

CASE REPORT

Spontaneous Rupture of Intra Cranial Dermoid Cyst: A Case Report

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ABSTRACT

Intracranial dermoid cysts are benign, slow growing, congenital lesions that comprise less than 1% of all intracranial lesions. They are usually asymptomatic unless they get ruptured, either spontaneously or after head injury. We present a case of a ruptured intracranial dermoid cyst, at sellar/suprasellar region, in a middle aged male with multiple known comorbidities. He presented in Accident and Emergency Department with severe headache, behavioral changes and vomiting. Radiological investigation were carried out showing ruptured intracranial dermoid cyst with dissemination of its contents into the ventricles and the sub arachnoid space causing hydrocephalus.

Key words: Ruptured, Intra Cranial, Dermoid Cyst, Sellar/Suprasellar.

INTRODUCTION

Intracranial dermoids are uncommon congenital lesions. They have accounted for less than 1 % of all intracranial masses.¹ These are known to originate from ectodermal inclusions of the primitive pluripotent cells due to defects in the closure of the neural tube at around 3rd to 5th week of gestation.² They are slow growing due to the active production of hair and oil from the internal dermal elements. After rupture of these cysts, the fatty content of the cyst can disperse into the subarachnoid space and find it in any of the cisterns and ventricles. The symptoms of these midline structures are variable with headache and seizure being the most common presentation.⁵ Occasionally, they are picked up incidentally when computed tomography (CT) or magnetic resonance imaging (MRI) brain is done for some other unrelated condition.⁷ Once considered fatal, such a benign lesion can now be dealt surgically.

CASE REPORT

A 56 years old Saudi male patient, known case of diabetes mellitus, hypertension, hypothyroidism with

hepatitis B and C, presented to Accident & Emergency Department with a five day history of headache and behavioral disturbance and one day history of multiple vomiting. He had been a chronic smoker with addiction to opium as well. He was brought in a state of confusion with GCS 14/15 that dropped to 7/15 within two hours of his arrival.

CT scan brain was carried out revealing a hypodense sellar mass (-30 to -40 HU) with extension to suprasellar region measuring 20x32x40 mm in anteroposterior, transverse and craniocaudal dimensions respectively (Figure 1).

This mass was causing pressure on the 3rd ventricle with obstructive hydrocephalus. There were also scattered hypodense droplets within ventricular system and subarachnoid spaces, highly suggestive of a ruptured dermoid cyst. The patient was thereafter taken to the operation theatre for External Ventricular Drainage (EVD), as a result of which his neurological status improved and he started obeying commands. Later MRI brain was done that revealed additional imaging features characteristic of a ruptured suprasellar dermoid cyst apart from resolving



Fig. 1: CT scan brain (Axial Image) showing a hypodense sellar mass with scattered hypodense droplets within ventricular system.

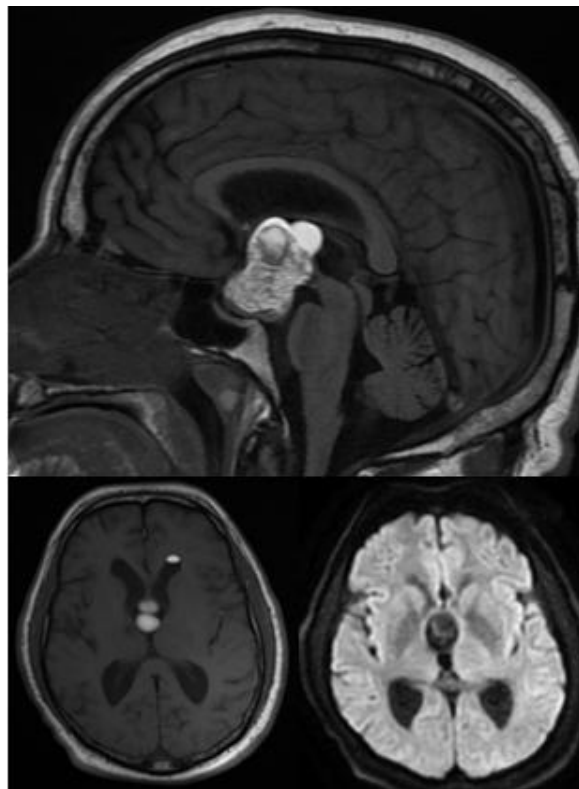


Fig. 2: MRI brain T1WI (Axial and Sagittal) having a heterogeneous hyperintense mass in the sella extending up to suprasellar region and DWI image showing restricted diffusion.

hydrocephalus, with heterogeneous hyperintense signals on T1-weighted image (T1WI) and T2-weighted image (T2WI) and restricted diffusion on diffusion weighted image (DWI) (Figure 2).

Three days after the placement of EVD, definitive surgery was performed and the patient underwent craniotomy for excision of the ruptured dermoid cyst. Per operatively, a right fronto temporal craniotomy flap was elevated for exposure. The dura was then opened along the sphenoid ridge for skull base visualization, exposing the extra axial lesion with a clear fat plane between the tumor and the brain parenchyma. The mass was too large for en bloc resection so it was incised and found to be amorphous and amenable to suction. Histopathology evaluation of the submitted tissue confirmed the diagnosis of a cystic dermoid tumor.

The patient was extubated the 2nd post op day with marked neurological recovery from surgical decompression. Postoperative CT scan Brain showed complete removal of tumor with post-operative changes (Figure 3). Follow-up MRI Brain at 6 weeks confirmed the complete removal of dermoid cyst but with persistent intra ventricular and subarachnoid fat foci.

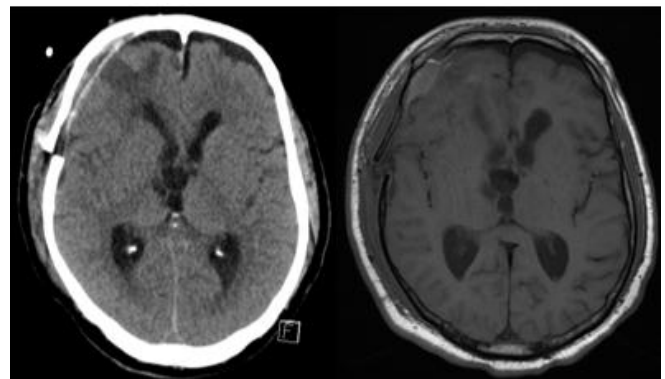


Fig. 3: Postoperative CT Scan and MRI brain (Axial Images) showing complete removal of tumor with post-operative changes.

DISCUSSION

An intracranial dermoid cyst is a benign lesion. It accounts for less than 1% of all the intracranial lesions.¹ This makes only 0.04%-0.06% of the primary intracranial tumors. These cysts are congenital and arise from the inclusion of ectodermal cells at the time of closure of neural groove, between the 3rd and 5th week of gestation, and carries a varying amount of

ectodermal derivatives.^{2,3} These mostly include apocrine, sweat, and sebaceous cysts as well as hair follicles, keratinized squamous epithelium, and possibly teeth.

Dermoid cysts are extra axial, well circumscribed, thick walled, containing thick, viscous greenish-brown fluid comprising of lipid metabolites and cholesterol. Such lesions are not to be confused with an **epidermoid cyst**, which contains **only squamous epithelium**. In contrast to epidermoids, **dermoid cysts** are usually present in the midline. Other differentials include arachnoid cyst and cystic Craniopharyngioma.

These lesions are mostly located infratentorially, either in cavity of 4th ventricle or in vermis.³ Supratentorial dermoid cysts are less common, mostly seen along the parasellar cisterns and in the fronto-basal locations.⁴ Cranial abnormalities such as bone defects and dermal sinus are not associated with supratentorial dermoids.

Supratentorial dermoids are often present in the second or third decades of life. Those located in the posterior fossa typically present in the first decade of life as a consequence of mass effect which they exert on the fourth ventricle with resulting hydrocephalus. However, the mean age of patients at presentation is fifteen years. There is a slight male predominance.

Although its rupture is usually spontaneous, it may occur iatrogenically during surgery or even after closed head injury. It has been hypothesized that the rupture is secondary to rapid enlargement of the cyst due to age-dependent hormonal changes.⁵ If ruptured, the contents may spill into the sub arachnoid space and/or ventricles.

The clinical presentation with dermoid cyst is highly variable, with headache and seizures being the predominant symptoms.⁵ It may present with hydrocephalus and rarely olfactory delusions. Neuro-behavioral symptoms have been reported in the literature.⁶ It can also present itself with symptoms pertaining to its locations, such as hearing loss, tinnitus, trigeminal neuralgias, hormonal changes, diplopia, blurred vision and visual field defects.

Cysts with an acute rupture, either spontaneous or iatrogenic, may cause severe headache and seizures. This is due to aseptic chemical meningitis from the disseminated cholesterol debris which has a reported incidence of up to 7%.⁴ Subsequently cerebral vasospasm ensues which may lead to ischemic events, and if left untreated, infarction and even death can ensue. Rarely the cyst ruptures without any neurological signs or symptoms. The persistence of fat

droplets in the subarachnoid spaces after rupture of the cyst wall may remain asymptomatic for years. Occasionally these cysts are picked up incidentally on brain computed tomography (CT) or magnetic resonance imaging (MRI) done for otherwise unrelated complaints.⁷

Since the era of diagnostic tools has revolutionized the dynamics of modern medicine, therefore, early diagnosis and appropriate management of the ruptured dermoid cyst is the corner stone in dictating the disease progression and its best possible outcome. At times small, asymptomatic, intact and incidentally picked up dermoid cyst may be followed up radiologically, since they grow slowly due to the active production of hair and oils from the internal dermal elements.⁸

On imaging studies, these lesions have pathognomic features. The wall of the cyst may calcify, becoming hyper dense on the CT scan brain, and occasionally enhances partially following administration of CT-iodinated contrast material. On MRI scan they are hyperintense on T1WI; but, may appear heterogeneous with minimal enhancement because of the presence of calcifications, hair follicles, epithelial debris and sebaceous secretion. They appear heterogeneous on T2WI. If the fat content of the cyst is relatively low, then the cyst will reveal cerebrospinal fluid (CSF) like signal intensity. In such cases, fluid attenuation inversion recovery (FLAIR) images will make fat appear hyperintense on a background of the suppressed fluid signal.

On plain CT ruptured dermoid cysts show hypodense fat droplets scattered in the subarachnoid spaces or in the ventricles, even floating within their independent portions with no enhancement on post-contrast studies.⁴ On MRI with gadolinium contrast chemical meningitis is detected as Hyper-intense pain and ventricular ependymal enhancement.^{7,8}

The final confirmation of the lesion is based on histopathology diagnosis. Rupture of the cyst with dissemination of the contents and progressive neurological deterioration warrants early surgical intervention. Even un-ruptured symptomatic dermoids require early surgical removal. Although the extent of resection is debated in terms of location of the lesion and postoperative neurological complications, gross total resection should be the aim of surgical decompression. The type of approach depends upon the site, location and extent of the disease. Recently endoscopic endonasal transsphenoidal approach for the removal of the suprasellar cysts have been reported to

decompress the optic pathways.

Meningitis and transient cranial nerve palsies are the most common postoperative complications being stated in literature after dermoid cyst excision, due to close proximity and adherence of the cyst wall to nearby structures.⁹ Hence, a subtotal excision should be considered in these cases, due to the low recurrence reported in such instances.⁹

Recurrence is rare, but is more common with retained portions of tumor wall because the active living part of the tumor resides in the capsule.⁹ Patients with subtotal resection of ruptured dermoid cysts should be followed up with the help of clinical examinations and serial MRI scans to detect possible recurrence or malignant transformation into squamous cell carcinoma.^{7,8,10} The resorption of the subarachnoid fat is variable and is reported to be present even after 6 years post-surgery.

CONCLUSION

Dermoid cyst is an unusual congenital disease with a wide spectrum of symptoms. An early radiological investigation will assist in timely diagnosis and prompt management. This is considered wise if the CT or MRI brain shows a ruptured cyst with disseminated disease and hydrocephalus. If symptomatic, then surgical resection, even subtotal excision, avoids potential complications and mandates early recovery. Close clinical follow up with serial imaging necessitates rare recurrences.

DISCLOSURE

The authors declare no conflicting interests, support or funding from any drug company.

Additional Information

Disclosures: Authors report no conflict of interest.

Human Subjects: Consent was obtained by all participants in this study.

Conflicts of Interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following:

Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.

Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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