Choroid Plexus Tumors

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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

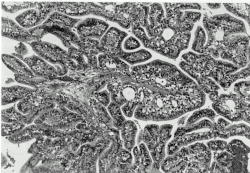
- corresponds to normal choroid plexus locations:

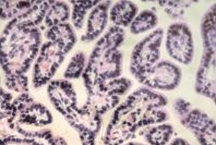
* 1. 60% - lateral ventricles (most common location for *children*) - lining of body, trigone, inferior horn
  2. foramen of Monro
  3. roof of 3rd ventricle
  4. 30% - posterior portion of roof of 4th ventricle → cerebellopontine angle (most common location for *adults*)

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**:
       1. increased CSF secretion by tumor cells (up to 4 times of normal)
       2. obstructed CSF flow.
       3. 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.

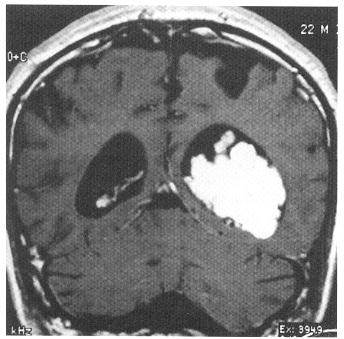
N.B. papilloma is within choroid plexus and thus outside BBB - *contrast enhancement is marked*!

* edema, invasion into surrounding parenchyma.
* ***punctate calcifications*** (20% tumors);

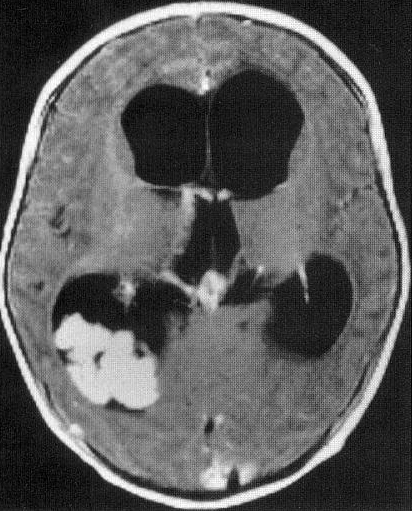
vs. *global calcification* throughout mass - more indicative of carcinoma!

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

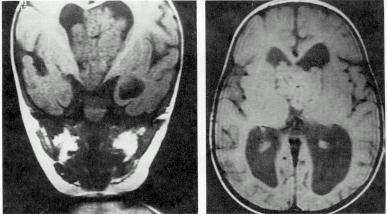
T1-MRI postgadolinium - lobulated, strongly enhancing tumour in trigone of left lateral ventricle:

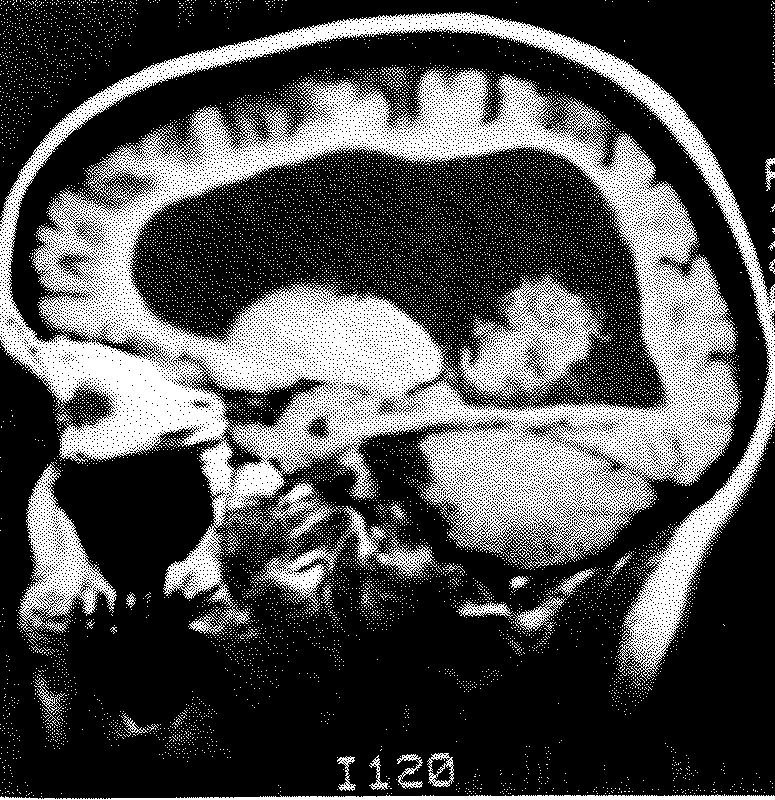
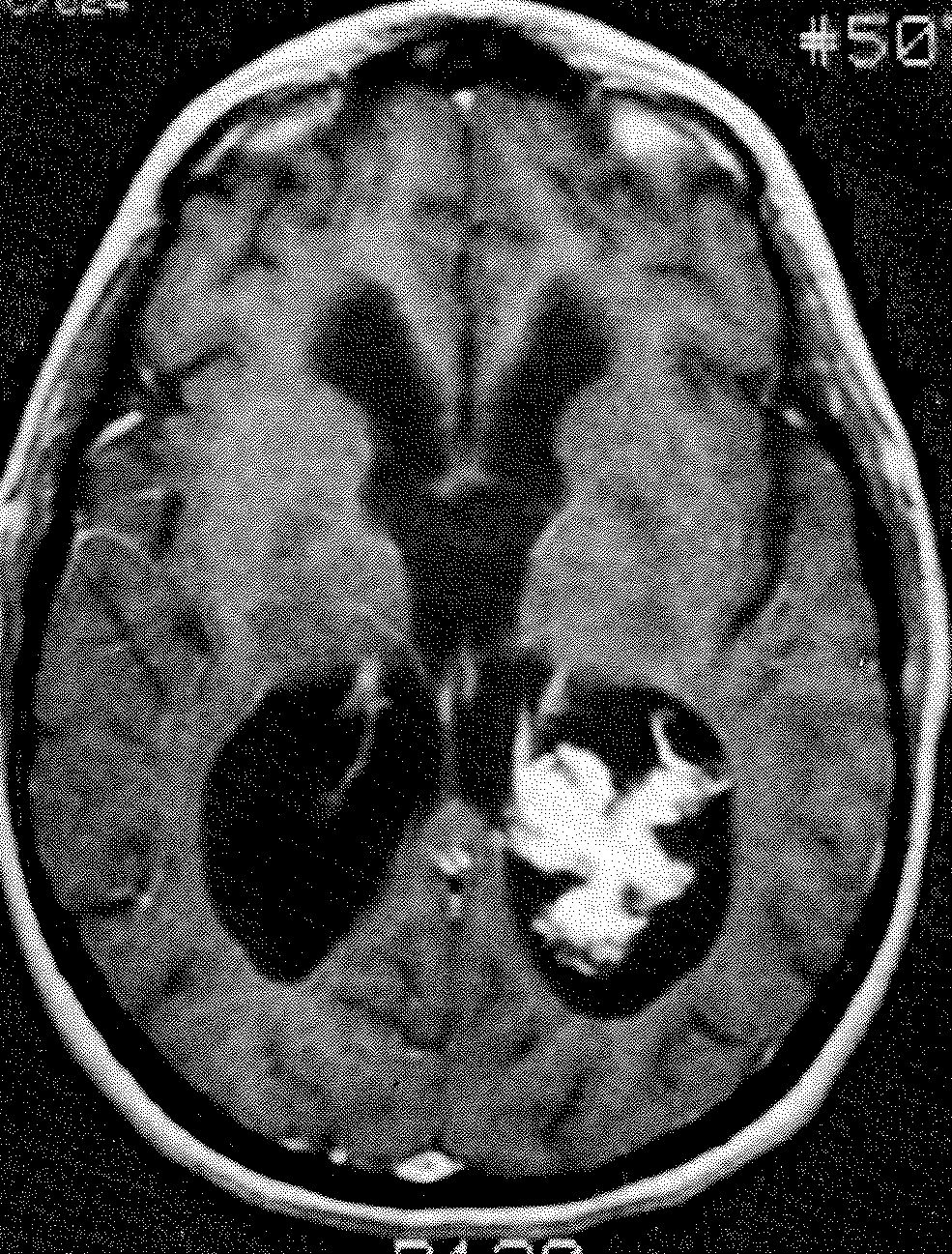






MRI – 3rd ventricular ch. plexus papilloma extending into lateral ventricles:



**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:
  + - 1. **reoperation**
      2. (intraventricular) **chemotherapy**
      3. (craniospinal) **irradiation**.

Choroid Plexus Carcinoma (s. Anaplastic Choroid Plexus Papilloma)

- extremely rare malignant tumor of choroid plexus.

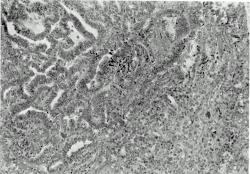
* usually found in children.
* significantly longer survival is noted in cases associated with +9p and -10q.

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 >>](http://www.neurosurgeryresident.net/Onc.%20Oncology\Onc.%20Bibliography.pdf)

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