Ruptured intracranial dermoid cyst manifesting as new onset seizure: a case report

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ABSTRACT

Intracranial dermoid cysts are rare tumors derived from ectopic epithelial cells. They are slow-growing benign entities, but can cause significant morbidity through compression of neurovascular structures and, rarely, rupture into the subarachnoid space. We present a rare case of a spontaneously ruptured intracranial dermoid cyst presenting as new onset seizures due to chemical meningitis caused by dissemination of fat droplets.

CASE REPORT

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A 19-year-old male presented to the ER with a history of 2 episodes of new onset seizures. Both were complex partial seizures involving lower extremity spasms prior to loss of consciousness. On presentation, the patient had a normal physical exam, including a complete neurological and cranial nerve exam.

CT of the brain (Figure 1) revealed a large low-density (-25 HU) suprasellar lesion causing widening of the interpeduncular cistern. Numerous low-density fat droplets were noted in the subarachnoid space.

Subsequent MRI (Figures 2-13) of the brain revealed a non-enhancing suprasellar mass partially encasing the right internal carotid artery. The mass was of high T1 and T2 signal with signal dropout on fat suppression imaging. Scattered droplets with similar signal characteristics were noted in a fronto-temporal spread within the subarachnoid space. The mass was hyperintense on DWI, but demonstrated an ADC that was similar to brain parenchyma (Figures 10-11). In addition, mild leptomeningeal enhancement was noted on the post-contrast images (Figure 13).

Given the clinical history and the MRI demonstration of disseminated particles, a ruptured intracranial mass seemed most likely. The signal characteristics of the mass on T1 and DWI, coupled with the insidious clinical history steered us towards the diagnosis of an intracranial dermoid cyst.

The patient was placed on anticonvulsant therapy, and on follow-up visits at 4 and 10 months he was noted to be neurologically asymptomatic with no further seizures. Followup imaging (Figure 14) remained unchanged.

DISCUSSION

Intracranial dermoid cysts are rare, comprising 0.04-0.7% of intracranial tumors [1]. They are derived from ectopic epithelial cells that are part of the neural tube, which also explains their typical location close to the midline [2]. Although they are benign, slow-growing tumors, they may cause focal neurologic signs through encroachment of neurovascular structures and, rarely, rupture [3]. Dermoid cysts contain lipid material [4], and previous reports have discussed that they may have fatty tissue peripherally and fluid centrally [5]. They can contain hair follicles, sebaceous and sweat glands, and the presence of these structures helps

distinguish a dermoid from the more common epidermoid cysts [6]. They are not true neoplasms, as they enlarge through accumulation of desquamation products and sebaceous secretions inside a cystic cavity [7] rather than via cell division.

Rupture of intracranial dermoid cysts is a rare phenomenon (5 out of 2707, or 0.18% of all new CNS tumors operated on during a 12-year period at a major tertiary care center) [8] and typically spontaneous, although can occur secondary to closed head trauma [9]. The pathophysiology behind spontaneous rupture is not clearly understood, and hypotheses have implicated glandular secretions caused by age-dependent hormones [6] as well as head movements and brain pulsations [10]. The dissemination of intracystic keratin and cholesterol breakdown products [11] following rupture can cause a wide variety of symptoms ranging from headache to hallucinations [12, 13, 14, 15]. Clinical presentation can vary depending on the cyst location, and in one analysis of available case reports by El-Bahy et al. [16] headache was the most common symptom (32.6%), followed by seizures (26.5%), cerebral ischemia with sensory and/or motor hemisyndrome (16.3%), and aseptic meningitis (8.2%). The case presented involved a spontaneously ruptured intracranial dermoid cyst causing new onset seizures due to presumed chemical meningitis and chemical irritation secondary to dissemination of fat droplets.

On CT scans, dermoid cysts can have mixed densities [8], and rarely enhance following contrast administration [17, 18, 19]. The intracystic fat and disseminated fat droplets appears hypodense, whereas calcifications in the wall are hyperdense. Hydrocephalus and fat-fluid level may be present following rupture into the ventricular system. On MRI, dermoid cysts are hyperintense on T1-weighted sequences and variable on T2weighted sequences, although the presence of cholesterol can often make them appear hypointense on T2 as well [17, 20]. Dermoid cysts can be differentiated from epidermoid cysts in that the former demonstrates fat signal on CT and MRI whereas the latter resembles CSF [21]. Although Fluid Attenuated Inversion Recovery (FLAIR) sequences and Diffusion Weighted Imaging (DWI) have been used to distinguish the two entities, dermoid cysts can resemble epidermoids due to their bright signal on DWI [22, 23].

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MRI is more sensitive than CT in the detection of dermoid cysts and due to the higher contrast resolution, the ease of multiplanar imaging and the lack of bone artifacts [24]. Orakcoiglu et. al. [25] in particular emphasizes the importance of using an MRI protocol involving T2-WI, T1-WI, T1 fat saturated-WI, Magnetic Resonance Angiography (MRA) and DWI. Conventional MRI is not very helpful in distinguishing dermoids from other cystic masses such as arachnoid cysts. On DWI, however, the dermoids are hyperintense to brain parenchyma, but demonstrate an ADC that is similar to that of parenchyma and CSF. This is in contrast to arachnoid cysts, which show the opposite pattern (low DWI, but elevated ADC) [26, 27]. Differentiating a dermoid cyst from craniopharyngiomas is relatively easier, as the latter enhances strongly on CT [28, 29]. In addition, the craniopharyngioma cyst walls also display strong enhancement on T1-weighted

MRI sequences [30, 31]. Teratomas help distinguish themselves via their calcifications, which are hyperintense on CT [32, 33] and show up as shadowing echogenic foci on ultrasound [34].

Dermoid cysts are benign entities, and have a generally favorable prognosis. Surgery is only indicated in cases where dermoid cysts cause mass effect and serious neurological deficits. In cases where the cyst is intact, the goal is complete surgical removal of the primary tumor capsule and intracystic contents and dissection from adjacent neurovascular structures [7, 10]. Unfortunately, dissemination of fat droplets following rupture is usually too extensive to allow for complete removal. However, Liu et. al. [8] and others [35, 36] have shown that long-term monitoring with serial MRI scans and clinical examinations of patients with extensive disseminated fat particles has not demonstrated progression or movement of the fat or new neurological deterioration. In those cases, medical management is indicated for symptom control.

TEACHING POINT

Intracranial dermoid cysts are benign rare slow-growing tumors that, when intact, are of mixed or predominantly low density on CT and hyperintense on MRI T1 with little to no contrast enhancement. The cysts are hyperintense on DWI and isointense to brain parenchyma on ADC, which helps distinguish them from other cystic masses. Upon rupture, however, widespread presence of T1 hyperintense droplets and leptomeningeal enhancement can be noted - making MRI the best imaging modality for diagnosis of this rare entity.

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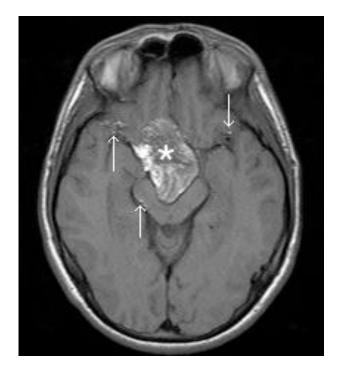


Figure 2: MRI T1 sequence Axial. 19 year old male with ruptured intracranial dermoid cyst. Axial MRI (1.5 Tesla magnet) T1 weighted image demonstrates a suprasellar mass of high T1 signal intensity (asterisk), which causes widening of the interpeduncular cistern. Scattered droplets (arrows) of similar signal characteristics were noted in the subarachnoid space. T1 Axial parameters: TR = 400, TE = 14, Slice Thickness = 5 mm, FOV = 24 X 24 cm, Magnet strength: 1.5 Tesla

FIGURES



Figure 1: Non-Contrast CT. 19 year old male with ruptured intracranial dermoid cyst. Axial non-contrast CT images of the head demonstrate a large low-density suprasellar lesion (asterisk). Numerous low-density fat droplets are seen in the subarachnoid space. Slice Thickness: 5.0, FOV: 24 X 24 cm, W: 80, L: 40, KVP = 120, mAs = 450



Figure 3: MRI T1 sequence Axial. Axial MRI (1.5 Tesla magnet) image of a 19 year old male with ruptured intracranial dermoid cyst. This image demonstrates partial encasement of the right ICA (arrow) by the mass. T1 Axial parameters: TR = 400, TE = 14, Slice Thickness = 5 mm, FOV = 24 X 24 cm, Magnet strength: 1.5 Tesla

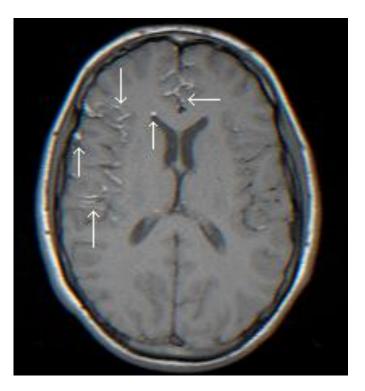


Figure 4: MRI T1 sequence Axial. 19 year old male with ruptured intracranial dermoid cyst. Axial MRI (1.5 Tesla magnet) T1 weighted image demonstrates scattered droplets (arrows) of high T1 signal in the fronto-temporal subarachnoid space. T1 Axial parameters: TR = 400, TE = 14, Slice Thickness = 5 mm, FOV = 24 X 24 cm, Magnet strength: 1.5 Tesla

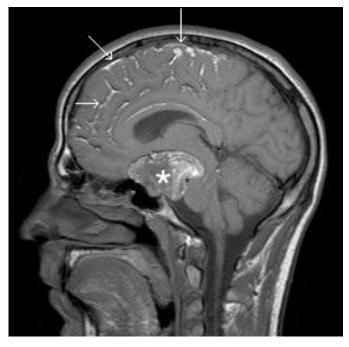


Figure 6: MRI T1 sequence Sagittal. 19 year old male with ruptured intracranial dermoid cyst. Sagittal MRI (1.5 Tesla magnet) T1 weighted image demonstrates a suprasellar mass (asterisk) of high T1 signal and scattered droplets (arrows) throughout the subarachnoid space of similar signal characteristics. T1 Sagittal parameters: TR = 453, TE = 12, Slice Thickness = 5 mm, FOV = 26 X 26 cm, Magnet strength: 1.5 Tesla

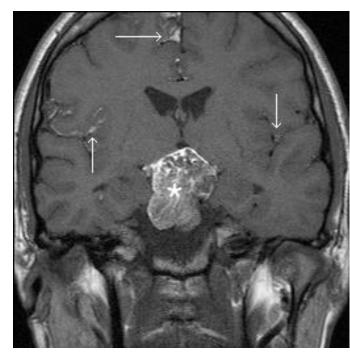


Figure 5: MRI T1 sequence Coronal. 19 year old male with ruptured intracranial dermoid cyst. Coronal MRI (1.5 Tesla magnet) T1 weighted image demonstrate suprasellar mass (asterisk) of high T1 signal. Scattered fat droplets (arrows) are re-identified. T1 Coronal parameters: TR = 446, TE = 13, Slice Thickness = 3 mm, FOV = 18 X 18 cm, Magnet strength: 1.5 Tesla

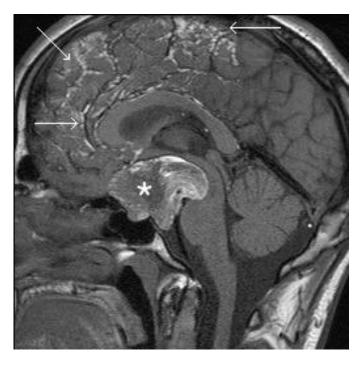


Figure 7: MRI T1 sequence Sagittal. 19 year old male with ruptured intracranial dermoid cyst. Sagittal MRI T1 weighted image demonstrates a suprasellar mass (asterisk) of high T1 signal and scattered droplets (arrows) throughout the subarachnoid space of similar signal characteristics. T1 Sagittal parameters: TR = 453, TE = 12, Slice Thickness = 5 mm, FOV = 26 X 26 cm, Magnet strength: 1.5 Tesla

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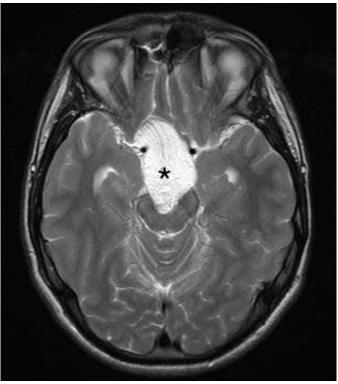


Figure 8: MRI T2 sequence Axial. 19 year old male with ruptured intracranial dermoid cyst. Axial MRI (1.5 Tesla magnet) T2 weighted image demonstrates a suprasellar mass of high T2 signal intensity (asterisk), which causes widening of the interpeduncular cistern. T2 Axial parameters: TR = 4790, TE = 113, Slice Thickness = 5 mm, FOV = 23.8 X 24 cm, Magnet strength: 1.5 Tesla

FLAIR TR = 8480, TE = 98, Slice Thickness = 5 mm, FOV = 23.8 X 24 cm, Magnet strength: 1.5 Tesla

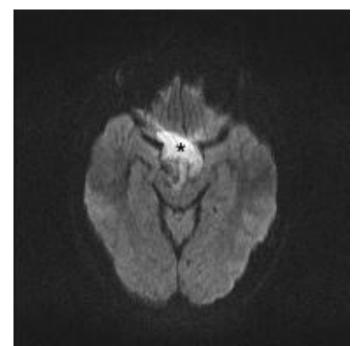


Figure 10: MRI DWI image. 19 year old male with ruptured intracranial dermoid cyst. The mass is denoted by an asterisk. A hyperintense signal is noted in the diffusion weighted images (DWI), whereas the Apparent Diffusion Coefficient (ADC) of the mass is isointense with brain parenchyma (figure 11). TR = 3800, TE = 102, Slice Thickness = 5 mm, FOV = 19.5 X 19.5 cm, Magnet strength: 1.5 Tesla

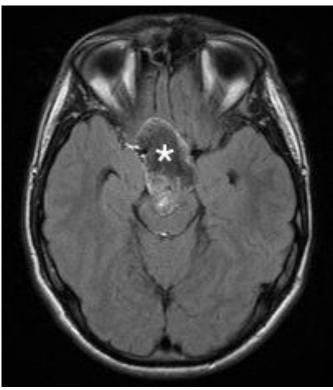


Figure 9: MRI T2 sequence Axial. 19 year old male with ruptured intracranial dermoid cyst. Axial MRI (1.5 Tesla magnet) T2 FLAIR image demonstrates a suprasellar mass of low FLAIR signal intensity (asterisk), which causes widening of the interpeduncular cistern. FLAIR Axial parameters:

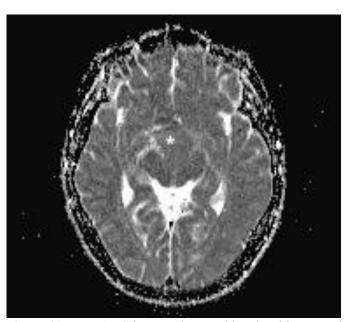


Figure 11: MRI ADC image. 19 year old male with ruptured intracranial dermoid cyst. The mass is denoted by an asterisk. A hyperintense signal is noted in the diffusion weighted images (DWI, figure 10), whereas the Apparent Diffusion Coefficient (ADC) of the mass is isointense with brain parenchyma. TR = 3800, TE = 102, Slice Thickness = 5 mm, FOV = 19.5×19.5 cm, Magnet strength: 1.5 Tesla

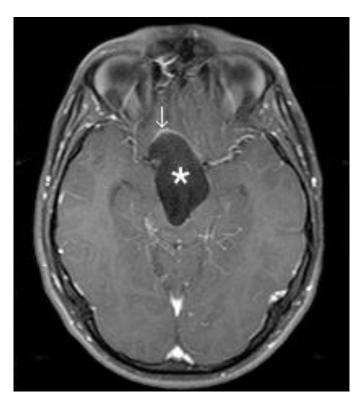
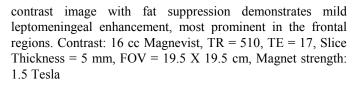


Figure 12: MRI T1 sequence Post Contrast Axial image with fat suppression. 19 year old male with ruptured intracranial dermoid cyst. Axial MRI (1.5 Tesla magnet) T1 weighted post contrast image with fat suppression demonstrates a nonenhancing suprasellar mass (asterisk) with signal dropout on fat suppression imaging. There is mild peripheral enhancement of the mass, as denoted by the arrow. Contrast: 16 cc Magnevist, TR = 417, TE = 17, Slice Thickness = 5 mm, FOV = 19.5 X 19.5 cm, Magnet strength: 1.5 Tesla





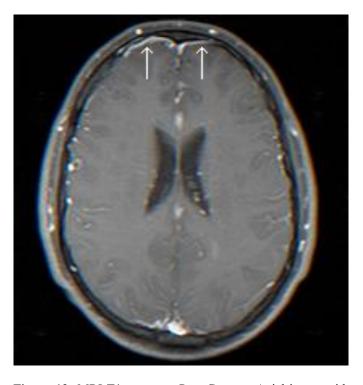


Figure 13: MRI T1 sequence Post Contrast Axial image with fat suppression. 19 year old male with ruptured intracranial dermoid cyst. Axial MRI (1.5 Tesla magnet) T1 weighted post

intracranial dermoid cyst. 4 month followup images. Sagittal MRI (1.5 Tesla magnet) T1 weighted post contrast image again demonstrates suprasellar mass (asterisk) with scattered droplets in the fronto-parietal (arrows) subarachnoid space. This was unchanged from the prior examination. Contrast: 16 cc Magnevist, TR = 435, TE = 15, Slice Thickness = 5 mm, FOV = 19.5 X 19.5 cm, Magnet strength: 1.5 Tesla

Diagnosis	General	СТ	MRI T1	MRI T2	Enhancement	Imaging Recommendation
Dermoid cyst	Benign midline cysts derived from ectodermal tissue. Less than 0.5% of primary intracranial tumors.	Well- circumscribed cystic mass with fat hypodensity, but rare "dense" dermoids can be hyperattenuating. 20% have capsular calcifications. Rupture my cause fat-fluid level within ventricles	Hyperintense fat appearance. Post-rupture droplets and "dense" dermoids very hyperintense	Unruptured: variable, range from hypo to hyperintense. Ruptured: Hyperintense droplets. Fine curvilinear hypointense elements represent hair.	CT: does not generally enhance. T1: Extensive enhancement from chemical meningitis after rupture.	Best imaging tool: MRI, especially with rupture. Can use fat- suppression to confirm diagnosis. Chemical shift- selective sequence useful in detection of tiny droplets.
Epidermoid cyst	Benign congenital inclusion cysts. 0.2- 1.8% of all primary intracranial tumors.	Round/lobulated mass resembling CSF. 10-25% can have calcifications.	75% slightly hyperintense to CSF (with lobulated periphery slightly hyperintense than center). Rarely, can be hypointense to CSF ("black epidermoid")	Isointense (65%) to slightly hyperintense (35%) to CSF.	CT and MRI (T1 sequence): usually none, margin may show minimal enhancement. Enhances after malignant transformation.	Best imaging tools: MRI. Diffusion definitively distinguishes from arachnoid cyst.
Cranio pharyngioma	Benign epithelial tumor derived from Rathke pouch. 1.2-4% of intracranial tumors (most common non- glial pediatric intracranial tumor).	Classic/adamanti nomatous type (more common): 90% iso to hypodense, and 90% have calcifications. Papillary type: Isodense, rare calcification	Signal depends on cystic contents. Classic: hyperintense cyst with heterogeneous nodule. Papillary: isointense solid component.	Cysts show hyperintense signal, while solid component show heterogeneous (iso to hyperintense) signal with calcified portions being hypointense. Hyperintense signal in adjacent brain parenchyma can be due to gliosis or edema from cyst leakage.	CT: 90% show enhancement. T1: solid components enhance heterogeneousl y while cyst walls enhance strongly.	Best imaging tools: MRI (especially sagittal and coronal sequences).
Teratoma	Tridermal midline supratentorial mass. 2-4% of pediatric intracranial tumors	Fat, soft tissue, cystic attenuation. Majority have calcifications	Increased signal from fat. Variable signal from solid tissue.	Soft tissue components iso to hyperintense. FLAIR: decreased signal from cysts, increased signal from solid tissue.	CT and MRI, post contrast: Soft tissue components enhance	Best imaging tool: CT, which shows soft tissue, fat and calcifications. MRI helps characterize relationship to midline structures.

Table 1. Differential diagnosis table for intracranial dermoid cyst

References used for differential table: Craniopharyngioma: 28-31, Teratoma: 32-34, Epidermoid cyst: 37-41

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Etiology	Inclusion of ectodermal tissue during embryological closure of neural tube			
Incidence	Rare (comprise < 0.5% of primary intracranial tumors)			
	4-9 times rarer than epidermoid cysts			
Gender Ratio	Slightly more common in males			
Age Predilection	Generally diagnosed between 30-50 years of age			
Risk Factors	Sporadic, but occurs with Goldenhar syndrome			
Presentation	Most common symptoms: headache, seizure and cerebral ischemia, focal neurological signs due te neurovascular encroachment. In addition, rupture can cause chemical meningitis			
Treatment	Definitive treatment: Surgical excision. However, benign nature, rarity of rupture and possibility of surgical complications and recurrence justifies delaying surgery and opting for regular imaging follow-up instead			
Prognosis	Benign, slow-growing tumors. Rupture (rare) can cause significant morbidity/mortality due to seizure, coma, vasospasm, and infarction. Can undergo malignant transformation into squamous cell carcinoma			
Differential	Epidermoid cyst, craniopharyngioma, teratoma			
Pathologic features	Outer wall of connective tissue, typically squamous epithelium surrounding collection of mixed lipid and sebaceous contents. Can have dermal appendages (hair follicles, sweat glands).			
Imaging findings	<u>CT:</u> Well-circumscribed cystic mass with fat hypodensity, but rare "dense" dermoids can be hyperattenuating. 20% have capsular calcifications. Rupture my cause fat-fluid level within ventricles.			
	MRI, T1 sequence: Hyperintense fat appearance. Post-rupture droplets and "dense" dermoids very hyperintense.			
	<u>MRI, T2 sequence</u> : Unruptured: variable, range from hypo to hyperintense. Ruptured: Hyperintense droplets. Fine curvilinear hypointense elements represent hair.			
	Enhancement:			
	<i>CT</i> : does not generally enhance.			
	T1: Extensive enhancement from chemical meningitis after rupture.			
	DWI: Pronounced hyperintensity on DWI. Demonstrate ADC that is similar to brain parenchyma.			

Table 2. Summary table for intracranial dermoid cyst

ABBREVIATIONS

CT = Computed Tomography MRI = Magnetic Resonance Imaging FLAIR = Fluid Attenuated Inversion Recovery DWI = Diffusion Weighted Imaging MRA = Magnetic Resonance Angiography

KEYWORDS

Adult; Dermoid Cyst/cerebrospinal fluid/diagnosis; Humans; Lipids/*cerebrospinal fluid; Magnetic Resonance Imaging/methods; Male; Rupture, Spontaneous/cerebrospinal fluid/diagnosis; Tomography, X-Ray Computed/methods

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