Choroid Plexus Papilloma

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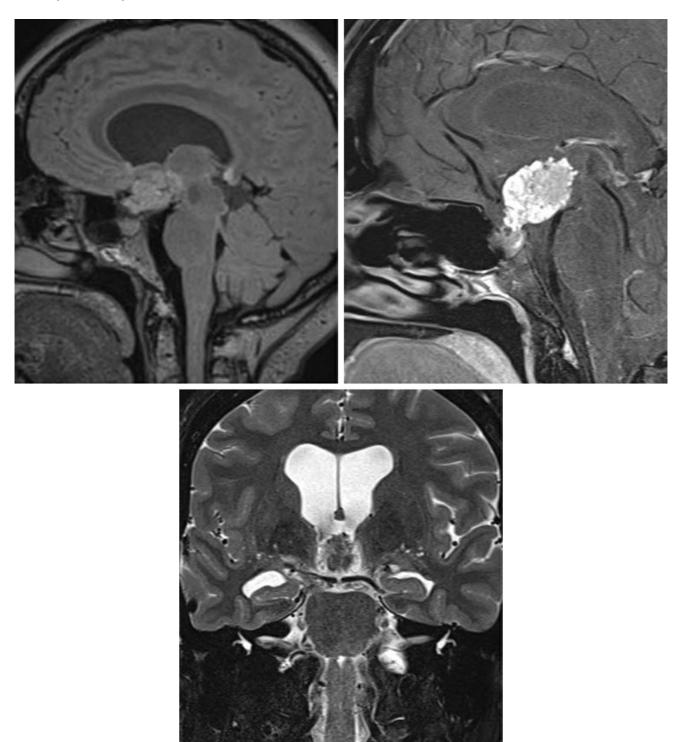


Figure 1: Sagittal FLAIR (top left) and T1-weighted postcontrast (top right) images demonstrate a lobular, avidly enhancing lesion within the third ventricle. (Bottom) T2-weighted coronal image illustrates dilatation

of the foramen of Monro and of the lateral ventricles due to obstruction by this CPP.

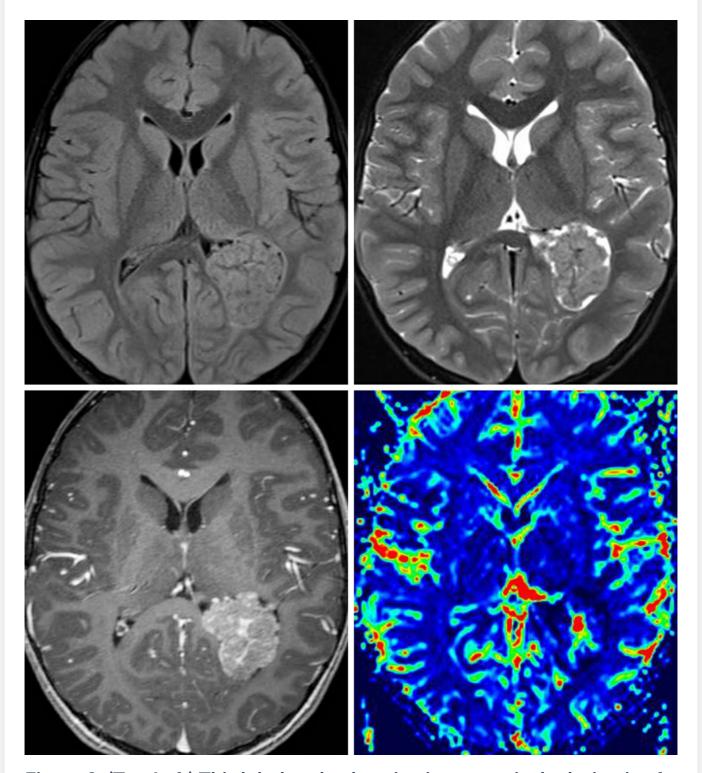


Figure 2: (Top Left) This lobulated enhancing intraventricular lesion is of intermediate signal on axial FLAIR image. (Top Right) Axial T2WI demonstrates surrounding CSF signal clefts that more easily characterize this lesion as intraventricular. This CPP also demonstrates avid enhancement on T1-weighted postcontrast imaging (bottom left) and hyperperfusion on cerebral blood volume imaging (bottom right).

BASIC DESCRIPTION

- Benign, lobulated, intraventricular mass arising from choroid plexus epithelium
- May disseminate via cerebrospinal fluid (CSF)

PATHOLOGY

- Fibrovascular connective tissue covered by cuboidal or columnar choroid plexus epithelium
- WHO grade I (typical choroid plexus papilloma [CPP]) or II (atypical CPP)
- Cysts and hemorrhage may be seen
- Rare malignant transformation
- Rare necrosis, invasion of adjacent brain parenchyma
 - Invasion suggests <u>choroid plexus carcinoma</u>
- Association with Li-Fraumeni and Aicardi syndromes
- Association with simian virus 40 (SV40) infection

CLINICAL FEATURES

- Usually arising in lateral or fouth ventricles
 - Lateral ventricle: majority of patients are <20 years old; no gender predilection
 - Fouth ventricle: adults are more commonly affected; slight male gender predilection
- Presenting signs/symptoms often related to increased intracranial pressure secondary to either CSF overproduction or obstruction/impaired CSF resorption (communicating hydrocephalus)
 - Macrocephaly, bulging fontanelles
 - Nausea, vomiting, headaches, ataxia
- Treatment
 - Gross-total resection

- Recurrence rare
- Excellent prognosis: 5-year survival rate, nearly 100%

IMAGING FEATURES

- General
 - Lobulated, frond-like, avidly enhancing intraventricular mass
 - Arising in regions of choroid plexus
 - Lateral ventricle atrium or trigone > foramen of Luschka or posterior medullary velum of fourth ventricle > third ventricular roof
 - Hemorrhage and calcification common
- CT
 - Isodense to hyperdense
 - ±Calcification, hydrocephalus
 - Avid homogenous enhancement on contrast-enhanced CT
- MRI
 - T1WI: isointense to hypointense
 - T2WI: isointense to hyperintense; ±flow voids
 - FLAIR: hyperintense periventricular signal secondary to transependymal CSF flow/interstitial edema from hydrocephalus
 - T2*/GRE/SWI: black signal blooming secondary to calcification or hemosiderin deposition
 - T1WI+C: avid homogeneous enhancement most common
 - MRA: may see flow-related signal within the tumor
 - MRS: elevated Cho, absent NAA, lactate peak if necrotic, elevated myoinositol may differentiate from <u>choroid plexus</u> <u>carcinoma</u>

IMAGING RECOMMENDATIONS

MRI with contrast, include both brain and spine due to risk of CSF

dissemination

For more information, please see the corresponding chapter in Radiopaedia.

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