Intracranial teratoma

They are considered intracranial germ cell tumors, and are comprised of cells originating from at least two and usually all three embryonic layers: ectoderm, mesoderm and endoderm. The histological subtype may not necessarily determine the biological behavior.

Epidemiology

Intracranial teratomas are nongerminomatous germ cell tumors and they account for 0.3 to 0.9% of all intracranial tumors.

Classification

They can be divided into two broad categories, intra- and extra-axial, which differ in epidemiology and clinical presentation. Another method of classifying an intracranial teratoma is as mature teratoma, immature teratoma, and mature with malignant transformation.

Epidemiology

Encephalic teratomas are rare and account for 2 to 5% of infant teratomas and 0.3 to 0.6% of all ventricular tumors. Teratomas may occur at any age, but with a higher incidence rate in younger patients.

The incidence rate in males is marginally higher than that in females. Encephalic teratomas always occur in the midline, where they are typically detected in the pineal and suprasellar regions and rarely in the posterior cranial fossa.

see Pineal teratoma.

see Posterior fossa teratoma.

see Suprasellar Teratoma.

Brain tumors presenting during the first year of life are a different subset of tumors than that presenting later in childhood. In a busy neurosurgical unit in a children’s hospital, they represented ≈ 8% of children admitted with brain tumors, an average of only ≈ 3 admissions per year.

90 % of brain tumors in neonates are of neuroectodermal origin, teratoma being the most common. Some of these tumors may be congenital.

Clinical features

Obstructive hydrocephalus is a common complication of teratomas.

This disease is difficult to diagnose due to its rarity. Furthermore, encephalic teratomas are occasionally misdiagnosed as highly malgnant tumors. For this reason, the family members of patients reject treatment and infant patients may lose the chance to be cured of obstructive hydrocephalus. An accurate pre-operative diagnosis is important and patients are usually in an extremely dangerous condition when they are admitted to hospital. As such, effective treatment strategies create conditions that promote the post-operative recovery of patients and lay the foundation of the desired long-term prognosis.

Intracranial teratomas exhibit different clinical features according to the growth zones. In the majority of infant patients, the main clinical feature of these tumors is intracranial hypertension. Intracranial teratomas are commonly accompanied by symptoms that include obstructive hydrocephalus, which results in headaches, vomiting, papilledema, outreach paralysis and progressive increases in head size. A small number of infant patients also experience epileptic attacks, even when they are in a coma, which may be associated with the intracranial hypertension and brainstem compression caused by the tumor. Infant patients with posterior cranial fossa teratomas do not exhibit dystaxia since their motor function is not yet fully developed. The reason that a medical consultation is required for these infant patients is typically the presentation of hydrocephalus or epilepsy.

Clinical presentation Clinical presentation varies according to whether they are intra- or extra-axial.

Intra-axial Intra-axial teratomas typically present either antenatally or in the newborn period. They are large tumors that increase head circumference and therefore often present with difficulty in childbirth. They tend to occur more commonly supratentorially.

Extra-axial Extra-axial teratomas usually present in childhood or early adulthood and are typically smaller. They most often arise in the pineal or suprasellar regions, and present due to the mass effect: obstructive hydrocephalus due to impingement on the midbrain, Parinaud syndrome, optic chiasm compression, etc.

Diagnosis

Intracranial teratoma diagnosis.

Differential diagnosis

A meaningful differential depends to a degree on location:

intra-axial

embryonal tumors with multilayered rosettes (ETMR)

atypical rhabdoid/teratoid tumor (ATRT): older age group

choroid plexus carcinoma

extra-axial

intracranial lipoma: fatty components only

intracranial dermoid: more mature tissue

craniopharyngioma: particularly for suprasellar lesions

other pineal region tumors