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CASE REPORT

Ruptured Suprasellar Dermoid Cyst presenting with Chemical Meningitis and Blindness

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Abstract

Background: Intracranial dermoid cysts (ICDC) are rare benign lesions. They typically originate from the ectoderm and contain dermal elements like hair follicles, sweat glands, sebaceous glands, and sometimes teeth. Ruptured suprasellar dermoid cysts with intracranial dissemination are extremely rare. **Case presentation:** We report one such patient who presented with headache and blindness, imaging suggestive of ruptured intracranial dermoid cyst. We performed a pterional craniotomy and excised the tumor and the diagnosis was consistent with dermoid cyst on histopathology. **Conclusion:** Rare cases should be reported for further scientific advancements and a better understanding of physicians in the future if they encounter.

Keywords: dermoid cyst, chemical meningitis, blindness, suprasellar

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Introduction

Dermoid cysts, also called mature cystic teratoma, are congenital benign tumors arising from embryonic ectodermal cells and contain dermal elements like retained hair follicles, sweat glands, sebaceous glands, and sometimes teeth. The incidence of ICDC is less than 1% of all intracranial tumours [1]. ICDC are typically seen in the suprasellar region, Sylvian fissure, and posterior fossa including cerebellopontine angle or within the fourth ventricle. Extracranial dermoid cysts are seen in the orbit and spine. Malignant transformation to squamous cell carcinoma is extremely rare [2]. ICDC usually presents with headache, seizures, visual loss, cerebral ischemia with neurodeficit, meningitis when ruptured, obstructive hydrocephalus, and lower cranial nerve palsy, as per their locations [3]. Craniotomy and excision of the lesion with a capsule is the treatment of choice [4]. We report a case of a middle-aged gentleman with ruptured suprasellar dermoid cyst presenting with headache, blindness, and aseptic meningitis.

Case Report

A 49-year-old male patient presented with headache for one month and progressive drowsiness and disorientation for one week before his presentation, without any history of seizures or vomiting. He had impaired vision in his left eye since childhood. On examination, there was no focal motor or sensory deficit. He was confused but obeying commands, pupils were equal and reacting to light and accommodation. Fundus examination showed primary optic atrophy in his left eye. NCCT scan of the patient showed an extra-axial fat density lesion in the suprasellar and right frontal region (Figure 1) with fat attenuation in the subarachnoid spaces and areas of calcification around the lesion. The lesion measured 65 mm x 52 mm x 45 mm (AP x TR x CC).

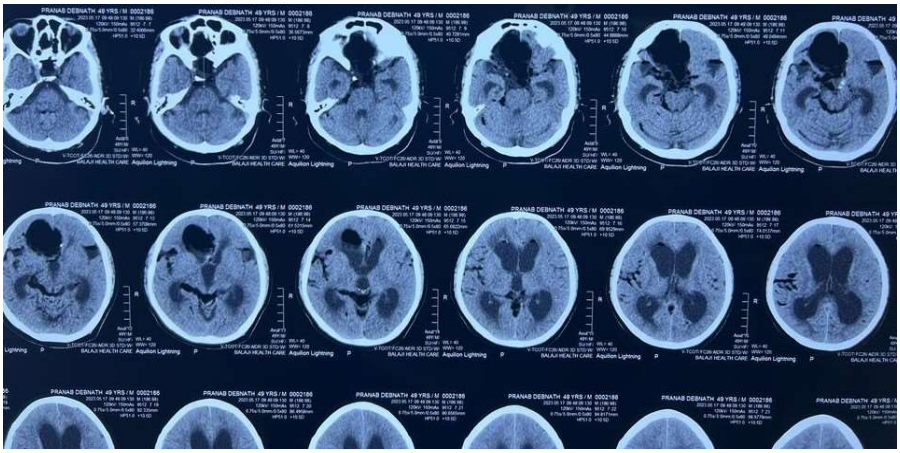
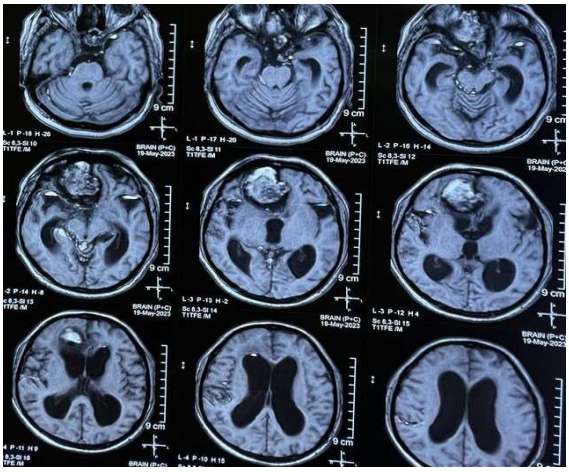
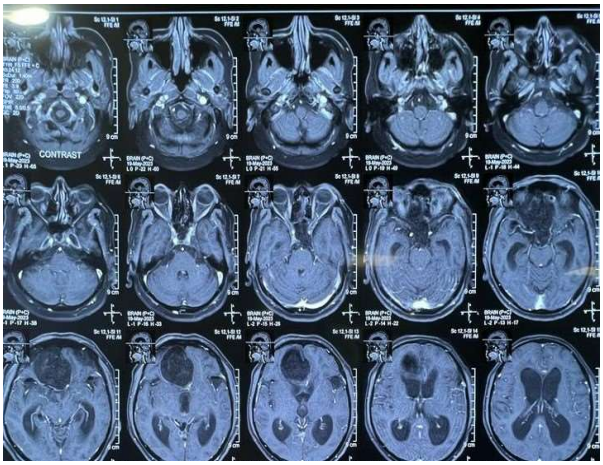


Figure 1. NCCT of brain showing extra-axial fat density in the suprasellar and right frontal region with fat attenuation in the subarachnoid spaces, areas of calcification around the lesion, and obstructive hydrocephalus.

MRI Brain with contrast (Figures 2 and 3) revealed a fairly marginated lesion in T1 sequence & T2W hyperintense lesion, in midline suprasellar and right basi-frontal region, with patchy restricted diffusion, minimal post-contrast enhancement.





Figures 2 and 3. MRI showing a fairly marginated T1 hyperintense lesion, in midline suprasellar and right basi-frontal region, minimal post-contrast enhancement.

Multiple T1 & T2W hyperintensities were seen in both lateral ventricles, quadrigeminal cistern, Sylvian cisterns, and basal cisterns which showed blooming on gradient echo. Radiologically it favoured the diagnosis of a ruptured intracranial dermoid cyst (Figure 4).

After obtaining proper written consent from the patient, we performed a right pterional craniotomy, with EVD inserted through Paine's point. After wide splitting of Sylvian fissure and basal cistern, a large dermoid cyst was removed from the subarachnoid space. The tumor capsule was adherent to branches of the middle cerebral artery, right optic nerve, and optic chiasm. Post-operative recovery was uneventful. Histopathology confirmed a Dermoid cyst. His vision improved significantly at the 3-month follow-up-up (Figures 5 and 6).

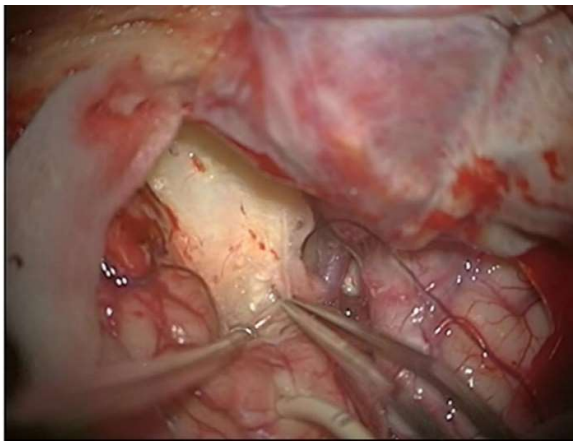


Figure 4. Intraop image showing under microscope after right pterional craniotomy and sylvian fissure dissection, lesion can be seen (yellowish).

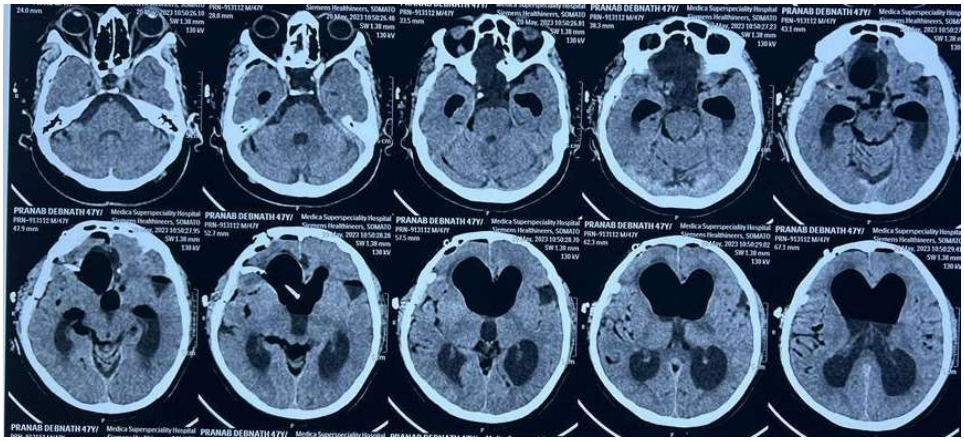


Figure 5. NCCT brain showing post op changes with pneumocephalus, EVD tip in situ.

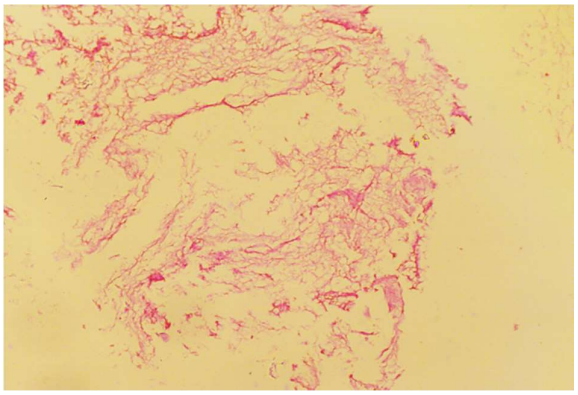


Figure 6. Microscopy (10X magnification) on H&E staining section showing keratinous debris with no lining squamous epithelium

Discussion

Dermoid cysts are congenital benign tumors arising from ectodermal cells during 3rd to 5th week of embryogenesis at the time of neural tube closure5. They contain apocrine glands, sweat glands, sebaceous glands, squamous epithelium, hair follicles, and sometimes teeth6. Epidermoid cysts, different from dermoid cysts, have only squamous epithelial lining. ICDC are more commonly infratentorial in location, in the midline, in the vermis, or inside the 4th ventricle. The supratentorial location of dermoid cysts is less common. Other rare sites are suprasellar and pineal regions.

ICDC accounts for 0.04% to 0.25% of all intracranial tumors4. The duration of presentation of ICDC is months to years. Suprasellar dermoid cysts present early because of the mass effect on the optic apparatus. Usual presentations of unruptured ICDC are due to mass effect on neurovascular structures mainly optic apparatus, presenting late due to their slow-growing nature. Ruptured ICDC presents with headache, seizures, cerebral ischemia with focal deficits, hydrocephalus,

vasospasm resulting in infarcts, fat embolism, visual deficits, aseptic meningitis, increased intracranial pressure (ICP), chronic granulomatous arachnoiditis and can be fatal. Headache is the most common symptom (32.6%) [7]. Rupture occurs spontaneously in the majority of cases but head trauma can also be a cause. Intraventricular fat can cause motion-dependent intermittent raised ICP by blocking the CSF pathways, resulting in acute hydrocephalus8.

Radiological features of ICDC are very characteristic. Non-contrast CT scan of the brain shows mixed density of ICDCs with hyperdense areas of calcification and hypodense areas of fat density (negative Hounsfield units). They do not enhance post-contrast CT. T1W MRI typically shows high signal intensity within the lesion (and in the subarachnoid space in case of ruptured ICDC). FLAIR shows subtle sulcal hyperintensity. T2WI, GRE, and SWI show sulcal "bloom". In T1 Post Gadolinium MRI, lesions commonly do not enhance but can show leptomeningeal reaction and contrast enhancement, if complicated by chemical meningitis [9].

Close differentials of ICDCs are epidermoid cysts, cystic craniopharyngiomas, and arachnoid cysts.

Surgical excision with copious lavage of subarachnoid space is the mainstay of treatment. In many cases, total removal is not possible when the capsule is densely adherent to neurovascular structures, in which case subtotal resection is performed to avoid neurovascular complications. There are instances when ruptured ICDCs have been treated with a lumbar drain for prevention of post-operative hydrocephalus [10].

The recurrence following the excision of ICDCs is rare, in contrast to epidermoids that frequently recur.

The prognosis is relatively good if the tumor is completely excised. However, ruptured ICDC can result in many complications and can turn fatal.

Conclusion

Dermoid cysts are congenital benign tumors arising from embryonic ectodermal cells and dermal elements. ICDCs are rare brain tumors and suprasellar/supratentorial are extremely rare locations for ICDC. It usually presents with mass effect on neurovascular structures and compression of optic apparatus. Radiological diagnosis is straightforward and is diagnosed pre-operatively in most of cases, more so if it is ruptured. Treatment is craniotomy and micro-neurosurgical excision with copious irrigation and lavage. We recently encountered one such case.

List of Abbreviations

ICDC – Intracranial dermoid cysts
NCCT – Non contrast Computed Tomography
AP x TR x CC – Anteroposterior x Transverse x Craniocaudal
MRI – Magnetic Resonance Imaging
ICP – Intracranial pressure
CSF – Cerebrospinal fluid
FLAIR – Fluid Attenuated Inversion Recovery sequence

Statements and Declarations

Conflicts of interest

The authors declare that they do not have conflict of interest.

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References

1. Jamjoom AB, Cummins BH. The diagnosis of ruptured intracranial dermoid cysts. Br J Neurosurg. 1989;3(5):609-12.
2. Nickl RC, Nickl V, Schindehütte M, Monoranu CM, Ernestus RI, Löhr M. Malignant transformation of a cerebral dermoid cyst into a squamous cell carcinoma with malignant intraperitoneal spreading along a ventriculoperitoneal shunt: illustrative case. J Neurosurg Case Lessons. 2022 Oct 17;4(16):CASE2254.
3. Furqan M, Akinleye A R, Ravella P Mahendra (May 16, 2013) Ruptured Intracranial Dermoid Cyst: Unusually Rapid Disease Course Leading to a Fatal Outcome . Cureus 5(5): e117.
4. Alam K, Varshney M, Aziz M, Maheshwari V, Haider N, Gaur K, Srivastava VK. Dermoid cyst in brain. BMJ Case Rep. 2011 Oct 4;2011:bcr0520114203.
5. Murrone D, Romanelli B, Ierardi A (2016) Ruptured intracranial dermoid cyst: a case report. Integr Mol Med 3
6. Ramlakhan R, Candy S. Traumatic rupture of an intracranial dermoid cyst. Radiol Case Rep. 2015 Dec 3;10(1):1053.
7. Jacków J, Tse G, Martin A, Sąsiadek M, Romanowski C. Ruptured intracranial dermoid cysts: a pictorial review. Pol J Radiol. 2018 Oct 19;83:e465-e470.
8. Jain, Pooja; Aggarwal, Ankita; Jain, Swarna G; Jain, Savyasachi. Ruptured Intracranial Dermoid Cyst – Unusual Cause of Obstructive Hydrocephalus. Neurology India 70(4):p 1712-1713, Jul–Aug 2022
9. Balasundaram P, Garg A, Prabhakar A, Joseph Devarajan LS, Gaikwad SB, Khanna G. Evolution of epidermoid cyst into dermoid cyst: Embryological explanation and radiological-pathological correlation. Neuroradiol J. 2019 Apr;32(2):92-97.
10. Blitz SE, Bernstock JD, Dmytriw AA, Ditoro DF, Kappel AD, Gormley WB, Peruzzi P. Ruptured Suprasellar Dermoid Cyst Treated With Lumbar Drain to Prevent Postoperative Hydrocephalus: Case Report and Focused Review of Literature. Front Surg. 2021 Aug 11;8:714771.