Intracranial neurenteric cysts

General information

Rare, most common in posterior fossa. Initially, may be difficult to rule out metastasis from an extremely well-differentiated primary adenocarcinoma of unknown origin (absence of progressive disease suggests NEC).

Locations:

1. posterior fossa

a) cerebellopontine angle (CPA): usually intradural, extraaxial (case report of extradural lesion with bone destruction)

b) in midline anterior to brainstem

c) cisterna magna

2. supratentorial: only 15 case reports as of 2004.

Locations: suprasellar (possible confusion with Rathke's cleft cyst), frontal lobe intraparenchymal, quadrigeminal plate region, duralbased extra-axial. Source of endoderm is controversial since the primitive foregut extends cranially only to the midbrain.

Theory: colloid cysts, Rathke cleft cysts, and supratentorial NECs may all arise from remnants of Seesel's pouch, a transient endodermally derived diverticulum of the cranial end of the embryonic foregut.

Histology

Most are simple cysts lined by cuboidal-columnar epithelium and mucin-secreting goblet cells. Less common types of epithelium described include stratified squamous and pseudostratified columnar, and ciliated epithelial cells. Mesodermal components may be present, including smooth muscle and adipose tissue, and some have called these teratomatous cysts, not to be confused with teratomas, which are true germinal cell neoplasms. Maybe histologically identical to colloid cysts.

Clinical

Most commonly present during the first decade of life.

Pain or myelopathy from the intraspinal mass are the most common presentations in older children and adults. Neonates and young children may present with cardiorespiratory compromise from an intrathoracic mass, or with cervical spinal cord compression. Meningitis may occur from the fistulous tract, especially in newborns and infants.

Imaging

Intracranial NEC:

● CT: usually low density, nonenhancing

● T1WI MRI: isointense or slightly hyperintense to CSF (may be hyperintense if there are blood products). T2WI isointense to CSF. Nonenhancing

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Treatment

Spinal NEC

Surgical removal usually reverses the symptoms. Recurrence is uncommon with complete removal of cyst wall.

Intracranial NEC

Capsule adherent to brainstem may prevent complete resection, which predisposes to delayed recurrence. Apparently successful treatment by evacuation of contents and marsupialization has been reported (5 cases, mean follow-up: 5 yrs). Incomplete removal requires long-term follow-up. Hydrocephalus is shunted if indicated.