Temporal Bone encephalocele

- Encephaloceles are the presence of meninges, cerebrospinal fluid (CSF), and/or brain extending beyond the cranial cavity through a bony defect in the skull.

- Formerly called brain fungus

- an encephalocele commonly involves portions of brain tissue that have become necrotic after herniation.

- The term encephalocele encompasses meningocele (herniation of meninges and CSF), encephalomeningocele (herniation of brain and meninges), and hydroencephalomeningocele (brain, ventricles, and meninges).

- The incidence of congenital encephaloceles is estimated at 1:3000 to 10 000 live births, favoring females 2.3:1.

- Occipital encephaloceles are most common, with cranial base lesions accounting for only 5% of encephaloceles.

- The most common cranial base encephaloceles involve the temporal bone and middle fossa.

- Rarely, temporal bone encephaloceles arise from the posterior cranial fossa.

Pathogenesis of temporal bone encephaloceles:

- spontaneous, iatrogenic, inflammatory, and traumatic.

- spontaneous encephaloceles are infrequently encountered, usually associated with a CSF leak and can be divided into congenital and idiopathic.

@ Congenital encephaloceles are best explained through understanding the development of the temporal bone.

- The temporal bone is formed by the joining of 4 ossification centers: the squamous, tympanic, mastoid, and petrous portions.

- These 4 centers begin ossification at different times and are not fully ossified at birth. Tegmen: joining of the superior portion of the petrous bone with the caudal squamous portion of the temporal bone.

- Fusion of this petrosquamous suture is usually complete by 1 year of age, but can be delayed by growth abnormalities, chemotherapy, or radiation therapy, leaving a potential space for the extension of an encephalocele.
The pathogenesis of idiopathic spontaneous encephaloceles remains enigmatic.

- Small sites of herniation through natural defects in the tegmen of the temporal bone (20%-34% of temporal bones have multiple small dehiscences)
- Aberrant arachnoid granulations slowly eroding the temporal bone through years of CSF pulsations.
- Inflammation, aging, and increased intracranial pressure may play a role in tegmental and/or dural attenuation.
- Dural weakening is necessary for transmission of an encephalocele, which would explain why although small bony dehiscences occur in up to 34% of the population, tegmental encephalocele remains a rare occurrence.

Most commonly, tegmental encephaloceles are iatrogenic, often seen as adverse sequelae of mastoid surgery for chronic ear infection.

- This condition typically occurs as a result of drill-induced defects of the tegmen often overlying the antrum and/or epitympanum.
- Revision mastoid surgery ➔ a higher incidence of tegmen defect
- Although dural exposure is a common occurrence, an intact dura mater can support the brain over large bony defects.
- A dural weakening as small as 2 mm at the site of the bony defect is suggested as necessary for encephalocele formation.
- Encephaloceles: preventable through careful surgical technique and appropriate tegmental and/or dural repair.

Temporal bone encephaloceles can arise secondary to chronic otitis media.

- Chronic inflammation and enzymatic degradation of tissues ➔ bone resorption and dural dehiscence.
- Tegmental erosion due to cholesteatomas frequently leaves the dura exposed and weakened as a result of concomitant infection.

Trauma to the temporal bone is the fourth mechanism of tegmental encephalocele formation.

- The dura is so tightly adherent at the skull base, any fracture of the temporal bone ➔ tegmental deficiency or a dural weakening.
- This type of injury is typically associated with CSF otorhinorrhea, resolve within 2 weeks.
Clinical presentation of tegmental encephalocele

* conductive hearing loss, intermittent or continuous CSF otorrhea and/or rhinorrhea,
* CSF masquerading as serous otitis media (most common presentation), headache, meningitis (often recurrent), and sometimes as a bluish gray mass behind the tympanic membrane, temporal lobe seizures, expressive aphasia, and facial nerve weakness.
* The differential diagnosis includes cholesteatoma, chronic otitis media, postsurgical granulation tissue, cholesterol granuloma, and serous mastoiditis.

Diagnosis of tegmental encephalocele

* Based on a high index of suspicion.
* Otologic examination can delineate serous otitis media, a common illness, and cholesteatoma.
* If a CSF leak is suspected, the fluid can be tested for glucose (limit of normal: <60% of serum glucose), protein (limit of normal: <200 mg/ml), and beta-2-transferrin levels (only present in CSF).
* High-resolution CT: bony dehiscence. Coronal section.
* A CT scan: poorly in differentiating soft tissue swellings.
* Pregadolinium and postgadolinium T1-weighted magnetic resonance imaging (MRI) scans provide wonderful differentiation of encephaloceles from cholesteatomas and inflammatory tissue.

* Sagittal MRI with pregadolinium and postgadolinium sequences show encephaloceles to be isointense compared with brain,

* postcontrast images: cholesteatomas (hypointense) and granulation tissue (hyperintense). Isolated tympanic glomera chemodectomas (hyperintense)

* facial neuromas, intratympanic meningiomas, and small metastases, are localized and well-delineated on CT scans, similarly on postgadolinium MRI

Treatment:

* Prolapsed brain tissue: non-functional, result strangulation, ischemia and resultant edema ➔ safely excised
Small amount of encephalocele: replace, except infection

Approach:
- Posterior and lateral defect (tegmen mastoideum), small defect < 1 cm → Transmastoid approach:
- Medial and anterior defect (tegmen tympani), Large defect > 2 cm, multiple defect → Middle cranial fossa approach

Combination:
- Dura repair: temporalis fascia, pericranium and fascia lata
- Tegmen repair: bone, cartilage graft
- Alloplastic material: Surgical silastic sheeting, oxycel cotton
- Autograft: Pedicled or free muscle flap
- Small defect: abdominal fat
- Small to moderate defect: pedicle of temporalis muscle
- Defect > 1 cm: place bone or cartilage
- Multilayered closure: high successful rate

REFERENCES

8. Glasscock ME, Dickens JRE, Jackson CG. Surgical management of brain tissue herniation into the middle ear and mastoid. Laryngoscope. 1979;89:1743-1754.