Subependymal Giant Cell Astrocytoma

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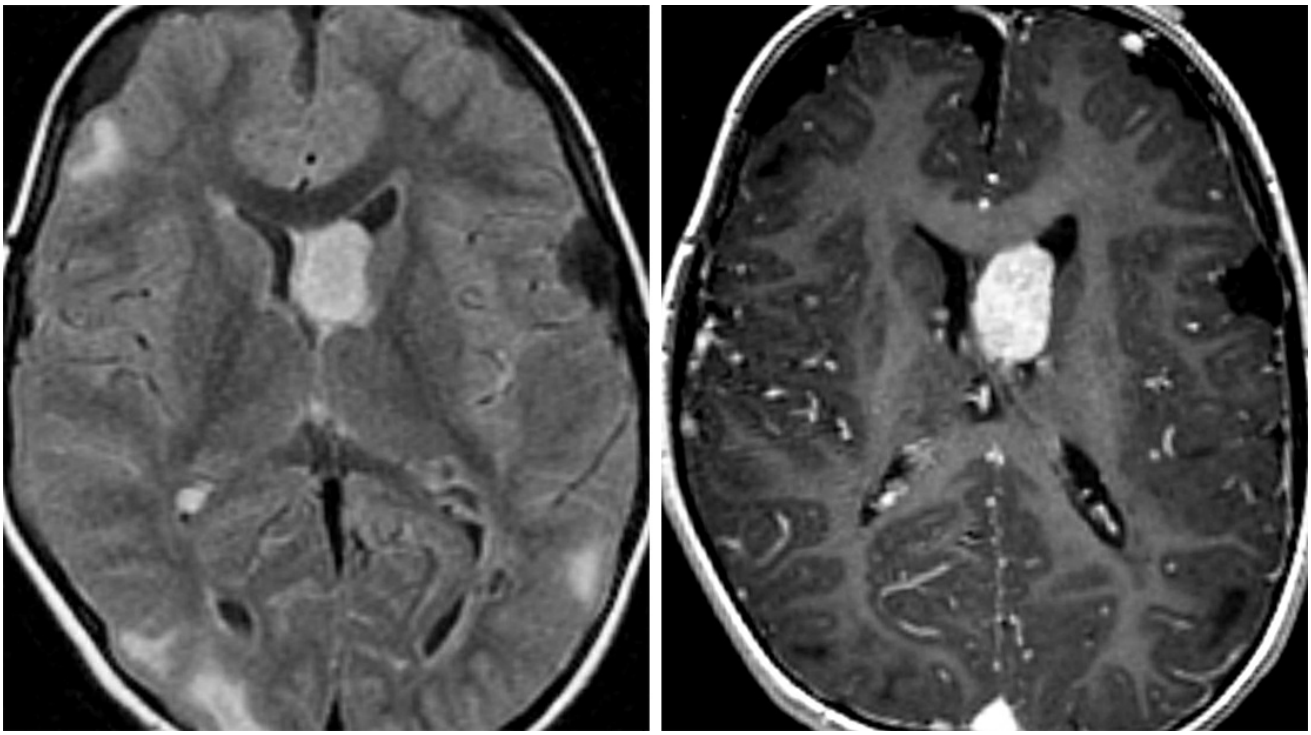

Figure 1: This subependymal giant-cell astrocytoma (SEGA) is present in its typical location at the foramen of Monro. This lesion is larger than should be seen for other subependymal nodules in tuberous sclerosis. (Left) A FLAIR image demonstrates multiple subcortical hyperintense tubers. A hyperintense right posterior periventricular nodule is also visible. Other calcified subependymal nodules are less evident. (Right) Postcontrast T1WI demonstrates the avid enhancement typical of SEGAs.

BASIC DESCRIPTION

- Benign tumor of neuroglial origin classically arising in patients with tuberous sclerosis complex (TSC)
- Slow-growing tumor arising in region of germinal matrix, usually near the foramen of Monro, where it can obstruct ventricular outflow, resulting in noncommunicating hydrocephalus

PATHOLOGY

- WHO grade I
- May arise from subependymal nodules (SENs) near germinal matrix
    - Represents a neuroglial migration abnormality
- Most common are CNS tumors in patients with TSC
    - Uncommonly arises in the absence of TSC
- Similar histology to SENs with diagnosis of SEGA based on tumor size and growth
- Autosomal dominant inheritance or *de novo* mutations of TSC1 and TSC2 genes

CLINICAL FEATURES

- Afflicts children and young adults with TSC (mean age, 11 years)
- Presenting symptoms
    - Obstructive hydrocephalus
    - Headache, nausea, vomiting from increased intracranial pressure
    - Rarely, spontaneous tumoral hemorrhage with intraventricular extension
    - Worsening seizures/epilepsy
- Good prognosis/curable with total surgical resection
- Treatment: rapamycin and/or surgical resection

IMAGING FINDINGS

- General
    - Slowly enlarging, avidly enhancing mass arising near the lateral ventricles/foramen of Monro in patients with TSC
    - Well-defined tumor margins, often lobulated or “frond-like”
    - Variable size

    - Additional findings of TSC (cortical tubers/dysplasias, SENs)
- CT
    - Heterogeneously hypodense to isodense to gray matter
    - May show calcification, hydrocephalus
    - Strong but heterogeneous enhancement on contrast-enhanced CT imaging
- MRI
    - T1WI: hypointense to isointense to gray matter; areas of calcification can appear hyperintense
    - T2WI: heterogeneously isointense to hyperintense; calcification appearing hypointense
    - FLAIR: heterogeneously hyperintense, periventricular hyperintensity secondary to acute hydrocephalus and transependymal flow of cerebrospinal fluid
    - T2*/GRE/SWI: black susceptibility artifact from foci of calcification
    - T1WI+C: avid enhancement
    - DWI: relatively reduced diffusion compared with TSC hamartomas

IMAGING RECOMMENDATIONS

- MRI with contrast, imaging surveillance of enlarging SENs/SEGA

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

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