Rare disease

'Teeth in the brain' – a case of giant intracranial mature cystic teratoma

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Correspondence to Mr Chandrasekaran Kaliaperumal, ckaliaperumal@gmail.com**Summary**

The authors describe a case of a giant intracranial mature cystic teratoma in a 16-year-old girl presenting acutely with a severe headache, vomiting and a complex generalised seizure with a background history of intermittent headaches for 3 years. CT and MRI brain demonstrated a ruptured large cystic teratoma encapsulating two large teeth within the diffusely dense fatty heterogeneous lesion. Surgical debulking of the cyst was performed and the calcific remnants were left behind owing to dense adhesion to the brain. The procedure was complicated by postoperative hydrocephalus and needed a ventriculoperitoneal shunt. She is currently asymptomatic and undergoing rehabilitation.

BACKGROUND

Giant intracranial mature cystic teratoma is a rare phenomenon. Even more unusual is the occurrence of asymptomatic rupture of such cysts in young individuals. Small cysts can be observed unless they show signs of enlargement along with symptoms of raised intracranial pressure. Surgical excision remains the main modality of treatment however the contents in the cyst could be densely adherent to brain parenchyma making complete excision difficult.

CASE PRESENTATION

A 16-year-old young girl with a 3-year history of 'migraines', presented with an acutely progressively worsening temporo-occipital headaches associated with vomiting, dizziness and disorientation. This was then followed by collapse, eye rolling, jerking of all four limbs and urinary incontinence. She denied any photophobia, neck stiffness or limb weakness.

On clinical examination she was alert, orientated in time, place and person, with a Glasgow Coma Scale (GCS) of 15/15 and neurological examination revealed no neurological deficit. Visual acuity, fields and optic discs were all normal, indicating no ocular involvement. Neuro cognitive assessment was normal. She was noted to be of short stature for her age. Her medical history included febrile convulsions.

INVESTIGATIONS

Haematological investigations revealed a raised white cell count of 14.6×10^9 cells/l with predominant neutrophils (12.9×10^9 cells/l). Inflammatory markers, urea and electrolytes were otherwise normal. Pituitary function hormones revealed a very low morning cortisol level (57 nmol/l) and decrease in thyroid stimulating hormone (0.28 μ IU/ml). Free T4 levels, luteinising hormone, follicle stimulating hormone and prolactin levels were normal.

CT brain showed a large (5.6×9.3 cm) midline intraventricular partially calcified and predominantly fatty lesion with associated hydrocephalus. There were scattered areas

of fatty foci throughout the cisterns and subarachnoid spaces and a fat fluid level within the lateral and fourth ventricles. The calcifications were seen to represent teeth within the lesion. A subsequent MRI confirmed the findings and was highly suggestive of a ruptured dermoid cyst (figures 1–4). Chest x-ray and cervical spine x-ray showed no evidence of any other cystic lesions.

DIFFERENTIAL DIAGNOSIS

- ▶ Ruptured dermoid cyst (benign cystic teratoma)
- ▶ Epidermoid cyst.

Figure 1 Axial fluid attenuated inversion recovery MRI demonstrating large intraventricular mass with teeth and fat filling the lateral ventricles superiorly.

Figure 2 T2 MRI demonstrating the hypointensities in the cyst (teeth).

TREATMENT

She underwent a craniotomy via transcallosal route and debulking of the dermoid cyst. Perioperative findings were indeed consistent with a calcified lump within which a tooth was visible. Soft material was successfully removed, leaving the calcified portion behind. Samples were sent for pathology, which confirmed the diagnosis of a ruptured benign cystic teratoma (dermoid cyst). The cyst was shown to display a large range of tissue components, including skin, sebaceous glands, apocrine glands and hairs. Additionally, fat, smooth muscle, nervous tissue and respiratory type epithelium were present.

OUTCOME AND FOLLOW-UP

Initial postop recovery was complicated by the development of intraoperative diabetes insipidus and hyperglycaemia, from which she made an uneventful recovery. Two days postoperatively she developed secondary hydrocephalus (figure 5) for which she required bilateral ventriculo-peritoneal shunt placement. Currently she is asymptomatic, GCS 15/15, and continues to improve towards her baseline. She requires some assistance with mobility and activities of daily living has some difficulty with short-term memory and is mildly dysphasic with a MMSE of 24/30. She was listed for rehabilitation.

Long-term outcome

Six months postoperatively her short-term memory has improved, however she has occasional difficulty in and the dysphasia has significantly improved. She is currently able to mobilise independently. Input from physiotherapy and occupational therapy have improved her overall postoperative state with a current MMSE of 28/30. She is for a 6

Figure 3 T1 postcontrast MRI with a non-enhancing large cyst.

monthly follow-up and due for an MRI brain in 6 months to assess the status of the cyst and ventricles.

DISCUSSION

Intracranial mature cystic teratoma or dermoid cysts are rare lesions comprising of 0.04%–0.25% of all intracranial tumours.¹ Certain features make this presentation unique. First, rupture was evident preoperatively and the patient had no symptoms of cyst rupture. Despite this, the patient remained well and was amenable to surgical intervention suggesting the possibility of asymptomatic cyst rupture. Her symptom at presentation may well be due to increase in the size of the cyst secondary to raised intracranial pressure. Cyst content initiates an inflammatory process causing meningeal irritation and may cause changes in peripheral blood picture. One may argue that it could be the cyst rupture secondary to increase in size, which led to her presentation, however there was no evidence to support this clinically as she lacked meningeal signs and with a normal erythrocyte sedimentation rate and C reactive protein. Second, the size of the teratoma following rupture was 5.6×9.3 cm and this may have been much more before rupture. This also emphasises the point that these tumours can grow to an enormous size in the midline without causing any cognitive or hormonal changes. Third, the technical difficulty encountered in the removal of the cyst contents (teeth) pointing to the fact that though benign in nature, these can be adherent to the brain causing radical excision a challenge.

Aetio-pathogenesis and literature review

During neural tube closure, weeks 3 to 4 of embryonic development, tissue implantation leads to cyst formation. This includes entrapment of ectodermal and mesenchymal cells.² Dermoid cysts are most often found at the midline, posterior fossa or at third ventricle.^{3 4} Clinical presentation varies depending on the cyst location, acting as a space-

Figure 4 Sagittal MRI demonstrating a large heterogeneous cyst with hyperintensity in the frontal horns of the lateral ventricle.

Figure 5 Postoperative CT brain with residual calcific cyst content and hydrocephalus.

occupying lesion. Headache is the most common symptom (32.6%), followed by seizures (26.5%), then cerebral ischaemia with sensory and/or motor deficit (16.3%) and aseptic meningitis (8.2%).⁵ Our patient had a history of migraine and this may have been a symptom of intermittent raised intracranial pressure.

Dermoid cysts may contain hair follicles, sebaceous and/or sweat glands. The presence of these structures

Figure 6 CT brain postventriculoperitoneal shunt insertion demonstrating residual calcified component and collapsed ventricles.

within the cyst helps to differentiate it from epidermoid cysts.⁶ Rupture of a dermoid cyst is rare and most often occurs spontaneously. Hormonal changes at certain ages are believed to lead to spontaneous rupture by causing rapid enlargement of the cyst.⁷ Photophobia, right trigeminal nerve-type pain, nausea, vomiting and confusion are described as features of rupture.^{8–10} The increase in size is due to a build up of desquamation products and sebaceous secretions inside the cystic cavity.¹¹ In our case, the patient was 16 years of age when she presented to us and there may be a possible hormonal influence attributing to change in her cyst size and subsequent rupture.

Features diagnostic of rupture on neuroimaging consist of diffuse fat droplets in the subarachnoid space and ventricles,⁶ which was very evident in the preoperative MRI, and CT brain in our case. MRI has been shown to be more sensitive than CT in the detection of a dermoid cyst, due to the higher contrast resolution, multi-planar imaging and the lack of bone artefacts.¹²

Where the cyst is intact, surgical removal of the tumour capsule as well as intracystic contents and dissection from adjacent neurovascular structures is an appropriate treatment.¹¹ Postoperative scan depicts satisfactory excision of the cyst (figure 6). Following rupture; however, dissemination of fat droplets is usually too extensive to allow for complete removal. In such cases medical management of patients is appropriate.⁶ Follow-up of cases describing features of rupture, has shown good response to treatment, both conservative and surgical.^{8 13 14}

Early radical excision may be the treatment of choice for better prognosis with this rare syndrome.⁹ Treatment of intracranial cystic teratomas is often difficult. This is because of the heterogeneity of the tumour cells posing

difficulty during surgery as described above. The most important factor with regard to outcome is histological diagnosis of the teratoma subtype. This helps ensure proper therapeutic protocols are adhered to for treatment.¹⁰

Learning points

- ▶ Giant intracranial mature cystic teratoma is a rare condition.
- ▶ Asymptomatic dermoid cyst rupture can occur with or without signs of inflammation (chemical meningitis).
- ▶ Radical surgical excision is recommended and may prove beneficial in patients with symptoms.
- ▶ Calcified intracystic components may pose a technical challenge and may have to be left alone to avoid complications.
- ▶ Postoperative hydrocephalus needs cerebrospinal fluid diversion in the form of VP shunt and patients need long-term follow-up.

Competing interests None.

Patient consent Obtained.

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