Dermoid, Epidermoid, Cysts, Lipoma

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DERMOID tumor, EPIDERMOID tumor (s. CHOLESTEATOMA)

- 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. *DERMOIDS*)
  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).

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* has pearly appearance ("pearly tumors") due to capsule of stratified squamous epithelium.

> *EPIDERMOIDS* 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).

vs. *TERATOMAS* - composed of *ectoderm*, *mesoderm*, and *endoderm*

- *expand slowly over many years* due to central accumulation of epithelial debris and glandular secretions.
- *DERMOIDS* frequently calcify; *EPIDERMOIDS* 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):

DERMOIDS (INTRACRANIAL) - most commonly **midline**: posterior fossa (extradural, vermian, or within 4th ventricle), suprasellar region, subfrontal areas; other sites – scalp (commonest location in childhood), skull, orbit, nasal, oral cavity, neck.

DERMOIDS (SPINAL) - most commonly near thoracolumbar junction, tends to involve conus medullaris and cauda equina:

intramedullary ≈ 50%
intradural extramedullary ≈ 50%
extradural ≈ least common

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

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

EPIDEMIOLOGY

DERMOID - uncommon (≈ 0.3% of all brain tumors).
EPIDERMOID - 4-10 times more frequent than dermoid (≈ 2% of all intracranial tumors).

CLINICAL FEATURES

Patient's age at diagnosis:
DERMOID - up to 20 yrs
EPIDERMOID (enlarges more slowly than dermoid) - 40-50 yrs.

Symptoms & signs are associated with slowly progressing **mass/pressure effect** (seizures, diabetes insipidus, hypopituitarism, etc).
- blockage of CSF flow occurs only rarely!
- cyst **rupture** → intense granulomatous chemical meningitis (rarely results in infarction from vasospasm).
- associated dermal sinus tracts / dimples are common:

> Any infant with dermal sinus tract → neuroradiological evaluation!

congenital lumbar dermal sinus may terminate in *EPIDERMOID* (less frequently *DERMOID*) within or near conus medullaris or cauda equina; often associated with spinal dysraphism and vertebral abnormalities.
congenital nasal dermal sinus may be associated with *DERMOID* or *EPIDERMOID*.
- if associated *dermal sinus tract* becomes **infected** → recurrent bacterial meningitis.

DIAGNOSIS

Fat components (glandular secretions of dermoids) are 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* fat gives very low density (may be slightly heterogeneous due to additional ectodermal 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 – midline mass hyperintense on T1 and hypointense on T2.

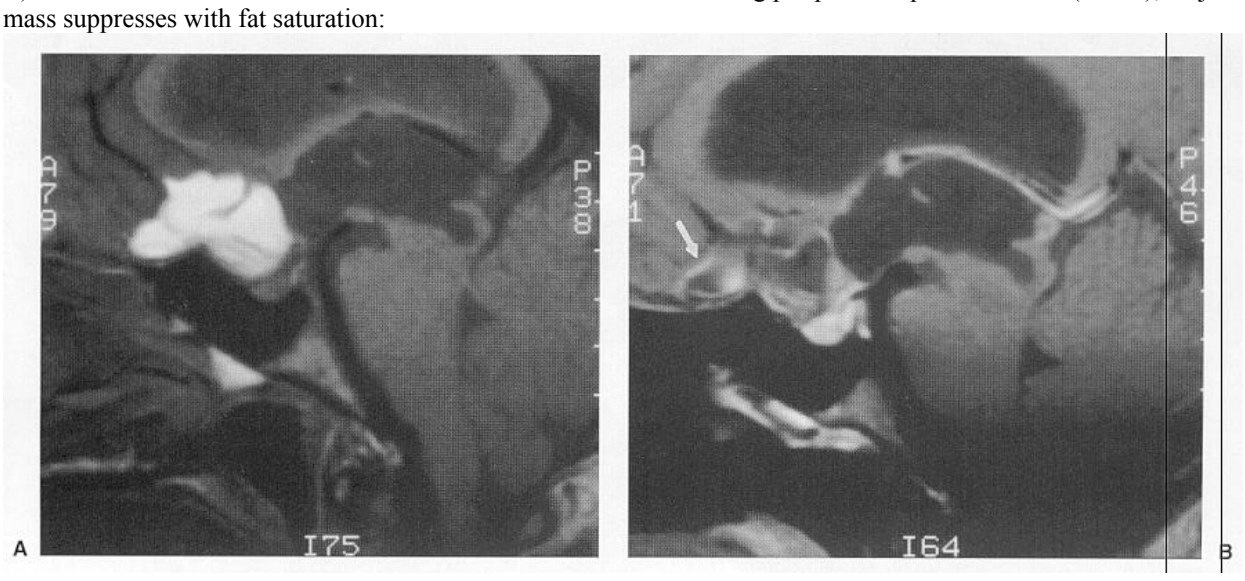
- *chemical-shift artifact* is often present on T2 images as markedly hypointense band posterior at fat-fluid interface.

EPIDERMOID - characteristics similar to CSF – variably hypointense on T1 and variably hyperintense on T2.

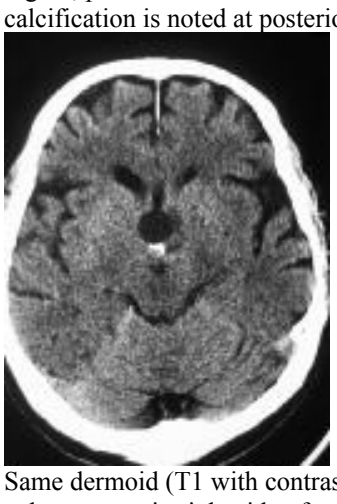
Angiography - avascular mass.

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

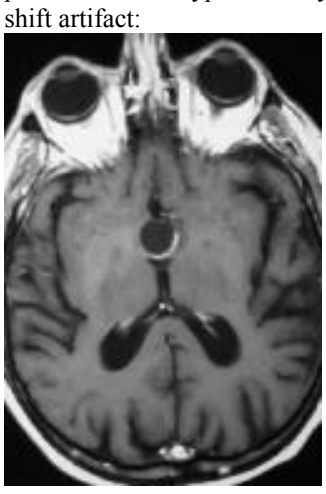
Suprasellar dermoid:
A) noncontrast T1 - high-signal-intensity suprasellar mass extending along planum sphenoidale.
B) contrast T1 with fat saturation - small amount of enhancement along peripheral aspects of lesion (*arrow*); majority of mass suppresses with fat saturation:



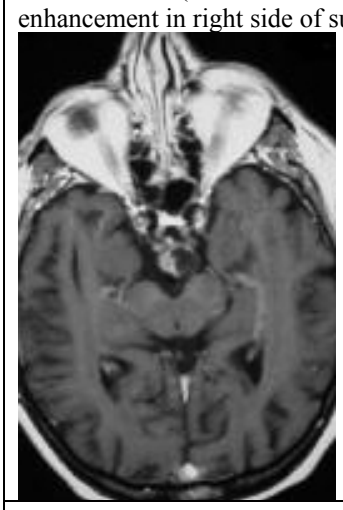
Dermoid (nonenhanced CT) - well-circumscribed, cystic, low-attenuating lesion at midline in suprasellar region, posterior to 3rd ventricle; small focus of calcification is noted at posterior margin of tumor:



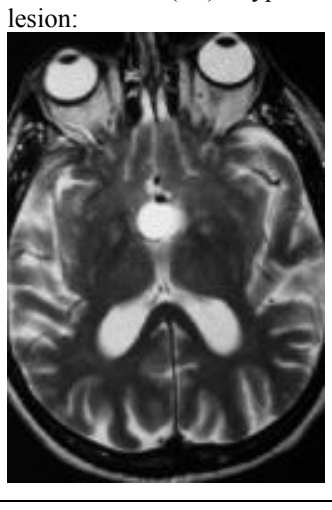
Same dermoid (T1) - hypointense lesion; crescentic posterior rim of hyperintensity represents fat chemical-shift artifact:



Same dermoid (T1 with contrast) - nodular focus of enhancement in right side of suprasellar lesion:



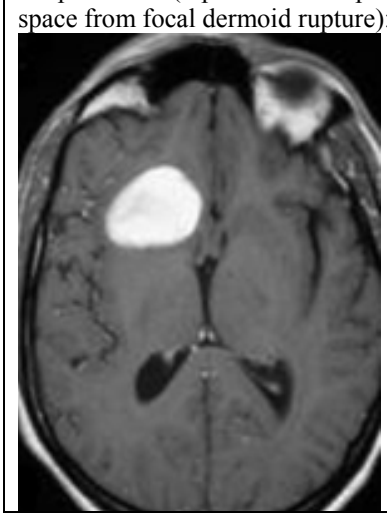
Same dermoid (T2) - hyperintense cystic component in lesion:



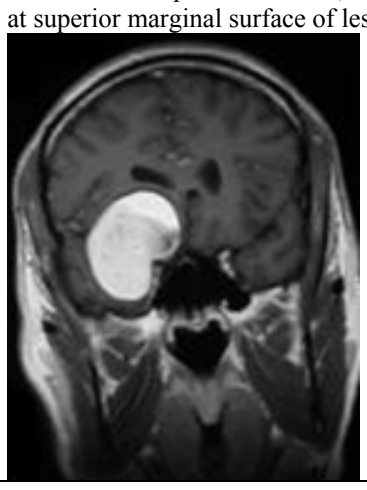
Dermoid (nonenhanced CT) - large, well-circumscribed low-attenuating cystic lesion in right temporal lobe lateral to cranial midline; peripheral marginal calcification; no erosion in adjacent bone of sella:

Same dermoid (contrast CT) - partial marginal enhancement; attenuation degree in center of lesion consistent with fat:

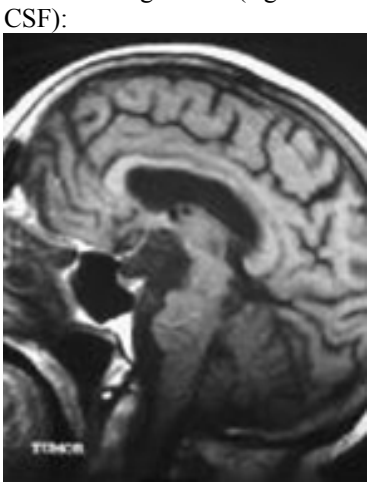
Same dermoid (T1) – hyperintense signal in lesion; multiple small hyperintense foci along sulci of right temporal lobe (represent fat droplets in subarachnoid space from focal dermoid rupture):



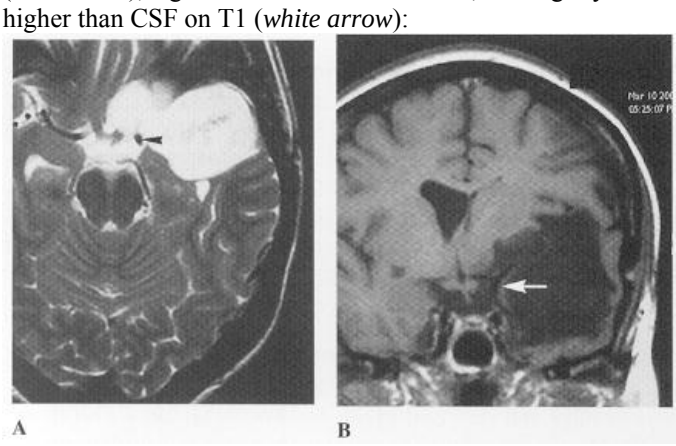
Same dermoid (T1 with contrast) - hyperintense lesion (hyperintensity is due to short T1 of fat); multiple hyperintense foci (fat droplets) in subarachnoid spaces; mild midline septal shift to left; chemical-shift artifact at superior marginal surface of lesion:



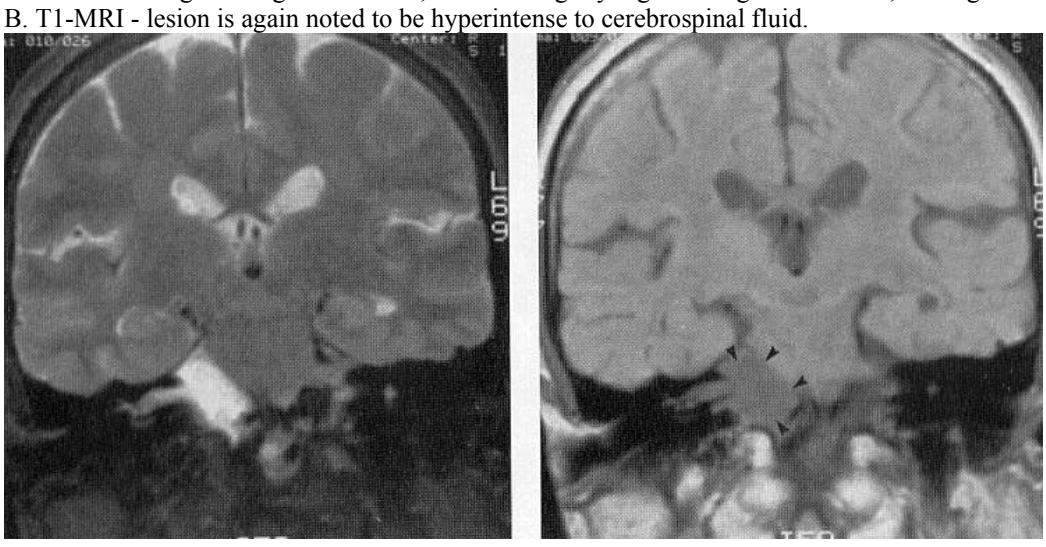
Epidermoid (T1 with contrast) - suprasellar, prepontine, and interpeduncular location of nonenhancing tumor (signal intensity similar to CSF):



Epidermoid (A- T2-weighted; B - T1-weighted MRI): 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-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.



TREATMENT

- complete **surgical excision** is curative.
  - epidermoids can interdigitate around vital neuronal structures, complicating surgical removal.
  - avoid spilling of contents (→ chemical meningitis).
  - associated dermal sinus should be removed completely.

Chemotherapy and radiotherapy are not useful.

COLLOID CYSTS

- congenital benign tumors 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 1910, Sjovall 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 *paraphysial cysts* for 50 years.
  b) diencephalic ***ependyma***
  c) invagination of ***neuroepithelium*** of ventricle
  d) ***respiratory epithelium*** of endodermal origin.

PATHOLOGY

- arise in anterior superior portion of 3rd ventricle between fornices, immediately dorsal to Monro 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) → cyst enlargement.

CLINICAL FEATURES

- classic intermittent obstructive hydrocephalus with paroxysmal headache associated with changing head position (large cyst obstructing Monro foramen).

Positional headache!

- usually present in age 20-50 yrs (youngest reported case - 2-month-old infant).
- other reported symptoms (sometimes related to changes in posture):
  - sudden weakness in lower limbs associated with falls without loss of consciousness.
  - symptoms similar to normal pressure hydrocephalus (dementia, gait disturbance, urinary incontinence).

  Mental changes are common (may persist after surgery if fornix is damaged)!
- ***sudden death*** (incidence appears to be low) has been reported; may not correlate to tumor size, degree of ventricular dilatation, or duration of symptoms.

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.

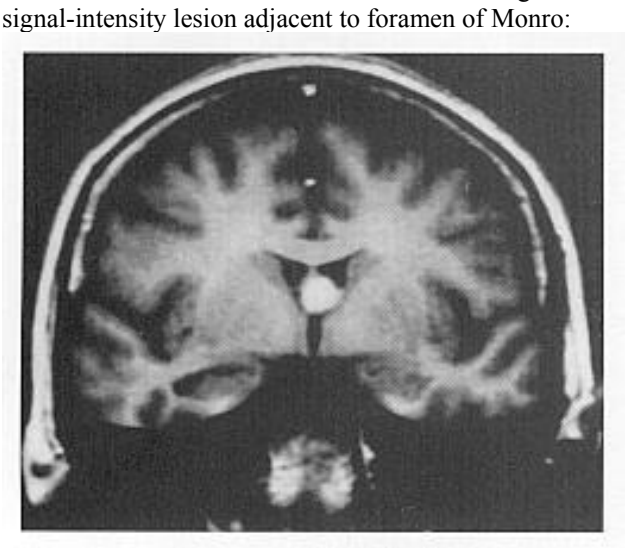
  **viscosity determines most appropriate surgical approach*; hyperdense cyst is more likely to have solid contents - more difficult to drain, but reduced capacity to enlarge over time.

MRI - hyperintense on T1 and hypointense on T2.

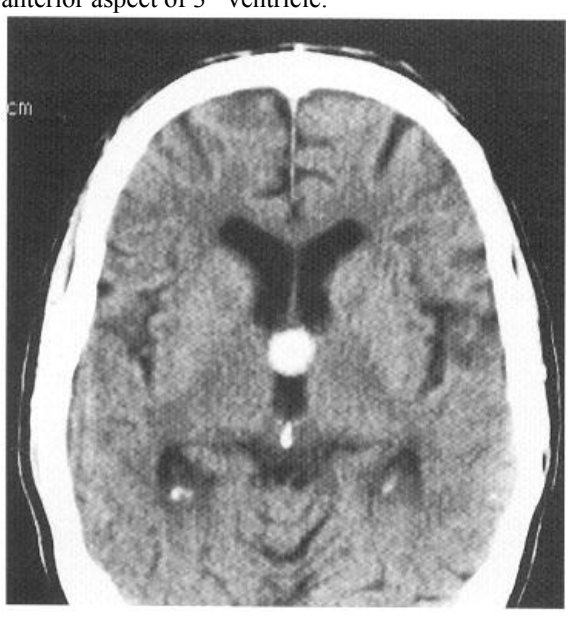
N.B. CSF flow artifact at Monro foramen can mimic colloid cyst!

- amount of rim enhancement is variable.
- MRI differentiates colloid cyst from basilar tip aneurysm (may have similar appearance on CT).

T1-MRI without contrast: well-circumscribed high-signal-intensity lesion adjacent to foramen of Monro:



CT - dense, rounded mass in region of foramina of Monro causing enlargement of lateral ventricles, and indenting anterior aspect of 3rd ventricle:



TREATMENT

Immediate attention to hydrocephalus!
Strategy:

a) large cyst & hydrocephalus → **surgery**.
   - *if patient is too ill* → bilateral CSF diversion (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!

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 contents are 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.

Intraoperative photograph through operating microscope 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:

Transcallosal approach

The transcallosal approach avoids incising cortex. A right frontal craniotomy is made two thirds anterior and one third posterior to coronal suture, crossing midline to expose superior sagittal sinus. The right frontal lobe is then retracted laterally, and corpus callosum is exposed. Draining cortical veins must be avoided if possible. The preoperative MRI can help identify potential veins. A 1-centimeter incision is made in corpus callosum, allowing entry to lateral ventricle. The Monro foramen can then be visualized, and septum pellucidum may be divided to see contralateral foramen. Either ventricle may be entered through standard right frontal transcallosal approach. Close inspection of orientation of choroid plexus, caudate nucleus, and Monro foramen helps determine which of ventricles has been entered.
The transcallosal approach decreases risk of postoperative epilepsy but risks venous infarction and contralateral leg weakness from prolonged retraction. An extensive callosal resection may also cause temporary mutism. Excessive manipulation of fornix may affect memory.

Endoscopic approach

The endoscopic approach is same as transcortical approach, with exception that former is accomplished through burr hole. The cyst is punctured and aspirated through working channels of endoscope.
The endoscopic 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.

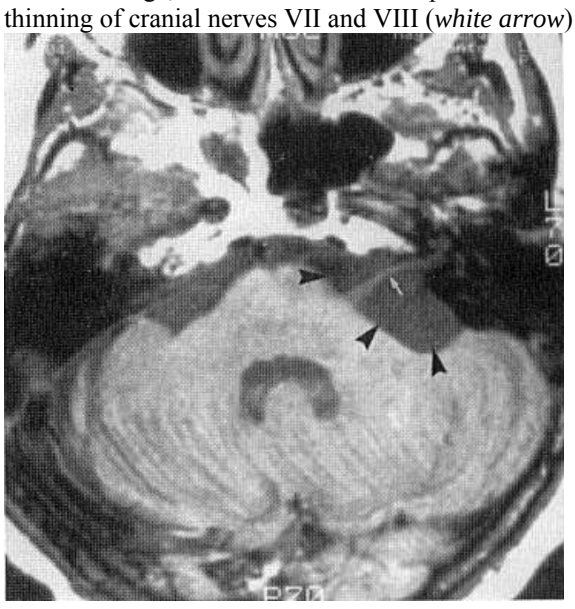
POSTOPERATIVE FOLLOW-UP

Hydrocephalus may develop despite cyst removal; H: periodic CT.

ARACHNOID CYSTS

- arise ***anywhere on brain surface***.
- some grow to remarkable size.
- smooth surface (vs. *EPIDERMOIDS* - cauliflower-like deep clefts).
- never calcify!
- majority are *incidental findings* - best left alone.

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



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.

BIBLIOGRAPHY for ch. "Neuro-Oncology" → follow this LINK >>