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Intracranial dermoid cyst

J.Sales-Llopis; J. Abarca-Olivas; I.Verdú-Martinez

Neurosurgery Department, University General Hospital of Alicante, Foundation for the Promotion of Health and Biomedical Research in the Valencian Region (FISABIO), Alicante, Spain

Intracranial dermoid cysts generally occur along the midline and are derived from the trapped somatic ectoderm during embryological development during third to fifth week of gestation.

Epidemiology

The tumors typically arise in infants to young adults because of their congenital origin ^{1) 2) 3)}. Intracranial dermoid cyst are very rare, constituting less than 1% of intracranial tumors ⁴⁾, and are relatively rare in middle-aged or older people ⁵⁾.

Many reports have mentioned the intradural posterior fossa and the midline as the preferential localization of these tumors ^{6) 7)}. In contrast, extradural dermoid cysts are a much rarer entity ^{8) 9)}.

see Parasellar dermoid cyst.

see Posterior fossa dermoid cyst.

see Asterional dermoid cyst.

Pathology

Dermoid cysts are thought to occur as a developmental anomaly in which embryonic ectoderm is trapped in the closing neural tube between the 5th-6th weeks of gestation.

Dermoid cysts, like epidermoid cysts, are lined by stratified squamous epithelium. Unlike epidermoid cysts, however, they also have epidermal appendages such as hair follicles, sweat and sebaceous glands. The latter handles the secretion of sebum that imparts the characteristic appearance of these lesions on CT and MRI.

A common misconception is that dermoid cysts contain adipose tissue. This is not the case, as lipocytes are mesodermal in origin, and dermoid cysts (by definition) are purely ectodermal. A dermoid cyst with adipose tissue would be a teratoma.

Clinical features

Associated dermal sinuses cause earlier onset of clinical symptoms such as infection ¹⁰⁾. Other common symptoms including headaches, seizures, and chemical meningitis, and visual disturbances occur late in the clinical course because of its slow-growing nature ^{11) 12) 13)}.

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Many intracranial dermoid cysts are asymptomatic and only found incidentally. Often there is a long history of vague symptoms, with headache being a prominent feature

In case of rupture (spontaneous, traumatic, or iatrogenic (at resection)) leakage of sebum into the subarachnoid space results in an aseptic chemical meningitis.

The presentation is variable, ranging from a headache, to seizures, vasospasm and even death ¹⁴).

Diagnosis

Occasionally, dermoid tumors are incidentally discovered on computed tomography (CT) of the brain or magnetic resonance imaging (MRI) following unrelated clinical complaints. They are also discovered during radiologic investigations of unexplained headaches, seizures, and rarely olfactory delusions.

On imaging, they are usually well-defined lobulated midline masses that have low attenuation (fat density) on CT and hypersignal on T1-weighted MRI images. Typically they do not enhance after contrast administration.

Although dermoid cysts are pathognomonic in appearance on a CT examination, the MRI is also of value in helping to understand the effect of extension and pressure of the mass. DWI is also important for support of the diagnosis and patient follow-up.

Radiograph

Historically, when skull x-rays were routinely used in the assessment of suspected intracranial pathology, a focal lucency due to the fatty sebum

CT

Typically dermoid cysts appear as well defined low attenuating (fat density) lobulated masses. Calcifications may be present in the wall. Enhancement is uncommon, and if present should at most be a thin peripheral rim.

Very rarely they demonstrate hyperdensity thought to be due to a combination of saponification, microcalcification and blood products. This is usually the case when present in the posterior fossa, although why this is the case is not certain.

MRI

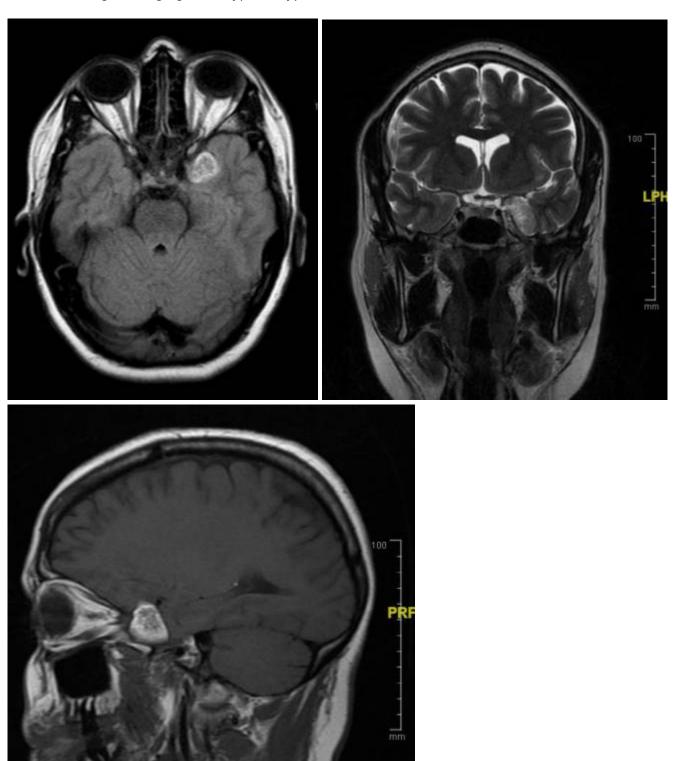
Unlike intracranial lipomas that follow fat density on all sequences, intracranial dermoids have more variable signal characteristics:

T1 typically hyperintense (due to cholesterol components) droplets in the subarachnoid space may be visible if rupture has occurred

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T1 C+ (Gd): generally do not enhance extensive pial enhancement may be present in chemical meningitis caused by ruptured cysts

T2: variable signal ranging from hypo to hyperintense.



Left parasellar extraaxial lesion 2.2 x 1.9 x 1.5 cm without evidence of contrast uptake.

Slight mass effect on the anterior aspect of the left temporal lobe.

Differential diagnosis

Epidermoid cysts at one end (containing only desquamated squamous epithelium) and teratomas at the other (containing essentially any kind of tissue from all three embryonic tissue layers).

Intracranial lipoma: homogeneous fat attenuation/signal intensity, chemical shift artefact

Intracranial teratoma: immature, usually occur in the pineal region

Craniopharyngioma most are strikingly hyperintense on T2, most enhance strongly

Treatment

Can be surgically excised and provided complete excision is achieved recurrence is uncommon. Sometimes due to local adhesion of the capsule to vital structures, incomplete excision must be performed. Recurrent growth, in either case, is slow ¹⁵⁾.

Complications

Spontaneous rupture of dermoid tumor is a potentially serious complication that can lead to meningitis, seizures, cerebral ischemia and hydrocephalus.

Rupture of these benign lesions occurs in only a small percentage of patients, and usually occurs spontaneously $^{16)}$ $^{17)}$

Traumatic rupture of an intracranial dermoid cyst is an exceedingly rare event, with only three cases reported in the literature to date ^{18) 19) 20)}.

Extremely rare malignant transformation into squamous cell carcinoma has been reported ²¹⁾.

Case reports

Intracranial dermoid cyst case reports.

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