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. Intracranial dermoid cyst are very rare, constituting less than 1% of intracranial tumors, and are relatively rare in middle-aged or older people.

Many reports have mentioned the intradural posterior fossa and the midline as the preferential localization of these tumors. In contrast, extradural dermoid cysts are a much rarer entity.

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. Other common symptoms including headaches, seizures, and chemical meningitis, and visual disturbances occur late in the clinical course because of its slow-growing nature.

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.
**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
- 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.

**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.

Traumatic rupture of an intracranial dermoid cyst is an exceedingly rare event, with only three cases reported in the literature to date.

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

**Case reports**

Borni et al. from the Department of Neurosurgery - UHC Habib Bourguiba -Sfax (Tunisia), presented a rare case of a spontaneously ruptured intracranial dermoid cyst in a 32-year-old man presenting as new onset epileptic seizures due to chemical meningitis caused by dissemination of fat or lipid droplets.

2013

A patient complaining of vertigo caused by spontaneous rupture of dermoid cyst, preoperatively diagnosed by CT and MRI. Cranial CT revealed a dense fatty lesion adjacent to the posterolateral parasellar region on the left with multiple small, dense fat droplets scattered in the subarachnoid space corresponding to a dermoid cyst rupture. Cranial MRI sections revealed a lesion with mixed-signal-intensity and multiple hyperintense droplets scattered through the cerebellar surface on the left. No enhancement was found on axial T1-weighted MRI after intravenous Gadolinium administration. Diffusion weighted image (DWI) and apparent diffusion coefficient map studies exhibited explicit restricted diffusion.

Many studies and literature case reports concerning the rupture of dermoid cyst have been reported. However, multimodal imaging of this rare pathology in the same patient is uncommon. 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.

A 47-year-old female presented to the hospital with a ruptured intracranial dermoid cyst following a mild head injury. The ruptured cyst contents were disseminated into the subarachnoid and intraventricular compartments, resulting in an obstructive hydrocephalus. After medical stabilization,
she underwent gross total resection of the cyst using combined transsylvian, transcortical-transventricular, and sub-frontal approaches. A ventriculo-peritoneal shunt was eventually also needed.

Traumatic rupture of an intracranial dermoid cyst is an extremely rare event and this is only the fourth such case reported in the literature. The authors presume that this rupture occurs due to sudden shifts in the cyst sac, which is adherent to some partially mobile intracranial contents. Although computed tomography (CT) is often adequate in making a diagnosis of this entity, magnetic resonance imaging (MRI) provides complete characterization of the extent of lipid dissemination, and is essential for operative planning. Intravenous steroids at presentation are helpful in managing the aseptic meningitis associated with rupture. Complete surgical resection is the goal, but must be weighed against the risk for injury to nearby vital structures. Hydrocephalus should be managed promptly, and patients should be monitored for it closely in the perioperative period. Even though the recurrence rate with subtotal resection is extremely rare, follow up should be done routinely.

References


