

Meningoencephalocele of the Temporal Bone-A Case Report

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Abstract: This paper reviews the diagnosis and treatment of temporal bone meningoencephalocele, defined as the herniation of meninges or brain tissue into empty spaces within the temporal i.e tympanic or mastoid cavity, through the tegmen tympani or antri respectively. Imaging is the mainstay of the diagnostic process. Management of the condition is surgical, special emphasis on the transmastoid approach. The tegmen was repaired with a multi-layered technique using cartilage, temporalis fascia and tympanomeatal flap.

Keywords: Temporal bone, meningoencephalocele, cerebrospinal fluid leak, transmastoid approach, multi-layered closure

I. Introduction

The most common soft tissue masses in the middle ear are cholesteatomas, granulation tissue, cholesterol granulomas, paragangliomas, high jugular bulb. Rare masses are facial nerve schwannomas, ectopic carotid arteries and meningoencephaloceles. Meningoencephaloceles of the temporal bone is rare and may be misdiagnosed. Autopsy study suggest that 15-34% contain single defect in tegmen of temporal bone[1]. The brain tissue usually becomes necrotic after herniation[2,3]. Meningoencephalocele of the temporal bone most commonly results from traumatic or iatrogenic injury to mastoid tegmen or posterior dura. A congenital encephalocele is rare and the incidence is estimated at 1:3000 to 10000 live births, with female to male ratios of 2.3:1[4]. Occipital encephaloceles are the most common. Frontoethmoidalencephaloceles comprise about 9.8% of all encephaloceles[5]. Skull base encephaloceles account for only 5%[4]. The most common skull base encephaloceles involve the temporal bone and middle fossa. Temporal bone defects may be secondary or primary in nature. Acquired tegmen defects are most commonly due to chronic otitis media with or without cholesteatoma or middle ear & cranial base surgery. Other causes include temporal bone trauma, neoplasia and radiotherapy. They occur predominantly in the floor of the middle cranial fossa at the level of tegmen[6]. Spontaneous temporal bone defects present in 2 major distinct categories based on the age of onset. Congenital temporal bone abnormalities in children include defect in the hyrtl fissure, wide fallopian canal, Mondini dysplasia & patent cochlear aqueduct[7]. Spontaneous temporal bone defects in adults are less common and generally present without previous history of petrous pyramid disease[8].

Patient with meningoencephalocele are either asymptomatic or complain of mild symptoms such as aural fullness, hypoacusia, tinnitus. In case of chronic elevation of the intracranial pressure, patient may describe pressure type headache, pulsatile tinnitus, balance problems & visual disturbances. If a CSF leak ensues orthostatic[9] or exertional headaches are frequent. Sudden sensory neural hearing loss are some times reported[11]. Recognition of the source of the leakage and its appropriate treatment are necessary to avoid rhinorrhoea or otorrhoea, low pressure headache and meningitis which are known complication of CSF leak[12]. Assessment of existence and extension of meningoencephalocele depends on computerised tomography(CT) and MRI[13]. Current imaging techniques include multidetector thin section CT with axial & coronal scans of the temporal bone[14]. It allows to assess the extent of bony resorption and helps in planning the reconstruction. However, in cases in which tegmen is thinned CT alone can be misleading because tissue averaging may hide an intact bony plate. Patients suspected of having a meningocele must then undergo an MRI of the brain in order to better define the pathology and differentiate among granulation tissue, cholesteatoma and brain hernia[15]. In this respect, T2 weighted coronal images clearly visualise brain and CSF entering the mastoid or middle ear[16]. Localization of the site of the CSF leak is a more challenging task and multiple imaging methods are required, such as digital subtraction myelography[17], intrathecal gadolinium MRI or MR cisternography[19].

of the presence of CSF in the middle ear requires a myringotomy and positive sampling for beta2 transferrin or the more specific trace protein[19,20]. Bony dehiscences in the lateral skull base are either congenital post traumatic or related with bone resorption caused by chronic infection of the adjacent structure i.e middle ear and mastoid to the otosurgical procedure performed to cure the pathology.

Numerous surgical approaches involving grafting the bony defects have been described and found to be highly successful for the management of tegmentympanidehiscence with CNS / ontological problems. Gubbles et al[21] reported the use of a fragment of calvarial bone to reconstruct the tegmen tympani with bone cement applied on the irregular surface and a muscle / fascia overlay to reconstruct the floor of middle cranial fossa.

Kenning et al[22] reported the use of autologous bone, pericranium and intradural collagen grafts. Seaman et al[23] suggested creating an epidural pocket with a cartilage locked in place & a fascia graft overlying the defect. Few reports have described surgical approaches for the treatment of small meningoceles without significant complications. As a consequence, it has been suggested that surgical intervention may not be required in asymptomatic patient.

II. Case Report

A 22 year old female had a H/O watery discharge from Rt ear since 6 months, insidious in onset, continuous, which was not blood stained, non foulsmelling, discharge was subsiding with medication. She had H/O trauma 6 months back. no H/O ear ache, tinnitus, vertigo, fullness of ear, itching sensation, hyperacusis. no H/O of nasal discharge, throat irritation, dysphagia, change of voice, no cerebellar abnormalities found.

Past history: history of Rt Csom With Attico Antral Disease 2 years back for which she was operated. Audiometric evaluation confirmed the presence of a mild conducting hearing loss.

Otoscopic examination revealed a cholesteatoma sac filling the attic, soft tissue bulge in Rt external auditory canal.

MRI of brain plain & contrast showed small defect in superior temporal bone with mild inferior herniation of brain parenchyma through the defect. hyperintensity on T2WI with in right mastoid air cells – s/o mastoiditis.

ct scan temporal bone plain & contrast showed post op. case of right mastoidectomy. Soft tissue attenuation with enhancement in right external auditory canal, middle ear and widening of external auditory canal, middle ear. Destruction of tympanic membrane, ossicular chain – cholesteatoma (? Recurrence). Erosion of tegmen tympani and communication with middle cranial fossa. Bony defect measuring 7.6*7.4mm.

She was posted for review mastoidectomy & CSF repair. Through post aural approach subcutaneous tissue, tympanomeatal flap was incised. soft tissue bulge seen occupying the external auditory canal. Skin over the soft tissue bulge peeled, using bipolar cautery arachnoid and brain parenchyma cauterised. A well was created in the outer cortex of the floor of middle cranial fossa. Dura was lifted from MCF. A preserved homologous septal cartilage of appropriate size passed through space created covering the bony defect all around and covered with temporalis fascia & tympanomeatal flap. Patient was followed for 6 months.

III. Discussion

The term encephalocele encompasses meningocele (herniation of meninges & CSF), encephalomeningocele (herniation of brain & meninges), hydroencephalomeningocele (herniation of brain, ventricle & meninges). There are varying degrees of herniation and the diagnosis may be obvious or quite difficult to establish. A mass extending from tegmen must be considered brain tissue until proven otherwise. On occasion, the mass may be quite large and can actually block the external auditory canal.

The temporal bone is formed by joining of 4 ossification centres squamous, tympanic, mastoid & petrous portion. These 4 centres begin ossification at different times and are not fully ossified at birth. Tegmen is formed by joining of superior portion of the petrous bone with the caudal squamous portion of the temporal bone[24]. Due to the nature of the remoulding, a small bony dehiscences frequently occur in the cortex of the tegmen & tegmen antri[25].

Etiology of skull base defects are

- 1) Iatrogenic – planned partial surgical removal of middle fossa dural plate either by transpetrous approach, subtemporal + transmastoid approach.
Accidental drilling during mastoidectomy
- 2) Neoplastic – glomus tumour, meningioma, middle ear carcinoma, metastasis.
- 3) Traumatic – temporal bone fractures, suppurative ear disease, temporal bone osteitis, invasive cholesteatoma, middle ear tuberculosis.
- 4) Others – Langerhans cell histiocytosis, temporal bone fibrous dysplasia, congenital dehiscence.

The pathogenic mechanism of production of a meningoencephalocele hernia through a bony defect in the skull base have been mainly related with increased intra abdominal / intra thoracic pressure, leading to increased central venous pressure and subsequent benign intracranial hypertension, a condition also known as pseudotumour cerebri. A long standing increase in intracranial pressure might produce a meningoencephalocele, as seen in obese patient[26].

A CSF leak is frequently associated with a meningoencephalocele. It occurs when direct communication of the subarachnoid space to the extracranial compartment, usually a paranasal sinus or the middle ear cavity, is established through an osseous and dural defects at the skull base

The clinical presentation of tegmental meningoencephalocele include conductive hearing loss, intermittent/ continuous CSF otorrhoea or rhinorrhoea, serous otitis media , headache, meningitis, temporal lobe seizures[27] ,expressive aphasia & facial nerve weakness[28].

diagnosis of tegmental meningoencephalocele is based on a high index of suspicion. Otologic examination can delineate chronic otitis media , cholesteatoma/ common illness. If CSF leakage is suspected , the fluid can be tested for glucose(limit of normal <60% of serum glucose),protein (limit of normal < 200mg/dl) and beta 2 transferrin levels[29]. If the index of suspicion is high, radiologic examination can usually provide the definitive diagnosis. HRCT of the tegmental plate , posterior and middle fossa & Eustachian tube provide the best evidence of a bony dehiscence[29]. Coronal section are most useful. However, CT poorly differentiates soft tissue swelling[30,31]. Pregadolinium and post gadolinium weighted MRI provide good differentiation of encephaloceles from cholesteatoma & inflammatory tissues[32].Surgical approach for meningoencephaloceleThese include a MCF craniotomy alone[33,34], a mini middle cranial fossa approach[35,36]. Transmastoidaooroach[37-39]& a combined MCF-transmastoid approach[39-41].

choice of surgical approach is driven by the etiology of the defect , by the site & extent of the bony defect and of thr brain hernia by the type & degree of hearing loss, the presence of chronic infection in the middle ear, by the intraoperative finding of active CSF leakage[42].

In general , the herniated tissue can be either resected or pressed back in to the intracranial compartment . if a middle cranial fossa approach is selected , the herniated cerebral tissue can be sectioned from above & left in middle ear. The remnants left in middle ear or Mastoid cavity progressively shrink , becoming part of scar tissue[43].

defect is sealed with autologous ,heterogenous or alloplastic material or their combination . each of these solution depends on surgeon experience, size of defect and volume of herniated brain. CSF drainage is warranted in large defects of the skull base or obese patients , bed rest , avoidance of straining and sneezing, fecalsoftners and cough sedation are sufficient preventive measures in all minor repair.

The Transmastoid Approach

tegmens plate defects causing CSF leaks and brain henias have conventionally been repaired otologists by using soft tissue grafts via a retroauriculartransmastoid approach. It is a simple , direct way to the lateral cranial base , with minimal morbidity , usually limited by the involvement of the ossicular chain in the attic , whose handling often leads only to a mild to moderate conductive deafness.

A simple or complete mastoidectomy with preservation of the outer ear canal wall is the preferred procedure .if a previous CWD mastoidectomy has been performed , its revision is mandatory. In case of chronic otitis media , eradication of the disease is enforced and a tympanoplasty with reconstruction of either the ear drum or ossicular chain is performed as needed. After the bone work is complete and the pathology is removed , the bony defect is exposed. The operative field is rinsed with antibiotic solution .the herniated tissue is generally small , even when dura is extensively unoveredand it is sufficient to coagulate the herniated tissue , reduce into the intracranial compartment. Cover the bony defect with bone dust & fascia. In rare instances, suturing the dura in an extradural fashion and cutting the redundant meningocele is required.

The bony dehiscence is the sealed by any of the material selected for reconstruction which must be trimmed and fitted precisely for the size & contour of the defect.

Technique for closure of the defect

1) Inlay technique 2)over lay technique 3) sandwich technique

Inlay Technique:the middle cranial fossa dura is smoothly detached , 3-4mm from the bony borders of the defect with a duck bill elevator. The final dimensions of the implant are slightly larger than the defect itself to permit limited overlapping.

A) If an alloplastic material is used , an autologous fascia temporalis graft is interposed in the intracranial compartment facing the dura

B) The implant is placed in the intracranial compartment between the dura and the petrous bone , thus held in place by gravity & physiological intracranial pressure.

Sandwich technique: further water tight sealing on the mastoidectomy side is achieved by means of another layer of temporal fascia, fibrin glue is added to inlay technique.

Overlay Tecnique: the graft or the implant is glued to the mastoid surface of the defect , leaving the intracranial compartment untouched. Unfortunately if not adequately supported by other filling material, the attached insert is prone to be dislodged into mastoidectomy under the CSF pressure.

Materials for reconstruction

They include autologous and homologous bone/ cartilage grafts , local pedicled soft tissue flaps or distant free vascularised flaps as well as many alloplastic material such as bony substitutes , cements , ceramics , metals , polymers , acrylic and their composites.

Organic tissue:

- 1) autologous bone grafts are oldest and preferred material to reconstruct any size of defect in petrous bone roof[44].
- 2) bone dust
- 3) cartilage – sufficient elastic but offer little resistance to deformity. They are not osteogenic and are often reabsorbed to variable extent or converted to fibroid tissue.

Alloplastic :metal mesh , ceramics fluoroplastic , methyl methacrylate either alone or in combination. Among ceramics hydroxyapatite is used extensively.

FIGURES

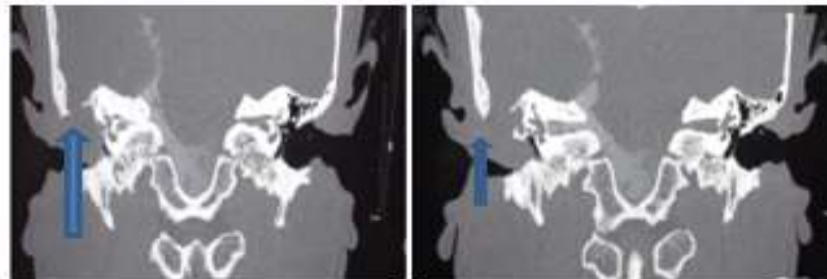


Figure 1

Figure 2

FIGURE 1 & 2 CT SCAN images (coronal view) of temporal bone showing the defect in tegmen.

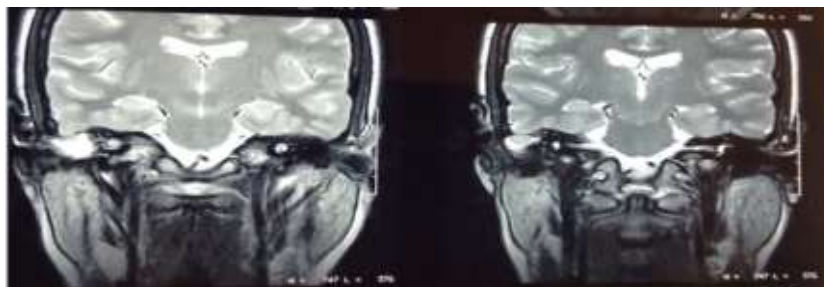
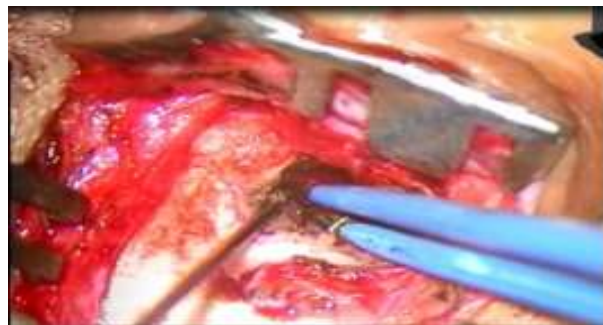


figure 3 { MRI showing the meningoencephalocele in mastoid cavity. }

A



B



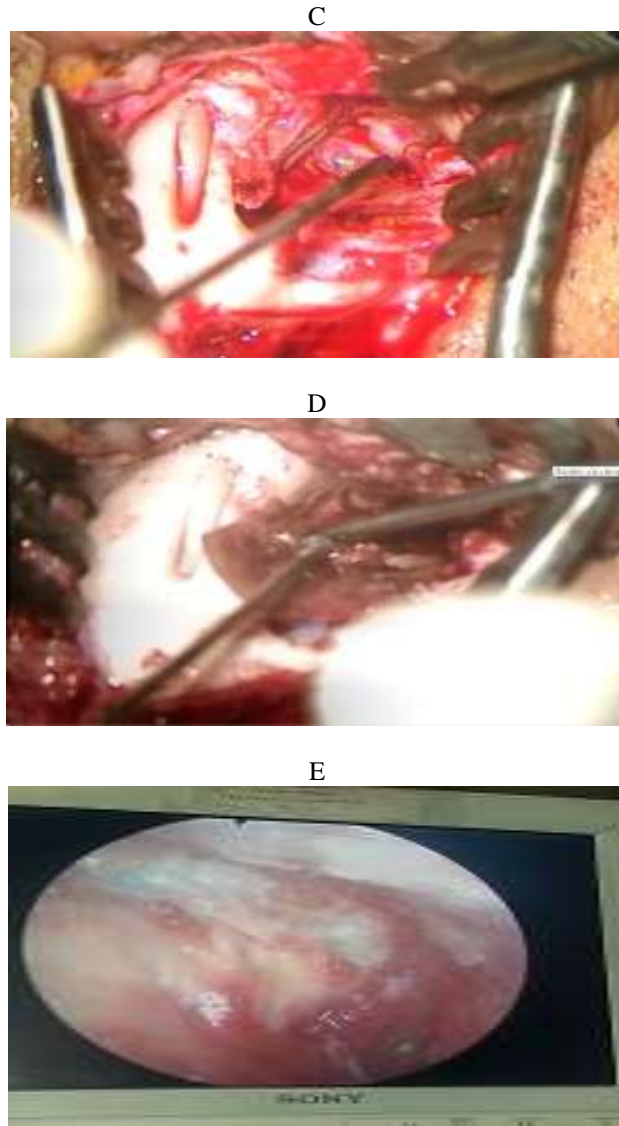


Figure (A to D) showing the surgical steps. Figure E – POST OP MASTOID CAVITY

Conclusion: Meningoencephalocele should be considered in the differential of a soft tissue mass in middle Ear. HRCT temporal bone will define the anatomy and MRI for soft tissue characterization. Cartilage / alloplastic implants provide rigid barrier to withstand the intracranial pressure. Never use mono polar cautery over the dura.

References

- [1]. Brown NE, Grundfast KM, Jabre A, Megerian CA, O'Malley BW jr, Rosenberg SI. Diagnosis and management of spontaneous cerebral fluid –middle ear effusion and otorrhea. *Laryngoscope* 2004;114:800-5
- [2]. KamererDB ,Caparosa RJ: temporal bone encephalocele- diagnosis and treatment. *Laryngoscope*1982; 92: 878-882.
- [3]. Glasscock ME 3rd, Dichinsjr , Jackson CG , Wiet RJ, Feenstra L : surgical management of brain tissue herniation into the middle ear and mastoid. *Laryngoscope* 1979;89:1743-1754.
- [4]. Gottlieb MB,Blaugrund JE, NiparkoJK : imaging quiz case 1. Tegmental encephalocele. *Arch otolaryngol head neck surgery* 1998; 124: 1274,1276-1277.
- [5]. Ba MC, KabreA ,Badiane SB, et al: fronto- ethmoidal encephaloceles in Dakar. Report of 9 cases. *Dakar med* 2003;48: 131-133.
- [6]. Lundy LB , graham MD, Kartush JM et al . temporal bone encephalocele and cerebrospinal fluid le[44aks. *Am J otol* 1996;17:461-9.
- [7]. Quiney RE, Mitchell DB, Djazeri B, et al .recurrent meningitis in children due to inner ear abnormalities. *J laryngolotol* 1989;103: 473-80.
- [8]. Schlosser RJ ,bolger WE , significance of empty sella in cererbrospinalfluid leaks. *Otolaryngol head neck surg* 2013; 128 : 32-38.
- [9]. Mokri b. spontaneous intracranial hypotension .*Currneurol neuro sci rep* 2001;1: 109-17.
- [10]. Mokri b, Maher CO ,sencakova D . spontaneous CSF lesks : underlying disorder of connective tiissue. *Neurology* 2002;58:814-16
- [11]. Gladstone JP ,doick DW . isolated sudden onst hearing loss as a manifestation of recurrent CSF leak. *Cephalgia*2005 : 25 : 550-53.
- [12]. SdanoMT ,pensak ML. temporal bone encephaloceles. *Curropinotolaryngol head necksurg* 2005; 13:287-89.
- [13]. Jackson CG , pappas DG jr, manolidis S , glasscock ME , von doersten PG , hampf CR , Williams JB, storper IS. Brain herniation into middle ear and mastoid; concepts in diagnosis and surgical management . *AM J otol* 1997; 18: 198-205.

- [14]. Obhay ,iguchi t , hirose y , takasu m , nishii N , maeda S , kitagawa H. computer tomography diagnosis of meningoencephalocele in a calf. *J vet med sci* 2008 ; 70: 829-31
- [15]. Johnson pl ,eckard DA , Chason DP , brecheisen MA , Batnitzsky S. imaging of acquired cerebral herniations . *neuro imaging clin N Am* 2002 ; 12: 217-28.
- [16]. BovoR ,ceruti S , padovani R , martini A . temporal bone herniation . *otol neurotol*2006 : 27 576 -77.
- [17]. HoxwarthJM ,patel AC , Bosch EP , nelson KD . localization of a rapid CSF leak with digital subtractionmyelography.*AJNR Am J neuroradiol* 2009 ;30: 516- 19.
- [18]. MRI with intrathecal gadolinium to detect a CSF leak: A prospective open label cohort study.*Jneuro/neurosurg psychiatry* 2010.
- [19]. Intra thecal gadolinium – enhanced MR cisternography in the evaluation of CSF leakage. *AJNR Am J neuroradiol* 2010; 31: 71-75.
- [20]. Preliminary study on the stability of beta -2 transferrin in extracorporeal cerebro spinal fluid .*otolaryngol head neck surg* 2011: 144 :101-3.
- [21]. Gubbels SP, Selden NR,Delashaw JB jr , Mc menomey SO. Spontaneous middle fossa encephalocele and cerebrospinal fluid leakage: diagnosis and management.*otolneurotol* 2007;28:1131-9.
- [22]. Kenning TJ ,Willcox TO, Artz GJ , Schiffmacher P, Farrel CJ ,evans JJ . surgical management of temporal meningoencephaloceles, cerebrospinal leaks and intracranial hypertension: treatment paradigm and outcomes. *Neurosug focus* 2012;32:e6.
- [23]. Seaman MT, GilpinDA ,HsuDP, WasmanJK, MegerianCA. Transmastoid extradural- intracranial approach for repair of transtemporalmeningoencephalocele: a review of 31 consecutive. *Laryngoscope* 2011;121:1765-72.
- [24]. Proctor B ,Neisen E, proctor C: petrosquamous suture and lamina . *otolaryngol head neck surg* 1981;89:482-495.
- [25]. Lang Dv: macroscopic bony deficiency of the tegmentympani in adult temporal bones. *J laryngolotol* 1983;97:685-688.
- [26]. Scurry WC jr, ort SA, Peterson WM, Sheehan JM, Isaacson JE. Idiopathic temporal bone encephaloceles in the obese patient. *Otolaryngol head neck surg* 2007: 136: 961-65.
- [27]. Hyson M ,Andermann F, Olivier A, Melanson D: occult encephaloceles and temporal bone epilepsy: developmental and acquired lesions in the middle fossa. *Neurology* 1984;34:363-366.
- [28]. Uri N, Shupak A, Greenberg E, Kelner J ; congenital middle ear encephalocele initially seen with facial paresis. *head neck* 1991;13:62-67.
- [29]. Patel RB, Kwartler JA , hodosh RM , Baredes S: spontaneous cerebrospinal fluid leakage and middle ear encephalocele in seven patients . *ear nose throat J* 2000: 79 : 372- 37
- [30]. LaRouere MJ, NiparkoJK ,Gebarski SS, Kemink JL: three dimensional x ray computed tomography of the temporal bone as an aid to surgical planning. *Otolaryngol head neck surg* 1990;103: 740-747.
- [31]. KamererDB ,caparosa RJ : temporal bone encephalocele- diagnosis and treatment . *laryngoscope* 1982 ;92:872-882.
- [32]. Martin N, sterkers O, Nahum H : chronic inflammatory disease of the middle ear cavities: Gd-DTPA enhanced MR imaging . *Radiology* 1990 ; 176: 399-405.
- [33]. Helms J , Geyer G . alloplastic materials in skull base reconstruction . in :Sekhar LN , JaneckaJp (eds) surgery of cranial base tumours: a colour atlas . newyork, raven Pr, USA 1993.
- [34]. PaparellaMM,MeyerhoffWF , Oliviera CA. mastoiditis and brain hernia (mastoiditis cerebri). *Laryngoscope*1978;88:1097-106.
- [35]. Johnson GD, Jackson CG , Fisher J , mastarSA, Poa DS. Management of large duralsefts in skull base surgery: an uptade .*laryngoscope* 1990; 100:200-02.
- [36]. Cheney ML ,Megerian CA , Brown MT, Mc kenna MJ, NadolJB. The use of the temporoparietal fascial flap in temporal bone reconstruction.*Am J otol* 1996;1742.
- [37]. RamdsenRT , Latif A , Iye RH, Dutton JE. Endaural cerebral hernia. *J laryngol* 1985;99643-51.
- [38]. Piattelli A , Franco M , Ferronato G, santello MT , Martinetti R , ScaranoA. A resorption of composite polymerthyroxyapatite membranes: a time – course study in the rabbit. *Biomaterials* 1977; 3: 629 -33.
- [39]. Jones NF ,sekhar H, Schramm VL . free rectus abdominis muscle flap reconstruction of the middle and posterior cranial base. *Plastreconstrsurg*1986 ; 78 : 471 -77.
- [40]. BoldreyE .stainless steel wire mesh in the repair of small cranial defects. *Ann surg*1945 : 121: 821-25.
- [41]. Van hasseltCA , Liu KC , Tong MC. The hongkong vascularised temporalis fascia flaps for optimal mastoid cavity reconstruction. *Rev larngolotolrhinol(bord)*1995; 116:57-60.
- [42]. SannaM ,Fois P , Russo A , Falcioni M, management of meningoencephalocephalic herniation of the temporal bone: personel experience and literature review. *Laryngoscope* 2009; 119: 1579-85.
- [43]. Arstegui M ,Falcioni M < Saleh E, tailbah A, Russo A, Landolfi M ,Sanna M. meningoencephalic herniation into the middle ear: A report of 27 cases . *laryngoscope* 1995 ;105:512- 18.
- [44]. KiyokawaK , Hayakawa K , Tanabe HY , Inoue Y , Tai Y, Shigemori M, tokutomi T. Cranioplasty with split lateral skull plate segments for reconstruction of skull defects. *J craniomaxillafacsurg* 1998; 26 : 379-85.