrence. Middle cranial fossa, mastoid, combined approaches and obliteration of mastoid, external auditory canal and middle ear are the surgical procedures for the repair of hernias. Various materials viz. fascia/ muscle/fat / cartilage/ bones/ synthetic materials are used. Recurrence of the repaired herniation has been reported. To overcome the problem of recurrence and have a foolproof method, a three tier management is described and suggested.

Abstract: Meningoencephalocele, brain herniation

INTRODUCTION

Meningoencephalic herniation into the middle ear (MHME) is a rare condition. It can result from ear surgery, infection, and head trauma or can be spontaneous. The usual sites of herniation are tegmen antri and tegmen tympani. Meningoencephalic hernia of the middle ear can appear as an insidious complication of chronic mastoiditis (atticoantral disease). It is usually asymptomatic and is coincidentally discovered at the time of surgery. Immediate and effective surgical treatment is indicated in order to avoid the recurrence and potential complications of this disease. Herniation of meningeal and/or encephalic tissue into the middle ear is pathology, which can be life-threatening for the patient because of eventual infective intracranial complication. These hernias are classified as localized pedunculated or sessile (a diffuse fungoid), with associated factors such as viability of herniated brain, infection, cerebrospinal fluid leak, and neurological complications. The associated complications require extensive surgery for eradication, and the potential for morbidity and mortality is significant. These hernias are repaired by otological,

neurosurgical and combined procedures using various implants and materials (autologous/homologous/synthetic). Even then these hernias can recur especially in large defects. Jackson et al in 1997 reported a recurrence after primary repair to be 10%¹. The main reasons for these recurrence and failures have been attributed to the slipping the material to one side or into the mastoid cavity if patient strains too much to increase the intracranial pressure. This problem can also recur; if the grafting material is smaller than the defect or the graft is placed on the mastoid side of the tegmen. This prompted us to report and suggest a three tier grafting to support the brain tissue with meninges and prevent recurrence.

CASE REPORT

S.C. 42 years female complained of a sense of fullness in the right ear, impaired hearing and discharging ear for the last so many years. She was operated upon thrice for chronic suppurative otitis media with cholesteatoma at different centers. Examination revealed a pulsatile soft mass filling the external auditory canal

Fig 1 (a): Pre-operative photograph showing mass in ear canal

Fig 1 (b): Pre-operative C.T. Scan, arrow showing the bony defect and mass in ear canal

Fig II (a): Pre-operative photo showing the bony defect.

Fig II (b): Line diagram showing three tier management

up to 90 percent. (Fig. 1a) There was scanty foul smelling blood stained discharge in the right ear. Tympanic membrane could not be visualized fully even on pressure reduction of the mass under microscope. Audiogram showed moderate conductive hearing loss in the affected ear. Computed tomography (Fig. 1 b) petrotympanic complex done showed an extensive opacity in the external auditory canal, middle ear cavity, mastoid antrum and mastoid cavity with wide bony defect in the tegmen tympani and tegmen antri with a possible communication to the posterior cranial fossa. Drum scutum and ossicles were absent (Radical Mastoidectomy). A soft tissue mass containing brain tissue and covering membranes was herniating in the external and middle ear. A diagnosis of Meningoencephalocele was established. The patient was taken up for surgery after necessary investigation. Via a wide post aural incision, mastoid cavity was explored. Manual reduction of the mass was tried after lowering intracranial pressure. The mass was adherent to the bony defect in the tegmen. The mass was excised. A wide bony defect (Fig. II a) was seen. The bony defect was repaired in three tiers as shown (Fig II b). Superiorly wired mesh was placed covering the margins of the bony defect on all sides. This was followed by locally available conchal cartilage with intact mucoperichondrium on one side and then covered by a temporalis fascia graft tucked onto the cranial side of the defect. Another piece of temporalis fascia was placed to cover the defect inferiorly. After wide meatoplasty the cavity was packed with antibiotic impregnated rolled gauge to provide extra support to the repaired defect. The packing was removed after three weeks. Antibiotic eardrops locally were advised. The patient was advised not to increase the intra-abdominal pressure and was prescribed stool softeners for four weeks. On three-year follow-up no recurrence was noticed. Repeat computed tomography scan petrotympanic complex done after eight months of surgery did not show previously seen soft tissue opacity containing brain tissue and meninges. (Fig. II c).

DISCUSSION

Meningoencephalic hernia of the middle ear is usually asymptomatic and diagnosis is accidental. This unusual condition may precipitate numerous otological problems such as hearing loss, trapped squamous epithelium, and the potential

for meningitis or encephalitis². The mastoid tegmen is the most common site of defects (41%) and the most common etiologic factor is cholesteatoma (84%).³ Cerebellar herniation into the mastoid through the posterior aspect of temporal bone as a result of chronic suppurative otitis media and mastoid surgery has been reported⁴. Among the 50 patients with tegmen defect associated with chronic otitis media, 15 patients presented with brain herniation associated with the bony defect and 10% developed complications. Fourteen patients had undergone previous mastoid surgery for chronic otitis media.⁵ In a series of 35 patients, the etiologic factor was previous surgery in twenty-six cases.⁶ Computerized tomography allows the suspicion of brain herniation, but a definitive diagnosis can best be established with an Magnetic Resonance Imaging (MRI) study. High Resolution computed tomography of the temporal bone in particular, can show the exact limits and location of the bone defect, while magnetic resonance imaging allows the nature of the tissue in the middle ear to be determined. MRI and CT scanning can often help clarify the diagnosis preoperatively.

Four different etiological types are possible, viz infective, post-surgical, traumatic and spontaneous. Brain herniation through the tegmen of the middle ear (tegmen antri and tegmen tympani) has a characteristic glistening, shiny appearance. All types are characterized by a bony and dural defect localized in the tegmen through which meningeal and encephalic tissue can herniate. A tegmen defect alone does not produce brain herniation into the middle ear cleft. Edema, hemorrhage and cyst formation invariably occur at the junction of cerebral cortex and grey matter, directly beneath the defect and these force the intervening brain to herniate.⁷ Infarction and necrosis develop in the herniations over the larger cysts, which resolves and scar tissue is formed after 2-7 weeks. Other factors relevant to the pathogenesis of brain hernia include the size of the defect, the thickness of the dura and its integrity and chronic inflammation of the middle ear cleft. Symptoms of brain herniation may include chronic watery ear discharge, earache, headache, hearing loss and epilepsy. Conductive hearing loss may occur due to reduced ossicular mobility or disrupted chain. Obstruction to the aditus ad antrum may provoke mastoid infection or an "aditus block syndrome"⁸. Increased tension on the dura of the herniated brain may induce

headache. The necrosed brain tissue in the herniated bag acts as a nidus for epilepsy. The complication arising from meningoencephalocele are meningitis, brain abscess and pneumocephalus. Treatment planning must avoid intra-cranial complications. Surgery is the only appropriate therapy. Transmastoid and middle cranial fossa are alternative or complimentary approaches, determined by several factors, including the size and the site of the bony defect and the presence of middle ear infection. Posterior tegmental deficiencies are suitable for transmastoid approach. Anterior and medial deficiencies require a combined middle fossa and mastoid approach. Repair of dural laceration requires watertight closure to prevent the occurrence of encephalocele. Options for managing a brain hernia fall into two basic categories, which include reconstruction of the mastoid cavity and obliteration of the mastoid cavity. Herniated brain is non-functioning and can be removed simply by sharply excising it. Any viable brain is reposted back into the middle cranial fossa. If the defect is large, both intradural and extradural repair of the dural defect must be considered. The various materials to repair the defect include autologous or homologous and heterologous.

The cortical bone, auricular, septal and conchal cartilages, autologous or homologous, is the preferred material with temporalis fascia. Wired mesh, silicon or silastic sheet are the heterologous materials used with or without auto/homologous materials available. A pedicled temporalis muscle has been used to support repaired dura through temporal craniotomy.⁹ The explanation for this technique is that muscle avoids the problem of graft necrosis or graft rejection. Different approaches have been described amongst which the transmastoid with or without temporal minicraniotomy and the middle cranial fossa (MCF) are the most frequently reported in literature. The middle cranial fossa approach in patients with large herniations is preferred to avoid infective complications. Of thirty patients, primary repair was successful in all but three patients, two of whom required a second repair¹. Failure of the repair is possible due to the slipping the material to one side or into the mastoid cavity if the patient strains too much to increase the intracranial pressure. This problem can also occur; if the grafting material is smaller than the defect or the graft is placed on the mastoid side of the tegmen. The second alternative is to simply obliterate the middle ear, external auditory canal and mastoid in patients with severe to profound hearing loss. Squamous epithelium is removed from the diseased ear, and the cavity is filled with abdominal fat after the Eustachian tube has been completely obstructed. The auditory meatus is sewn closed. Obliteration ensures a safe, well healed, problem free ear.

CONCLUSION

Meningoencephalocele, brain fungus and brain herniation are the synonymous terms. The herniated mass consists of meninges and brain tissue. It is usually asymptomatic and found accidentally on surgery. It may produce life-threatening complications. Proper, meticulous, prompt and effective surgical repair is necessary. A three tier management is suggested to prevent recurrence and complications in these cases.

Fig II (c): Post operative C.T. Scan done after four months

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