Choroid Plexus Tumors

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CHOROID PLEXUS PAPILLOMA 1
ETIOLOGY 1
LOCATION 1
PATHOLOGY 1
CLINICAL FEATURES 1
DIAGNOSIS 1
TREATMENT 3
CHOROID PLEXUS CARCINOMA (S. ANAPLASTIC CHOROID PLEXUS PAPILLOMA) 3
PATHOLOGY 3
DIAGNOSIS 4
TREATMENT 4

Papilloma (grade I) : Carcinoma (grade III) = 5 : 1
Practically, only in kids

CHOROID PLEXUS PAPILLOMA

- 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.

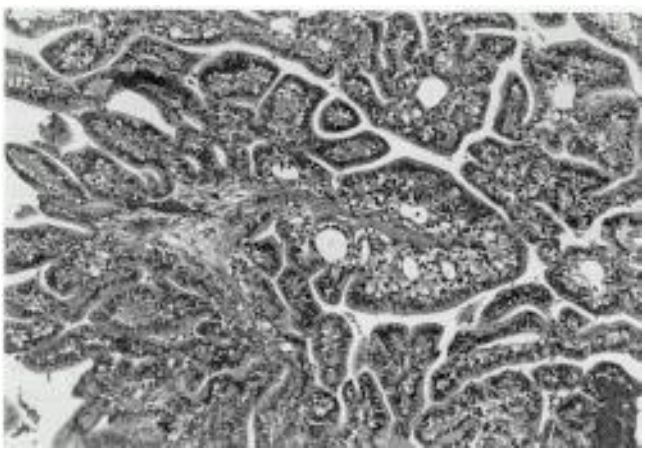
LOCATION

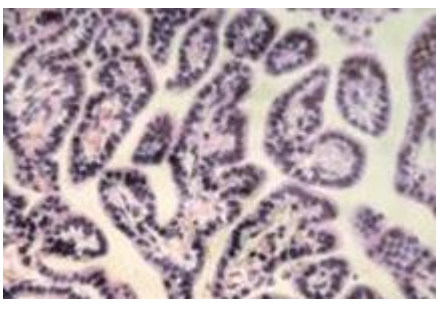
- corresponds to normal choroid plexus locations:
    1) 60% - lateral ventricles (most common location for *children*) - lining of body, trigone, inferior horn
    2) foramen of Monro
    3) roof of 3rd ventricle
    4) 30% - posterior portion of roof of 4th ventricle → cerebellopontine angle (most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

DIAGNOSIS

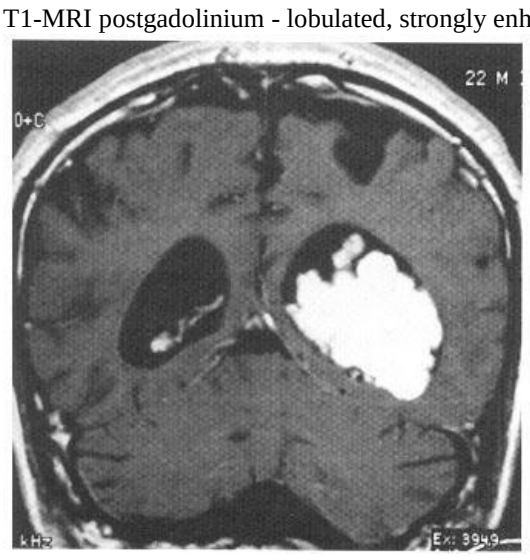
Imaging - lobulated, 'cauliflower-like' hypodense ÷ slightly hyperdense mass with cystic areas, located within or near ventricular system.

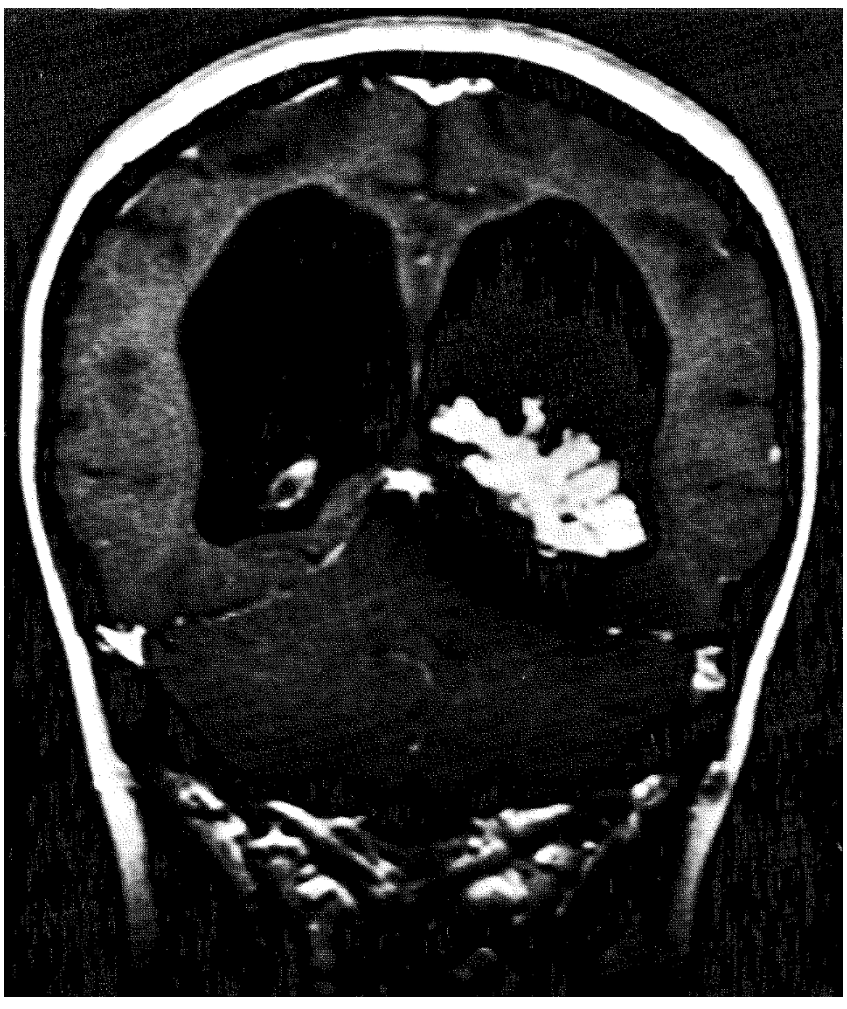
N.B. papilloma is within choroid plexus and thus outside BBB - *contrast enhancement is marked*!

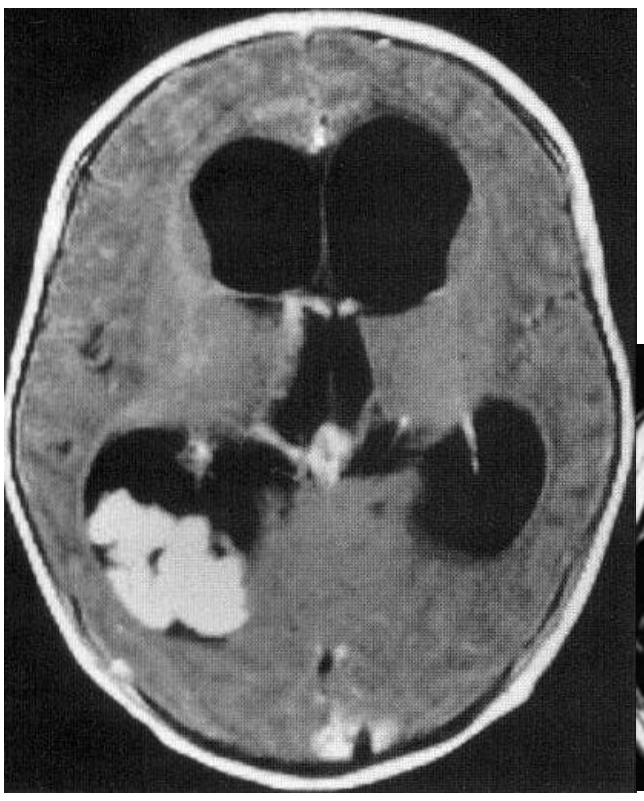
- edema, invasion into surrounding parenchyma.
- ***punctate calcifications*** (20% tumors);
    vs. *global calcification* throughout mass - more indicative of CARCINOMA!

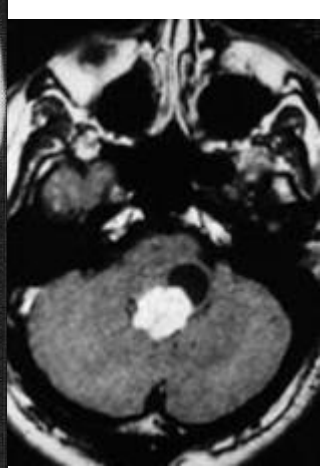
- heterogeneous MRI signal intensity – reflects high vascularity, calcifications, hemorrhages.
- determination of **tumor stalk** location is crucial – dictates surgical approach!

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

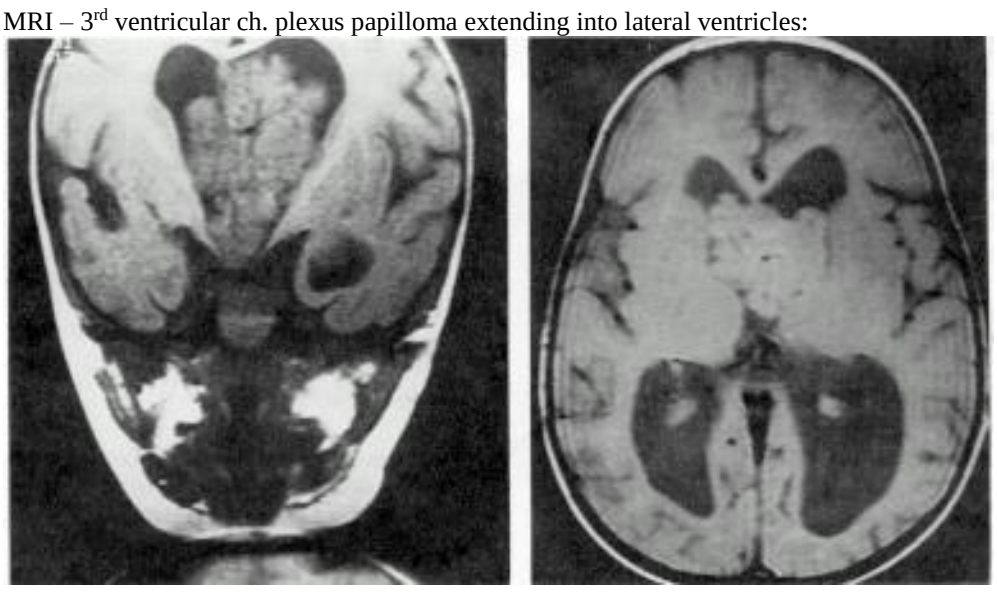


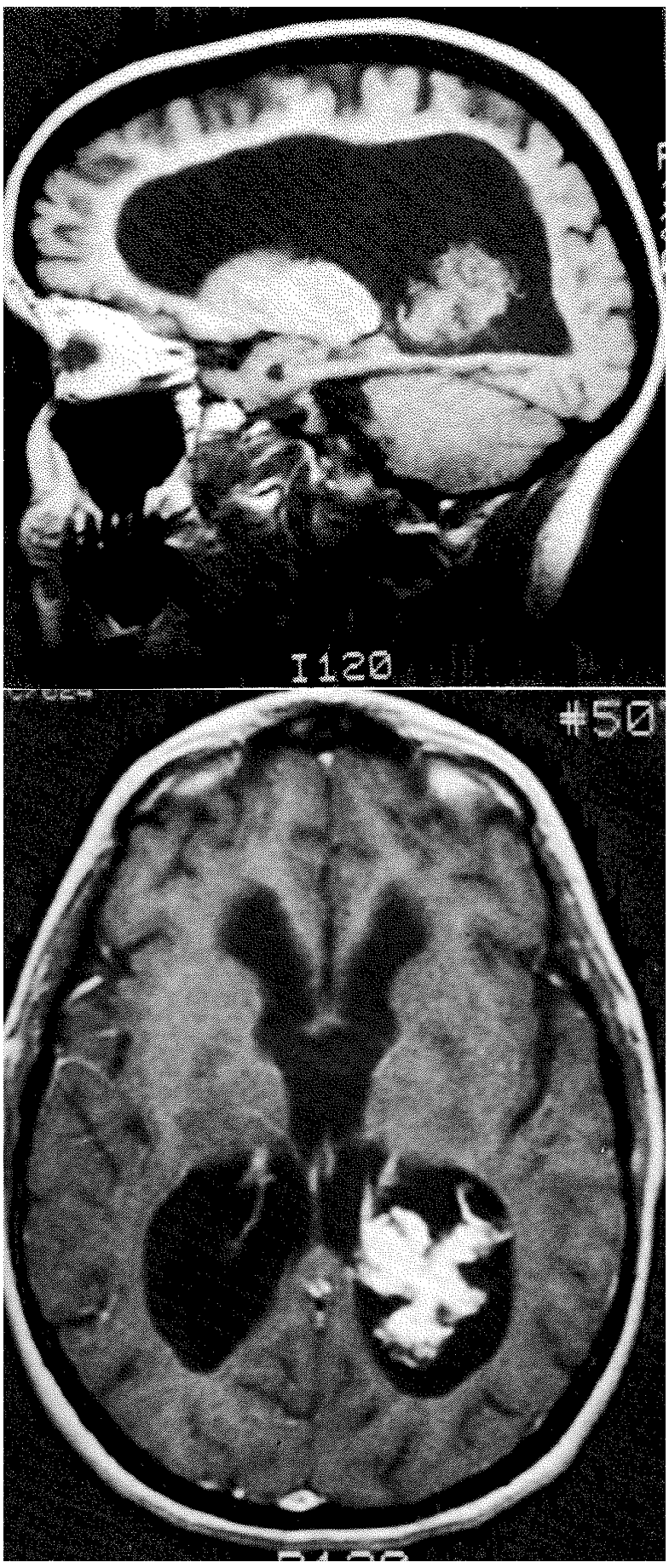






MRI – 3rd ventricular ch. plexus papilloma extending into lateral ventricles:





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**.

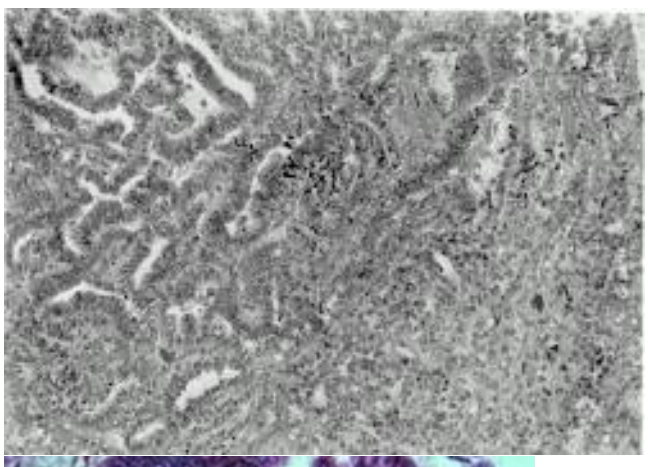
CHOROID PLEXUS CARCINOMA (s. ANAPLASTIC CHOROID PLEXUS PAPILLOMA)

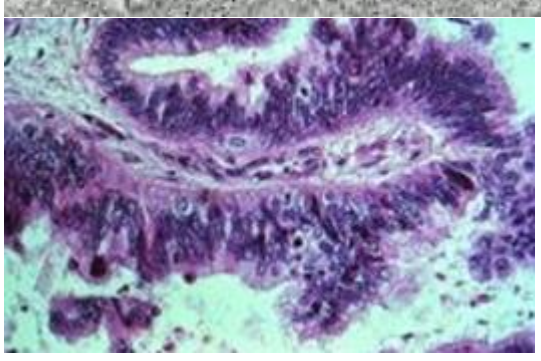
- extremely rare malignant tumor of choroid plexus.
- usually found in children.
- significantly longer survival is noted in cases associated with +9p and -10q.

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 >>