

Choroid Plexus Tumors

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CHOROID PLEXUS PAPILOMA

- rare benign neoplasm of cuboidal **neuroepithelial cells** lining papillae of choroid plexus.
- < 1% of brain tumors, but 3% of childhood intracranial neoplasms (esp. younger groups: 4-6% in children < 2 years and 10-20% in infants).
- male-to-female ratio = 2.8 : 1

ETIOLOGY

- have been associated with von *Hippel-Lindau syndrome*, *Li-Fraumeni syndrome*, *Aicardi syndrome*.
- one etiologic theory - presence of **simian vacuolating virus No. 40 (SV40)**–related viral DNA; SV40 is oncogenic virus with ability to inactivate both Rb and p53 proteins.

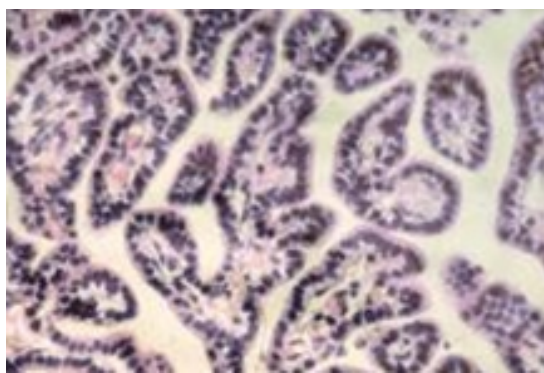
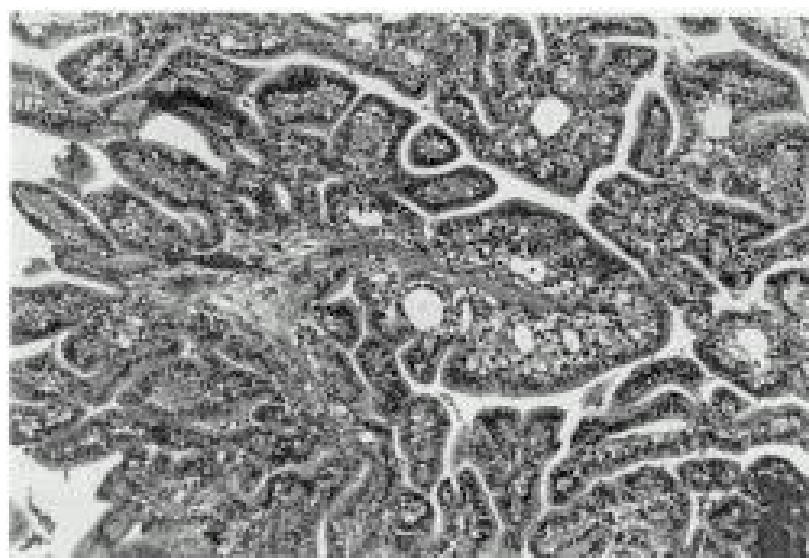
LOCATION

- corresponds to normal choroid plexus locations:
 - 1) **60%** - **lateral ventricles** (most common location for *children*) - lining of body, trigone, inferior horn
 - 2) **foramen of Monro**
 - 3) **roof of 3rd ventricle**
 - 4) **30%** - posterior portion of roof of **4th ventricle** → cerebellopontine angle (most common location for *adults*)

Most common in lateral ventricles of children!

PATHOLOGY

- fill ventricles and compress walls.
- often pedunculated, giving it some mobility.
- slow growing, does not invade brain parenchyma (often reach size of 60-70 g before are diagnosed).
- histologically differentiating normal choroid plexus from papilloma can be very difficult.
 - normal choroid plexus epithelium tends to have more "hobnail" shape on ventricular side, whereas papilloma epithelium is more flattened.
- occasionally may spread via CSF.



CLINICAL FEATURES

- **intracranial hypertension**:
 - 1) **HYDROCEPHALUS**:
 - a) increased CSF secretion by tumor cells (up to 4 times of normal)
 - b) obstructed CSF flow.
 - c) damage to CSF resorptive bed from recurrent hemorrhages (basilar arachnoiditis).
 - 2) later – **MASS EFFECT**.
- tumors in 4th ventricle frequently produce *cerebellar findings*.
- sometimes manifest in *cerebellopontine angle*.

DIAGNOSIS

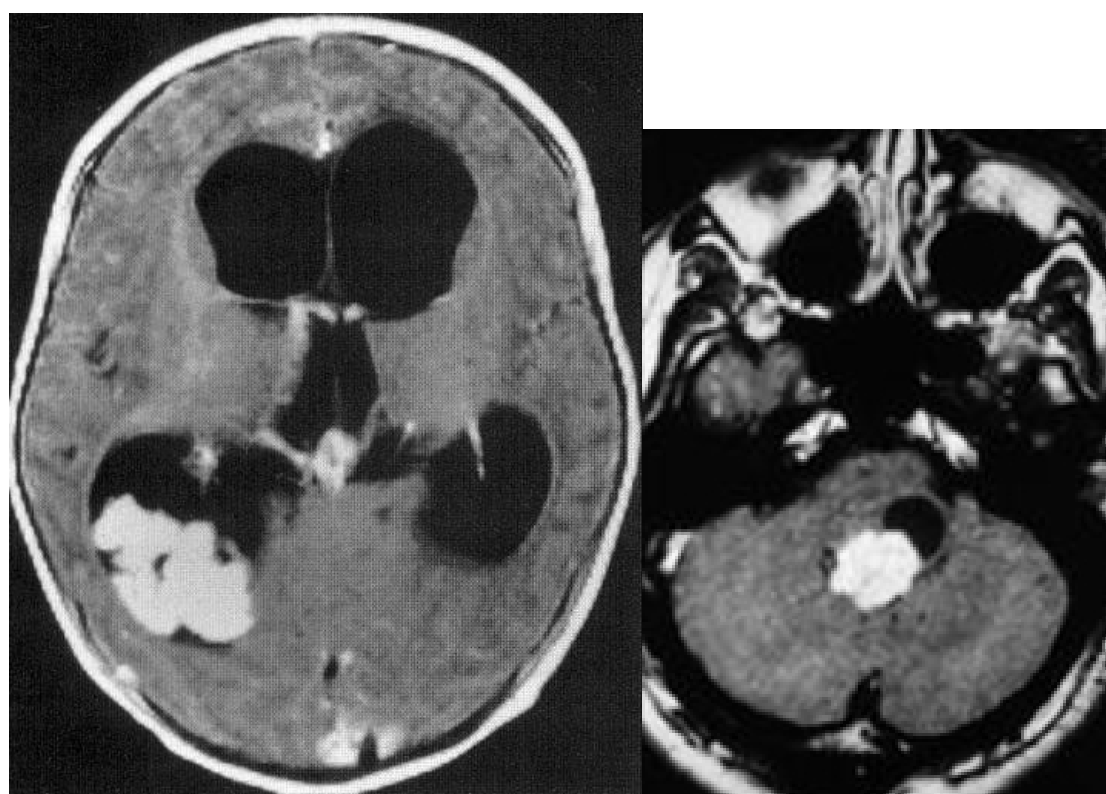
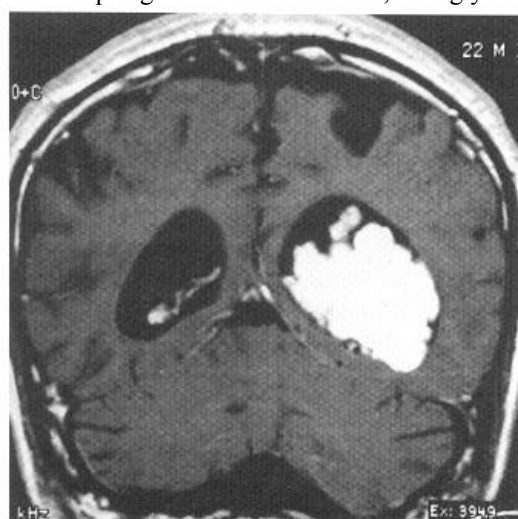
Imaging - lobulated, 'cauliflower-like' hypodense ÷ slightly hyperdense mass with cystic areas, located within or near ventricular system.

N.B. papilloma is within choroid plexus and thus outside BBB - *contrast enhancement is marked!*

- edema, invasion into surrounding parenchyma.

- **punctate calcifications** (20% tumors);
vs. **global calcification** throughout mass - more indicative of CARCINOMA!
- heterogeneous MRI signal intensity – reflects high vascularity, calcifications, hemorrhages.
- determination of **tumor stalk** location is crucial – dictates surgical approach!

T1-MRI postgadolinium - lobulated, strongly enhancing tumour in trigone of left lateral ventricle:



MRI – 3rd ventricular ch. plexus papilloma extending into lateral ventricles:



CSF - protein↑, xanthochromia.

Can be extremely vascular! (blood supply derived from choroidal arteries) - **angiography** is sometimes indicated before surgery.

TREATMENT

Persistently increased ICP is not compatible with life - watchful waiting is inappropriate!

Surgical removal of mass! (many intraventricular lesions can be totally resected through endoscope)

High incidence of surgical cure! (gross total resection nearly always effects cure)

- **normalize excessive CSF pressure** prior to surgery - *repeated lumbar punctures* or *ventricular shunt* (in older patient).

Intraventricular tumors outside posterior fossa are more easily removed if ventricles are large - preoperative shunts are usually not inserted in otherwise stable patients.

- **hydrocephalus may not resolve with surgery** (derangement of reabsorption mechanisms or blockage at other sites in ventricular system); H: shunting (required in up to 60% patients postoperatively!).

Treatment of hydrocephalus must be considered both before and after any surgery!

- **subtotally resected papillomas** require additional therapy:

- a) **reoperation**
- b) (intraventricular) **chemotherapy**
- c) (craniospinal) **irradiation**.

CHOROID PLEXUS CARCINOMA (S. ANAPLASTIC CHOROID PLEXUS PAPILOMA)

- extremely rare malignant tumor of choroid plexus.

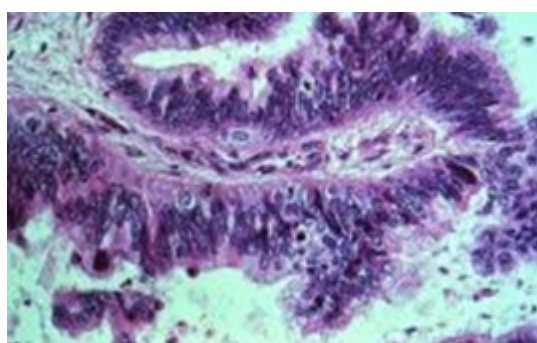
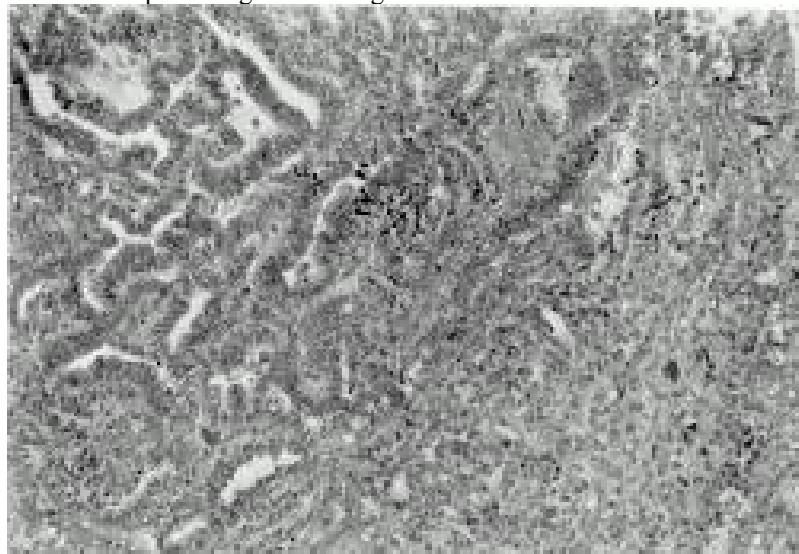
- usually found in children.
- significantly longer survival is noted in cases associated with +9p and -10q.

PATHOLOGY

- **anaplastic features**: nuclear pleomorphism and hyperchromasia, mitotic activity↑, tumor giant cells, tumor necrosis, frank invasion of brain parenchyma.

- extremely vascular.
- in adults, must be differentiated from much more common metastatic carcinoma.
- 44% cases disseminate along CSF pathways (diffuse and aggressive leptomeningeal spread).

Papillary character (partially retained in left side) has been lost on right side; note pseudostratified epithelium forming irregular glandular structures on left and diffuse epithelial growth on right:



DIAGNOSIS

Imaging - more heterogeneous than papilloma; *global calcification*; areas of necrosis.

- **craniospinal MRI** and **CSF cytology** - any evidence of seeding?

TREATMENT

Surgical removal of mass!

- resection may be facilitated by preoperative chemotherapy (**IPHOSPHAMIDE**, **CARBOPLATIN**, **VP-16**).
- adjuvant **chemotherapy** and **radiotherapy** have been demonstrated to increase survival;
 - **CARBOPLATIN** and **ETOPOSIDE** are commonly used.
 - in documented *leptomeningeal seeding*, use **craniospinal irradiation**.

BIBLIOGRAPHY for ch. "Neuro-Oncology" → follow this [LINK >>](#)