Dermoid, Epidermoid, Cysts, Lipoma

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NEURENTERIC CYST

- benign slow-growing cysts
  - location (intra and extra axial):
    A. Most commonly - spinal canal (particularly in low cervical or high thoracic region), can be found both ventral and dorsal to spinal cord, as well as in intramedullary location
    B. Intracranial (most often posterior fossa, pontomedullary region, cerebellopontine angle, parasellar area, and cranio-nasal junction)
  - N.B. N.B. most VENTRAL and EXTRA-AXIAL
  - cyst wall is composed of well-differentiated cuboidal, columnar, or ciliated epithelium, with or without goblet cells or mucrovi; mucin-containing cells show positive reaction with periodic acid-Schiff stain and are seen in 5% to 80% of specimens
  - epithelium may resemble intestinal or respiratory type, and there may be smooth muscle in underlying fibrovascular tissue.
  - cyst fluid - clear and colorless, milky, yellowish, gelatinous, xanthochromic, and blackish and viscous.
  - immunohistochemical staining - positive for cytokeratin and epithelial membrane antigen and negative for vimentin, glial fibrillary acidic protein, and S-100
  - cells also often test positive for carcinoembryonic antigen (CEA)
  - differentiate between neurenteric* and ependymal cysts.
  - presence of underlying basement membrane or collagenous tissue separating epithelial layer from neural tissue + lack of immunoreactivity to GFAP, S-100, neuron-specific enolase, and vimentin
  - malignant transformation is rare.

CLINICAL FINDINGS

- slow and insidious compression of adjacent spinal cord
  - persistent fistulas (between enteric structures and these cysts within CNS) — recurrent aseptic meningitis.

IMAGING

- MRI - variable and heterogeneous cyst signal (often parallel signal intensity of CSF), no enhancement (some variable wall enhancement may be present)
  - reports of TI signal changes in these tumors, resulting from changes in protein concentration or hemorrhage
  - cysts have tendency to insinuate themselves between surrounding structures.

Dermoid, Epidermoid, Tumor

A. T1-MRI - hypointense suprasellar mass
B. Hypointense midline suprasellar lesion
C. T1-MRI - cystic lesion
D. Contrast-enhanced T1-MRI - rim of cyst wall enhancement and region of intracyst enhancement.
E. T2-MRI - hypointense signal of cyst contents that follows signal change in cerebrospinal fluid.
F. FLAIR-MRI - increased intensity in cyst and some parenchymal hyperintensity around tumor, consistent with edema.
G. Protein density axial MRI - cyst contents somewhat heterogeneous.
H. Gradient echo axial MRI - cyst heterogeneity.

Pathology

- N.B. m.
- Neurenteric canal - normal embryologic transitory communication between neural tube (ectoderm), notochordal canal, and gut endoderm; canal persistence — paravertebral neurenteric cyst(s) or fistula(s).

- primary disruption of tissues derived from one or more of three germ cell layers:
  - PHARYNGOPHARYNX
  - digestive tract
  - neural tube
- frequently associated with mesodermal malformations (esp. vertebrae - hemivertebrae, absent or fused vertebrae, butterfly vertebrae, midline bony spurs).
- can manifest in patients of any age.
- clinically onset is usually insidious; occasionally cyst rupture and spillage of irritative contents into subarachnoid space — acute meningitic complaints or increased ICP.
- treatment of each of these entities is primarily surgical; radiotherapy and chemotherapy have little to offer.

GASTRULATION

- development of enteric structures and these cysts within subarachnoid space — acute meningitic complaints or increased ICP.
- treatment of each of these entities is primarily surgical; radiotherapy and chemotherapy have little to offer.

H, Gradient echo axial MRI - cyst heterogeneity.

Neurenteric cysts are almost pathognomonically associated with vertebral anomalies or diastematomyelia.
DERMOID, EPIDERMOID, CYSTS, LIPOMA

TREATMENT

- primarily surgical, complete resection (not always feasible given difficulty of completely removing cyst wall at locations where it adheres to adjacent structures)
- simple cyst aspiration, cyst wall marsupialization, or creation of cyst-subarachnoid shunt*, are considered in difficult cases.
- aseptic meningitis has not been problem one might expect.
- recurrence is possible even after gross total resection → reoperation.
- radiotherapy and chemotherapy have little to offer; whether there may be some role for radiosurgery in treatment of residual neurenteric cysts remains to be seen.

DERMOID tumor, EPIDERMOID tumor
- benign inclusion cysts (not true neoplasms!) composed of ectodermal elements.

ETIOLOGY
- congenital (embryonic remnants) - inclusion of ectodermal epithelial elements:
  a) during 3-5th embryonic weeks when neural tube closes at midline → midline tumors (esp. DERMIDS)
  b) during formation of secondary cerebral vesicles → lateral tumors (esp. EPIDERMOIDS).
- N.B. EPIDERMOIDS also may be acquired – due to trauma, frequently from lumbar puncture (epithelial cells deposited within spinal canal, mostly historical when spinal needle didn’t have stylet).

PATHOLOGY
- similar appearances and developmental origins;
  - both contain stratified squamous epithelium found in skin;
  - centrally, both contain desquamated epithelial keratin and some lipid material (cyst fluid may contain cholesterol crystals).
  - external surface is smooth, lobulated, EPIDERMOIDS have pearly appearance (“pearly tumors” or “keratin pearls” in wall) due to capsule of stratified squamous epithelium.
  - histo-cell nuclei among keratin (vs. pure keratin s. “wet keratin” in craniopharyngioma)
  - histologically, EPIDERMIDS have outer connective tissue capsule and are lined with stratified squamous epithelium (i.e. composed of ectodermal remnants).
  - DERMOIDS have outer connective tissue capsule and are lined with stratified squamous epithelium, which also contains hair follicles, sebaceous glands, and sweat glands (i.e. composed of ectodermal and mesodermal remnants).

- expand slowly over many years due to central accumulation of epithelial debris and glandular secretions – predictable linear growth due to accumulation of keratin (vs. tumors – grow exponentially due to cell multiplication).
- rarely calcify, CALCIMORS frequently calcify; CYSTOMORS calcify uncommonly (but when it occurs, it is feature that helps in distinguishing from arachnoid cysts).
- malignant transformation is rare.

LOCATION
- sites of epithelial deposition can occur anywhere between neural tube and overlying skin surface (depending on stage of intrauterine development at which they arise, they can lie within ventricular system, brain parenchyma, subarachnoid space, bones of skull, or even extracranially).
**DERMOID** (intracranial) - most commonly midline: 2/3 in posterior fossa (extradural, vermian, or within 4th ventricle); also suprasellar region, subfrontal areas, other sites - scalp (commonest location in childhood), skull, orbit, nasal, oral cavity, neck.

**DERMOID** (spinal) - most commonly near thoracolumbar junction, tends to involve conus medullaris and cauda equina.

- Intradural = 50% 
- Intradural extramedullary = 50% 
- Extradural = least common

Dermoids should be considered whenever lumbar puncture yields fat in CSF.

**EPIDERMOID** - most commonly lateral near cerebellopontine angle: may also occur in suprasellar and parasellar regions, choroidal, sylvian, and interhemispheric fissures, intraventricular, intradiploic (in cranial bones), inside spinal cord; intracerebral epidermoid is very rare.

**EPIDERMOID CLINICAL FEATURES**

**EPIDERMOID** - uncommon (≈ 0.3% of all brain tumors).

**EPIDERMOID** - 4-10 times more frequent than dermoid (≈ 2% of all intracranial tumors).

**EPIDERMOID DIAGNOSIS**

Absence of edema is characteristic!

Plain radiographs - local bone expansion or erosion, lytic lesions with thin sclerotic margin.

CT - well-circumscribed, unilocular cystic mass; calcifications in tumor wall.

- Contrast enhancement is uncommon! (EPIDERMOID wall may sometimes enhance).

**DERMOID** - gives very low density (may be slightly heterogeneous due to additional lipid-depleted elements - hair follicles, sebaceous glands, sweat glands).

- Fat-fluid level in ventricles or fat droplets in subarachnoid spaces strongly suggest dERMoid rupture.

**MRI**

- **DERMOID** - characteristics similar to fat (glandular secretions) - midline mass hyperintense on both T1 and T2 – unique tumor!
  - Chemical-shift artifact is often present on T2 images as markedly hypointense band posterior at fat-fluid interface.

- **EPIDERMOID** - characteristics similar to CSF - variably hypointense on T1 and variably hyperintense on T2 – mimics arachnoid cyst (H: DWI – epidermoid has diffusion restriction).

**DERMOID** cyst has diffusion restriction (bright on DWI, dark on ADC) vs. arachnoid cyst (normal DWI).

Abscess, stroke, and lymphoma (high cellularity) also have diffusion restriction vs. gliomas and metastases do not restrict diffusion!

**ANGIOGRAPHY** - avascular mass.

Prenatal diagnosis with ultrasound (and resection shortly after birth) are now possible.

Suprasellar dermoid:

A) contrast T1 - high-signal-intensity suprasellar mass extending along planum sphenoidale.

B) contrast T1 with fat saturation - small amount of enhancement along peripheral aspects of lesion (arrows); majority of mass suppresses with fat saturation.

**ANGIOGRAPHY**

- Same dermoid (T1) - hypointense lesion; crescentic posterior rim of hyperintensity represents fat chemical-shift artifact.

- Same dermoid (T2) - hypointense cystic component in lesion.

- DERMoid (nonenhanced CT) - well-circumscribed, cystic; low-attenuating lesion at midline in suprasellar region, posterior to 3rd ventricle; small focus of calcification is noted at posterior margin of tumor.

- DERMoid (enhanced CT) - well-circumscribed, nodular focus of enhancement in right side of suprasellar lesion.
**DERMOID, EPIDERMOID, CYSTS, LIPOMA**

**EPIDERMOID CYSTS** (from WHO manual)
- benign exostic inclusions of epithelial cells during gastrulation (i.e. malformations of surface ectoderm) → secondary disruption of neural tube closure (i.e. dysraphism is secondary).

**PATHOLOGY**
- Location: tend to be off midline.
  - SPINE: intradural extramedullary (rarely intramedullary)
  - CRANIAL: 0.2% to 1.8% of all intracranial tumors
    - a) intradural (usually extra-axial) - cerebellopontine angle (may extend into Meckel's cave) or paraspinal cisterns
    - b) extradural (usually arising in diploic space of calvaria)
- thin capsule of stratified, keratinized squamous epithelium.
- cyst contains accumulation of desquamated epithelial cells, keratin, and cholesterol (characteristic pearly flakes).
- malignant transformation is rare, but SQUAMOUS CELL CARCINOMA has been reported (reported 15 cases with leptomeningeal dissemination of squamous cell carcinoma); MALIGNANT MELANOMA has also been reported in temporal lobe epidermoid.
- immunohistochemical staining positive for carbohydrate antigen CA19-9* (also detected in serum - can be used to evaluate for tumor recurrence or progression).

**TREATMENT**
- complete surgical excision is curative.
  - epidermoids do not invade but interdigitate around vital structures - complicating surgical removal.
  - avoid spilling of contents → chemical meningitis (15%); prophylaxis - irrigation with dexamethasone solution intraop + 2 weeks of oral dexamethasone (↑↑↑risk of infection).
  - associated dermal sinus should be removed completely.
  - chemotherapy and radiotherapy are not useful.

**DESMOID (nonenhanced CT)** - large, well-circumscribed low-attenuating cystic lesion in right temporal lobe lateral to cranial midline; peripheral marginal calcification; no erosion in adjacent bone of sella.

**EPIDERMOID (T1 with contrast)** - suprasellar, prepontine, and interpeduncular location of nonenhancing tumor (signal intensity similar to CSF): left Sylvian fissure is filled by mass which extends into chiasmatic cistern and encases left internal carotid artery termination (arrowhead); signal is similar to CSF on T2, but slightly higher than CSF on T1 (white arrow).

**EPIDERMOID (A- T2-weighted; B- T1-weighted MRI):** left abducens nerve is seen to be hyperintense to cerebrospinal fluid.

**TREATMENT**
- complete surgical excision is curative.
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  - avoid spilling of contents → chemical meningitis (15%); prophylaxis - irrigation with dexamethasone solution intraop + 2 weeks of oral dexamethasone (↑↑↑risk of infection).
  - associated dermal sinus should be removed completely.

Chemotherapy and radiotherapy are not useful.

**DESMOID (contrast CT)** - partial marginal enhancement; attenuation degree in center of lesion consistent with fat.

**EPIDERMOID (T1)** - hyperintense signal in lesion; multiple small hyperintense foci along sulci of right temporal lobe (represent fat droplets in subarachnoid space from focal dermoid rupture):

**EPIDERMOID (T1 with contrast)** - hyperintense lesion (hyperintensity is due to short T1 of fat); multiple hyperintense foci (fat droplets) in subarachnoid spaces; mild midline septal shift to left; chemical-shift artifact at superior marginal surface of lesion.

**EPIDERMOID:**
A. T2-MRI – large homogeneous mass, which is slightly higher in signal than CSF, fills right cerebellopontine angle
B. T1-MRI – lesion is again noted to be hyperintense to cerebrospinal fluid.

**Cystic Medullary Chordoma** (from WHO manual)
- Usually presents as a large, smooth-surfaced, well-encapsulated mass in midline location, often with associated bone destruction.
- On MRI, characteristically shows high signal intensity on T2-weighted images and variable signal on T1-weighted images.
- Associated with a high risk of local recurrence and distant metastasis.

**MALIGNANT MELANOMA**
- Radiation therapy may be indicated for localized tumors.
- Vascular invasion, necrosis, and lymph node involvement are common.
- Treatment options include surgery, chemotherapy, and immunotherapy.

**MALIGNANT MELANOMA** (from WHO manual)
- May present as a pigmented skin lesion.
- Risk factors include fair skin, blue eyes, red hair, and family history of melanoma.
- Early detection and treatment are crucial for improving survival.

**IMMUNOCHEMICAL**
- Staining for S-100 protein and HMB-45 can be helpful in distinguishing melanomas from other tumors.
- Positive staining for NSE, CD56, and CD57 can also be observed.

**MALIGNANT MELANOMA**
- Radical excision with wide margins is the treatment of choice.
- Adjuvant therapy, including chemotherapy and immunotherapy, may be used in select cases.
- Prognosis is poor, with a high rate of recurrence and distant metastasis.
**CLINICAL FINDINGS**
- reported from infancy to adulthood + incidental findings at autopsy.
- average age at detection is 35±4 years.
- female preponderance
- grow linearly (similar to normal skin) → insidious onset (median duration of symptoms 4-14 years).
- picture of acute meningitis may indicate epidermoid cyst rupture.
- spinal epidemoid tumors are frequently associated with vertebral anomalies.

**EXTRADURAL LESIONS** - manifest as local mass.
**INTRAHDURAL TUMORS** - headache (because of common parasellar location), visual disturbance, and to lesser extent, hypothalamic alterations.
- tumors in middle fossa grow quite insidiously and are often asymptomatic.
- tumors in cerebellopontine angle may cause ataxia, dizziness, or cranial nerve deficits.

**DIAGNOSIS**
**CRANIAL XR** - for intradiploic tumors - typically in cranial vault (may also occur in orbital region).
- lytic erosion of skull, sharply delineated sclerotic edge and scalloped margins on plain radiographs.
- inner table of skull is usually destroyed and outer table thinned.

**CT** - homogenous nonenhancing hypodense lesion in subarachnoid space without surrounding edema.
- occasionally, high density masses (“white epidermoids”) - difficult diagnosis.

**DIFFERENTIAL DIAGNOSIS**
- arachnoid cyst, Rathke's cleft cyst, craniopharyngioma.
- differentiation from ARACHNOID CYST:
  1) diffusion restriction on DWI (vs. arachnoid cysts – no diffusion restriction)
  2) more fat density than CSF density
  3) extend into subarachnoid space and enlarge it (vs. arachnoid cysts cause more focal mass effect).

**MRI** - hypointense to hypertense, heterogeneous, multiloculated.
- T1 hypointensity + T2 hyperintensity
- some tumors show rim enhancement
- no surrounding edema
- hydrocephalus is rare
- DW imaging is superior to other types of MRI sequences in delineating borders of epidermoid cysts.

A. Contrast-enhanced, T1-MRI - hypointense mass without enhancing components in midline posterior fossa. The mass is compressing brainstem and obstructing egress of cerebrospinal fluid from fourth ventricle, thereby causing hydrocephalus.
B. More lateral extent of this tumor.
C. Mass in situating between normal posterior fossa structures and extending to involve both right cerebellopontine angle and midline structures.
D. Contrast-enhanced, FLAIR MRI - marked heterogeneous within cyst and discontinuous rim of cyst wall enhancement.

**TREATMENT**
- symptomatic patients benefit from surgery (technically difficult because of tumor adhesions).
- tumor is well demarcated, with smooth, hypovascular capsule
- primary intracapsular debulking (CUSA is extremely useful) → removal of capsule (fragments of capsule adherent to important structures are left when necessary to avoid neural or vascular injury).
- although subtotal resection increases risk for recurrence, slow growth of epidermoid cysts makes this less problematic.
- capsule should not be removed from intramedullary cysts because of risk of causing neurological deficits or preoperative cyst rupture (radiographic finding of small fat globules in subarachnoid and intraventricular spaces)
- CONVENTIONAL TREATMENT:
  - steroid
  - no established role for radiotherapy or chemotherapy for residual / recurrent epidermoid cysts.

**DERMOID CYSTS** (from WHO manual)
- manifest at earlier age than epidermoids and have shorter duration of symptoms (duration of symptoms average 8.5 years, vs. 16 in epidermoids).
- female preponderance
- denser tumors than epidermoids → more focal mass effect - patients are initially seen at younger age (average age at diagnosis - 15 years, vs. 35-40 in epidermoids).
- tend to occur at MIDSITE (when extradural, may arise at anterior fontanelle).
- reported in association with Klippel-Feil syndrome.
PATHOLOGY
- positive for CA19-9* in many cases (serum CA19-9 levels tend to be higher than in epidermoid cysts - means of follow-up for residual or recurrent disease). *tumor marker for pancreatic cancer
- dermoids contain elements of dermis (hair and hair follicles + apocrine, sebaceous, or sweat glands)
- epithelial cell lining may be less differentiated (than in epidermoids)

Squamous epithelial cyst with adherent keratin debris, consistent with dermoid cyst. H & E, magnification x 40.

CLINICAL FINDINGS
- local neural deficits, headache, or meningitis.
- dermoid cyst rupture - previously thought to be uniformly fatal.

IMAGING
CT - hypodense and avascular, no contrast enhancement.
- extradural cranial lesion shows typical bony erosive changes seen with epidermoids.

MRI - more signal heterogeneity than epidermoids; high fat content - high signal on both T1- and T2-weighted images – unique tumor!
- nonenhancing (rare cases of capsule contrast enhancement and even enhancing mural nodule should raise suspicion for neoplasm!).
- more solid (than epidermoid tumors) - less likely to grow between neurovascular structures and tend to demonstrate more of local mass effect.
- edema is lacking.
- dermoid cyst rupture → fat droplets may be seen throughout subarachnoid or intraventricular space, localized dissemination in sulci causing widening, perhaps contained by pia or inflammatory tissue; hydrocephalus secondary to CSF obstruction by fat droplets.

TREATMENT
- surgical extirpation is treatment of choice - less problematic (than epidermoid tumors) because of firmer consistency, adherence of tumor capsule to vascular and neural structures → more conservative surgical approach.
- complete excision decreases risk for both postoperative chemical meningitis and tumor recurrence.
- tumor recurrence has been reported → reoperate.
- no current role for radiotherapy or chemotherapy.

COLLOID CYSTS
- congenital benign tumor that can cause sudden death because of their location (almost always found in 3rd ventricle → obstructive hydrocephalus).
- 0.5-1% of all primary brain tumors (15-20% of all intraventricular masses).

ETIOLOGY
- possible sources:
  a) in 1930, Sjöwall hypothesized that colloid cysts are remnants of PARAPHYSIS (embryonic midline structure within diencephalic roof immediately rostral to telencephalic border, in posterior lip of foramen of Monro) - cells of paraphysis are similar to those found in colloid cysts (i.e. low columnar epithelial cells without cilia or blepharoplasts) - colloid cysts were called paraphysal cysts for 50 years.
  b) diencephalic ependyma
c) invagination of neuroepithelium of ventricle
d) respiratory epithelium of endodermal origin.

PATHOLOGY
- grise in anterior superior portion of 3rd ventricle between fornices, immediately dorsal to Mono's foramen
- also have been reported to arise in septum pellucidum, 4th ventricle, sella turcica.
- attached to roof of 3rd ventricle (and frequently to choroid plexus).
- gross appearance of small white ball
- lined with simple or pseudostratified cuboidal or low columnar ciliated epithelial cells (PAS-positive; stain positively for S100 and negatively for glial fibrillary acidic protein, vimentin, and neurofilament). epithelial lining secretes mucinous fluid (greenish, of variable viscosity and can be rubbery) → cyst enlargement.

CLINICAL FEATURES
- classic intermittent obstructive hydrocephalus with paroxysmal HEADACHE; associated with changing head position (large cyst obstructing Mono's foramen).

Pituitary headache - usually present in age 20-50 yrs (youngest reported case - 2-month-old infant).
- other reported symptoms (sometimes related to changes in posture):
The transcortical approach carries increased incidence of epilepsy. It may be necessary if it is attached to either thalamostriate veins or internal cerebral veins. After decompression, completely remove it in order to prevent recurrence. Leaving small portion of cyst behind is associated with amnesia. The cyst should be reaspirated and no hydrocephalus (occurrence appears to be low) has been reported; may not correlate to tumor size, degree of ventricular dilatation, or duration of symptoms; suggested mechanisms:

- acute hydrocephalus
- hypothalamic dysfunction

**Diagnosis**

CT - well delineated, round or ovoid, homogenous, 66% hyperdense (to surrounding parenchyma) and 33% isodense.
- most are 5-25 mm.
- typically nonenhancing and uncalcified (occasional thin rim of enhancement).
- viscosity* of cyst contents correlates more closely to radiodensity on CT than to density visible on MRI.

*viscosity determines most appropriate surgical approach: hyperdense cyst is more likely to have solid contents - more difficult to drain, but reduced capacity to enlarge over time.

MRI - hyperintense on T1 and hypointense on T2.
- amount of rim enhancement is variable.
- MRI differentiates collid cyst from basilar tip aneurysm (may have similar appearance on CT).

**TREATMENT - STRATEGY**

Immediate attention to hydrocephalus!

**Strategy**

- a) large cyst & hydrocephalus → surgery.
  - if patient is too ill → bilateral CSF diversion or unilateral septostomy (suboptimal because sudden death has been reported in absence of acute obstructive hydrocephalus)
- b) small cyst, large ventricles, few or no symptoms → observation with serial MRIs.
- c) small cysts and normal-sized ventricles → observation.

N.B. prevention of sudden death is not indication for surgery in asymptomatic patients with small cysts and no hydrocephalus!
- explain to patient that cyst stretches fornix - look for preoperative short memory deficits – those may worsen postop if fornix is further violated surgically.

**TREATMENT - SURGERY**

Surgical approaches:

**Transcortical approach**

The transcortical approach involves making corticectomy over middle frontal gyrus and proceeding to frontal horn of lateral ventricle. Intraoperative ultrasonography may aid in approach to ventricle. The Monro foramen is visualized at convergence of septal veins, thalamostriate vein, and choroid plexus. The fornix arches over superior and anterior margins of foramen. Avoiding fornix is important because unilateral fornix damage has been associated with amnesia. The cyst should be readily visualized through foramen. The cyst is punctured and aspirated, internally decompressing walls of cyst. Avoid excessive retraction of walls of lateral ventricle because genu of internal capsule is in subependyma. Other concerns include damaging thalamostriate veins, which can result in basal ganglia damage. After cyst has been decompressed, completely remove it in order to prevent recurrence. Leaving small portion of cyst behind may be necessary if it is attached to either thalamostriate or internal cerebral veins. The transcortical approach carries increased incidence of epilepsy.

**Approximate Photograph Through Operating Microscope**

- Transcortical approach shows colloid cyst in Monro foramen. Choroid plexus is observed overlying cyst, and thalamostriate vein is along inferior border.
Intraoperative photograph showing removal of cyst, leaving dilated Monro foramen. The third ventricle can be seen through opening.

Endoscopic approach
The endoscopic approach is the same as the transcortical approach, with the exception that former is accomplished through burr hole. The endoscope is passed into the cyst, and the cyst is aspirated through working channels of the endoscope. This approach is least invasive, but it can be used only on cysts that can be aspirated. Hydrocephalus can persist after surgery, even after resection of cyst. This complication may be secondary to spillage of cyst contents or to bleeding during surgery. A ventricular catheter may be placed intraoperatively to safeguard against ventricular dilatation.

Postoperative follow-up
Hydrocephalus may develop despite cyst removal. H: periodic CT. Cyst may recur if partially excised!

**ARACHNOID CYSTS**

References
Mark S. Greenberg “Handbook of Neurosurgery” 8th ed. (2016), ch. 15.1

**Features**
- Congenital - splitting of arachnoid membrane (thus they are technically intra-arachnoid cysts), contain fluid identical to CSF.
- Arise anywhere on brain surface or in spinal canal.
- Almost all occur in relation to an arachnoid cistern (exception: intrasellar - only one that is extradural).
- Incidence: 5 per 1000 in autopsy series (~1% of intracranial masses).
- Bilateral arachnoid cysts may occur in Hurler syndrome (a mucopolysaccharidosis).
- Lined with meningothelial cells positive for epithelial membrane antigen (EMA) and negative for carcinoembryonic antigen (CEA) - two types:
  1) “simple arachnoid cysts”: arachnoid lining with cells that appear to be capable of active CSF secretion (middle fossa cysts seem to be exclusively of this type).
  2) Cysts with more complex lining which may also contain neuroglia, ependyma, and other tissue types.
- Do not communicate with the ventricles or subarachnoid space.
- Uniloculated or have septations.
- Incidence of growth: ~2% (some grow to remarkable size).
- Smooth surface (vs. epidermoid - cauliflower-like deep clefts).
- May hemorrhage (into cyst or subdural compartment): middle fossa cysts are notorious for hemorrhage due to tearing of bridging veins - some sports organizations do not allow participation in contact sports for these patients.
- Obsolete term “temporal lobe agenesis syndrome” - findings with middle cranial fossa ACs; however, the brain volumes on each side are actually the same (bone expansion and brain matter account for the parenchyma that appears to be replaced by the AC).
- Clinically: majority are incidental findings; most commonly symptomatic location - suprasellar: a) Hydrocephalus (probably due to compression of the third ventricle).
  b) Endocrine symptoms: occurs in up to 60% (incl. precocious puberty).
  c) Head bobbing (the so-called “bobble-head doll syndrome” - suggested suggestive of suprasellar cysts, but occurs in as few as 10%).
  d) Visual impairment.
- Imaging: menancing, extraparenchymal cyst with characteristics exactly mimicking CSF:
  a) Hydrocephalus (due to compression of the third ventricle).
  b) Endocrine symptoms: occurs in up to 60% (incl. precocious puberty).
  c) Head bobbing (so-called “bobble-head doll syndrome” - suggested suggestive of suprasellar cysts, but occurs in as few as 10%).
  d) Visual impairment.
  e) No calcify!
DERMOID, EPIDERMOID, CYSTS, LIPOMA

Type I:
- Small, brown, located in subcutaneous tissue (no mass effect). Communicates with subcutaneous space on water-soluble contrast CT/sinogram (WS-C/CT).
- Type II involves normal extradural/sinusoid segments of Sylvian fissures. Completely open results in oval shape. Partial communication on WS-C/CT.
- Type III involves entire Sylvian fissures. Marked midline shift. Early expansion of middle fossa (absence of lesser wing of sphenoid; outward expansion of sphenoid sinus turbinates). Minimal communication on WS-C/CT. Surgical treatment usually does not result in total decompression of brain (may approach type I lesion).

T1-MRI: large, fluid-filled structure expands left cerebellopontine angle cistern (arrowheads); note elongation and thinning of cranial nerves VII and VIII (white arrow).

TREATMENT:
- Asymptomatic: best left alone regardless of size or location (in adults, a single follow-up imaging in 6–8 months is adequate to rule out any increase in size; children need to be followed until adulthood).
- Indications for surgical treatment:
  1. Accelerated head growth in younger patients.
  2. Significant midline shift.
  3. Enlarging over time.
  4. Obstructive hydrocephalus.
- Surgery in symptomatic cases yields good results:
  a) Cyst aspiration: high rate of recurrence.
  b) Cyst fenestration: best treatment!
  c) Cyst shunting: second best treatment (due to risk of shunt dependence); for shunting into peritoneum, use a low pressure valve; if there is concurrent ventriculomegaly, may also place a ventricular shunt (e.g., through a “Y” connector).
  d) Cyst excision: significant morbidity and mortality (may be due to abrupt decompression), subsequent scarring may block fenestration allowing reaccumulation of cyst, flow through subarachnoid space may be deficient.

Suprasellar cysts (endocrinopathies tend to persist even after successful treatment of suprasellar cysts):
- Subfrontal or transcaldosal cystectomy: dangerous and ineffective.

Middle fossa cysts: shunting may be accomplished through the lateral ventricle, thus shunting both compartments (distal shunt tubing should be routed behind the ear; do not tunnel in front of ear— injury to facial nerve or solicit the plastic surgeon to help avoid the facial nerve).

LIPOMA
- Derived from mesoderm.
- Occur chiefly in midline (esp. over corpus callosum*, vermis, quadrigeminal cistern, spinal dural sac).
- Often associated with callosal dysgenesis.
- Majority are incidental findings.
- Characteristic appearance on both CT and MRI - fat density, calcification is frequent in periphery.

MUCOCELE
- Frontal mucocele (contrast MRI) - large mucocele compressing frontal lobe, with chronic inflammation of nasal muco-mucocutaneous nasal sinuses.

CT Choroid of sylvian fissure anechoic cysts.
Type I: small, brown, located in extracerebral space (no mass effect). Communicates with subcutaneous space on water-soluble contrast CT/sinogram (WS-C/CT).
Type II: involves normal extradural/sinusoid segments of Sylvian fissures. Completely open results in oval shape. Partial communication on WS-C/CT.
Type III: involves entire Sylvian fissures. Marked midline shift. Early expansion of middle fossa (absence of lesser wing of sphenoid; outward expansion of sphenoid sinus turbinates). Minimal communication on WS-C/CT. Surgical treatment usually does not result in total decompression of brain (may approach type I lesion).

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