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.
- 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) 40% - 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 “hubnail” shape on ventricular side, whereas papilloma epithelium is more flattened.
- occasionally may spread via CSF:

CLINICAL FEATURES
- Intraventricular 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 hypodense mass with cystic areas, located within or near ventricular system.
  - Null: 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).
- 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!
- subtotaly 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.
**CHOROID PLEXUS TUMORS**

**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](#).