Choroid Plexus Carcinoma

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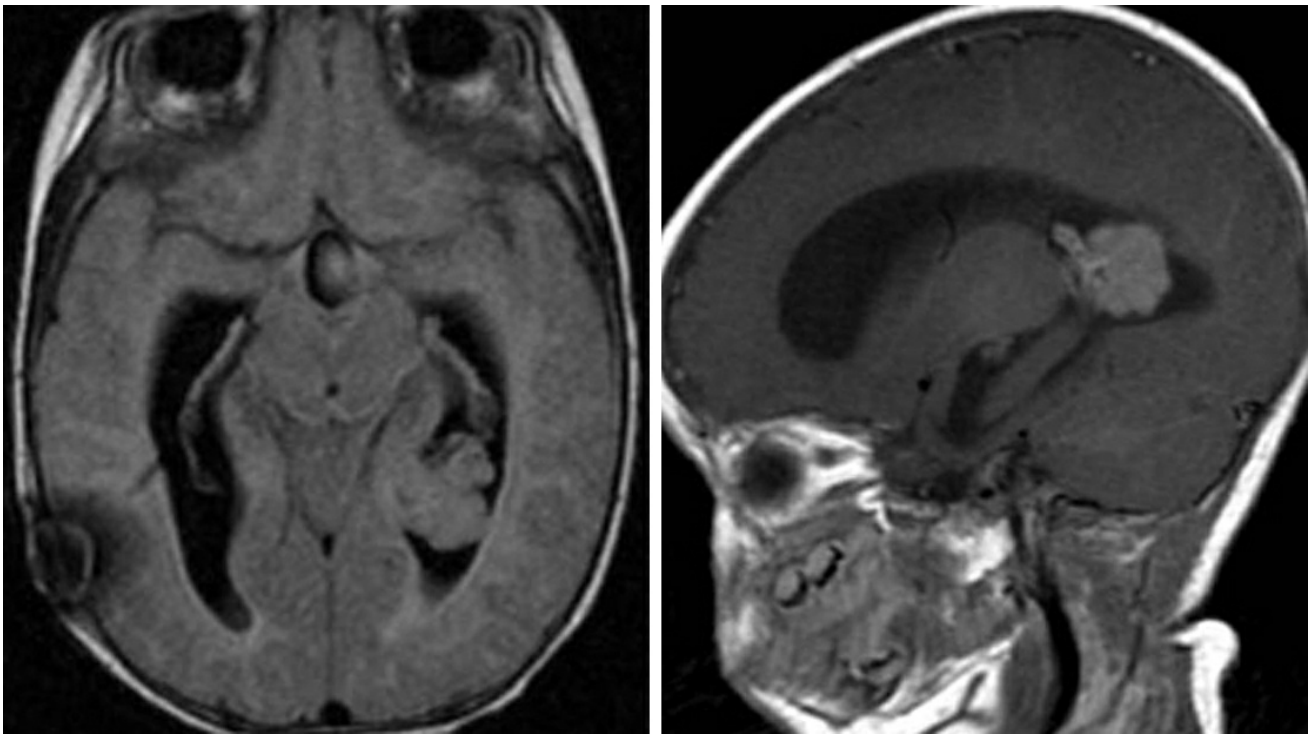

Figure 1: Axial FLAIR (left) and sagittal T1WI postcontrast (right) demonstrate a lobulated intraventricular enhancing tumor. This choroid plexus carcinoma is indistinguishable from choroid plexus papilloma given its lack of invasive features.

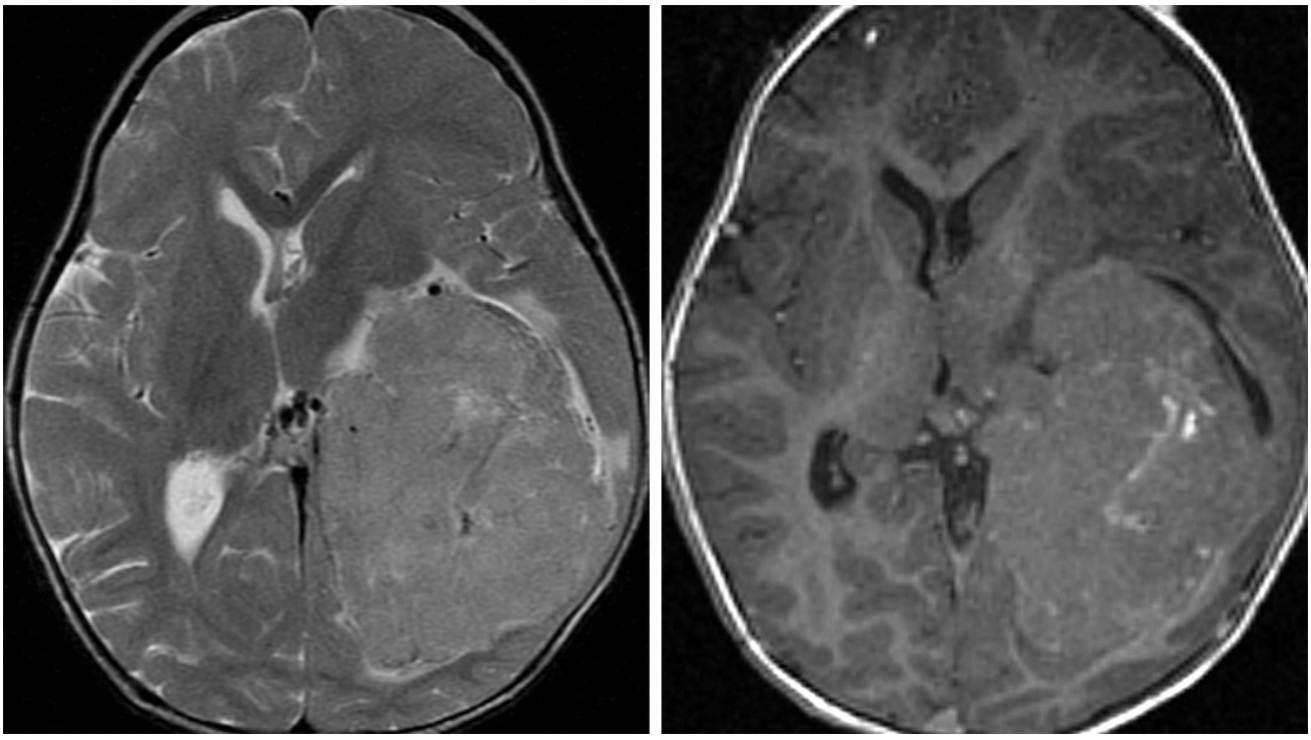

Figure 2: This choroid plexus carcinoma is quite large. As with many tumors of this size, the intraventricular origin of this tumor is difficult to determine. (Left) The hyperintense cleft surrounding CSF on T2WI can give a clue. (Right) The tumor demonstrates less enhancement on postcontrast T1WI than is usually present in choroid plexus tumors.

BASIC DESCRIPTION

- Malignant, rapidly growing intraventricular tumor arising from choroid plexus epithelium
- Less common than choroid plexus papilloma (CPP)

PATHOLOGY

- WHO grade III
- Hypercellularity, pleomorphism, and increased mitosis are characteristic microscopic features
    - Increased Ki-67 index
- Hemorrhage, cysts, calcification, and necrosis are common
- Invasion of adjacent ependyma
- Arises from malignant degeneration of CPP (10%–20% of CPPs)
- High association with simian virus 40 (SV40)

- Association with Li-Fraumeni and Aicardi syndromes

CLINICAL FEATURES

- Commonly afflicts infants and children (majority are <2 years old)
- Presenting signs/symptoms often related to increased intracranial pressure secondary to cerebrospinal fluid (CSF) overproduction/obstruction and decreased CSF resorption
    - Nausea, vomiting, headaches
    - Focal neurologic deficits
- Treatment
    - Gross-total resection followed by chemotherapy
    - ±Radiation after chemotherapy
- Poorer prognosis than for CPP: 5-year survival rate, 30% to 50% after resection
    - Ependymal invasion and CSF dissemination are poor prognostic findings

IMAGING FEATURES

- General
    - Lobulated or irregular, enhancing intraventricular mass with ependymal invasion
        - Lateral ventricle most common location
    - ±Hemorrhage, cysts, calcification, and necrosis common
    - Can be indistinguishable from CPP radiographically
- CT
    - Isodense to hyperdense
    - ±Calcification, hydrocephalus
    - Avid heterogeneous enhancement ± CSF dissemination on contrast-enhanced CT
- MRI
    - T1WI: isointense to hypointense
    - T2WI: variable, heterogeneous signal intensity due to cysts,

necrosis, blood, and calcification

- FLAIR: heterogeneous signal intensity, periventricular bright signal suggests invasion and/or transependymal CSF flow from hydrocephalus
- T2*/GRE/SWI: black signal blooming secondary to calcification or hemosiderin deposition
- DWI: solid tumor components show restricted diffusion
- T1WI+C: avid heterogeneous enhancement ± CSF dissemination
- MRS: elevated Cho and lactate, absent NAA

IMAGING RECOMMENDATIONS

- MRI with contrast, include both brain and spine because of the risk of CSF dissemination

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

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