Encephaloceles, Meningoceles, and Cranial Dermal Sinus Tracts
nosology

- cephalocele: herniation of any of the intracranial contents through a defect in the skull
- Encephalocele: contains either normal brain or gliotic tissue
- Meningocele: contains only cerebrospinal fluid (CSF) surrounded by a capsule of arachnoid tissue
- Encephalocystocele: contains ventricle
Encephalocele classification

- Encephalocele can be congenital or acquired due to trauma, tumor, inflammation, infection, or previous surgical procedure.
- Other classification based on the location of the defect anterior and posterior.
- Anterior divided into two groups: sincipital (or frontoethmoidal) when the herniation is anterior to the cribriform plate, and “basal,” when the herniation is through the sphenoid sinus.
Embyrology

- The exact error in development leading to the occurrence of occipital and sincipital encephaloceles is not entirely known.
- As encephaloceles are covered in skin, a defect in neurulation alone cannot completely explain the etiology, as a problem with neurulation would necessitate the absence of skin coverage.
- McClone show that the membranous bones of the skull and the three meningeal layers originate from a mesoderm that is locate between the closed neural tube and superficial squamous ectoderm.
In normal embryogenesis, a diverticula of dura normally projects anteriorly through a small fontanelle between the developing nasal and frontal bones (fonticulus nasofrontalis)
• Ossification of the sphenoid bone, which occurs during the first year of life. A developmental insult here could lead to nonossified membranous sphenoid ala allowing for protrusion of temporal lobe.

• Incomplete development of the medial sphenoid bone can lead to the persistent development of a lateral craniopharyngeal canal, known as Sternberg’s canal, which can act as gateway for encephaloceles to protrude into the lateral sphenoid sinus.
Epidemiology

• The incidence of encephaloceles and other neural tube defects is declining because of using folic acid and improve diagnose method.

• Over 80% of encephaloceles in the Western Hemisphere are occipital with most cases of frontoethmoidal and basal encephaloceles occurring in Southeast Asia and parts of Russia.

• Using a lot of tea and pesticide can be a reason of encephalocel.
• **Genetics**
  • High rate in female and association with some syndromic disorder

• **DIAGNOSIS**
  • maternal serum AFP
  • prenatal ultrasound
  • Amino synthesis
• in patient with nasal obstruction, obligate oral breathing, snoring, potential CSF rhinorrhea and meningitis, and purulent nasal discharge we should rule out encephalocell

• Frontonasal and frontobasal encephaloceles can also contain anatomic portions of the hypothalamus, optic apparatus and anterior cerebral arteries

• MANAGEMENT

• Mode of Delivery
Encephaloceles and Hydrocephalus

- Hydrocephalus has been reported in both posterior and anterior encephaloceles.
- In most cases, it is usually recommended to treat the hydrocephalus first.
Anesthetic Considerations

• ask anesthesiology team to perform a preoperative assessment of these patients, including an endoscopic evaluation to work out the airway anatomy

• Goals of Surgery

• The principles of surgical treatment of encephaloceles are reduction of the herniation with preservation of as much viable brain as possible, water-tight dural closure with adequate skin coverage, repair of any cosmetic deformity, and cranial or craniofacial reconstruction
Surgical Technique

Posterior Encephaloceles

• The encephalocele is grasped and raised with tissue forceps, and an elliptical incision is made around the sack close to the neck of the herniation.
Anterior Cephaloceles

• anterior encephaloceles often involves a multidisciplinary approach including OMFS, ENT and plastic surgery

• transcranial approach typically involves a bicoronal scalp flap, a bifrontal craniotomy, and an extradural dissection of the anterior cranial fossa to separate the sac from the surrounding tissue and skull base. Next, an intradural transection of the encephalocele stalk is performed then OMFS/ENT team to come from below
Prognosis

• The location and eloquence of the brain tissue comprising the encephalocele are also important prognostic factors.

• Ant encephalocele have better prognosis than post

• The coexistence of hydrocephalus and associated intracranial and extracranial anomalies with any type of encephalocele will also portend a poorer prognosis
CRANIAL MENINGOCELES

• Cranial meningoceles, by definition, are congenital herniations of meninges and CSF through the skull and are bereft of any cerebral tissue

• A common location for these lesions is directly over the anterior fontanelle

• These lesions commonly abut the sagittal sinus, and dissection in this area has to be done carefully

• These lesions can be mass effect, and source recurrent meningitis at the skull base and epileptogenic focus in the temporal lobe
CRANIAL DERMAL SINUS TRACTS

• In the presence of an unexplained meningitis, especially due to Staph aureus hemolyticus, the entire skin surface along the midline should be examined carefully for dimples, palpable thickening, or sinuopenings. This search should include shaving the head.
Embryology

- Cranial dermal sinus tracts are uncommon, potentially deleterious, congenital, epithelialized tunnels that originate at the skin surface and can occur anywhere along the cranial bones.
- The most common sites are at the occipital protuberance or nasion.
- Result from failure of dysjunction between neuroectoderm and cutaneous ectoderm.
Epidemiology

- Cranial dermal sinuses are uncommon dysraphic lesions with slightly more than 100 cases published as single reports or small series.
- Cranial DSTs have also been reported in non-midline locations in the head, including lateral, parietal, anterior temporal, lateral temporal (pterional), and retroauricular.

Genetics

- Usually sporadic but female predominance and be with Klippel-Feil anomaly show genetic basis.
Diagnosis

- Cranial dermal sinus tracts commonly appear as a small, asymptomatic cutaneous dimple or pit.
- They can also appear as fulminant meningitis or acute hydrocephalus or stridor and difficulty feeding in nasal DST.
- In an Iranian series of 18 patients with cranial DSTs, infection was the most common reason for referral (50%), followed by an asymptomatic skin dimple (33%), with only one patient (5.6%) exhibiting signs and symptoms of raised ICP due to mass or abscess.
Association with Dermoid and Epidermoid Cysts and Intracranial Abscesses

- Dermoid and epidermoid cysts can be found at any depth along the tract in up to 50% of cranial dermal sinuses.
- These lesions can be sources of mass effect or abscess or can rupture spontaneously and cause of Aseptic meningitis.
Management

• For asymptomatic cranial DSTs, most authors agree that prophylactic surgical removal of the tract and any associated dermoid or epidermoid cyst is warranted “as soon as the child can tolerate the operation

  • Goals of Surgery

  • Cranial DSTs should be removed entirely to eliminate the risk of meningitis and abscess formation
  • In the presence of CSF leak, infection, or raised intracranial pressure, surgery should be performed emergently
Prognosis

• When completely resected, cranial DSTs rarely recur, and the prognosis is usually favorable.

• Recurrence following partial removal has been documented as late as 20 years from the original operation.