

A Rare Case of Rathke Cleft Cyst Mimicking a Cavernous Carotid Aneurysm Hypopituitarism

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AUTHORS CONTRIBUTIONS

Evan Hartman wrote entire manuscript. Steve Nelson provided figures and figure legends as well as editing the manuscript.

DISCLOSURES

We have no disclosures to make.

CONSENT

Informed consent was obtained from the patient.

HUMAN AND ANIMAL RIGHTS

No human or animal research subjects were used in this case report.

ABSTRACT

Rathke cleft cysts (RCC) are non-neoplastic masses typically located around the pituitary gland. In many patients, RCCs are asymptomatic and found incidentally via brain imaging or after death via autopsy. Incidental findings are becoming more common with the increase in imaging, however, some RCCs cause symptoms consisting of headaches, visual disturbances, or pituitary dysfunction. Asymptomatic lesions may be followed with serial imaging, but symptomatic lesions are often removed surgically. Due to varying appearance of RCCs on imaging, it can be difficult to differentiate RCCs from other pathology, such as adenomas, until surgery is performed. General imaging characteristics exist that may be more suggestive of RCCs compared to other pathology. Here, we present a rare case of a patient with a large RCC resulting in hypopituitarism. Initial imaging suggested the possibility of a thrombosed cavernous aneurysm which was later ruled out.

CASE REPORT

BACKGROUND

Rathke's cleft cysts (RCC) are benign cysts that develop in the region of the pituitary gland of the brain when Rathke's pouch fails to develop properly [1]. These are non-neoplastic lesions located in sellar and suprasellar regions that are usually small, asymptomatic and incidentally identified on 5-33% of routine autopsies [2]. However, RCCs can increase in size, compressing brain structures which ultimately leads to various symptom development such as visual impairment when compressing the optic chiasm. Surgical intervention serves as the treatment for symptomatic cases whereas conservative management consisting of follow-up imaging is the usual course for asymptomatic RCCs [3]. Here, we present a 62-year-old female with a Rathke cleft cyst causing hypopituitarism that was initially suspected to be a possible thrombosed cavernous aneurysm on imaging.

CASE PRESENTATION

The patient is a 62 y/o female who presented to the ER for altered level of consciousness. CT scan at that time was negative for acute pathology but note was made of a possible peripherally calcified suprasellar aneurysm (Figure 1A,1B). A CT angiogram of the head was recommended for follow up. Head CTA was read to be negative for aneurysm as there was no contrast enhancement within the region of previously described calcifications (Figure 1C,1D). Approximately 16 months later the patient presents with new central hypothyroidism and central adrenal insufficiency. Gonadotropin was also deficient. An MRI sella was ordered to further evaluate for central causes of hypopituitarism. The MRI revealed a tubular hypointense structure on T2 weighted imaging that was also T1 hyperintense and without appreciable post contrast enhancement. There was some speculation that the mass might represent a thrombosed cavernous carotid aneurysm versus Rathke Cleft cyst. A pituitary macroadenoma was felt less likely given the effaced appearance of the adenohypophysis. The patient was referred to a tertiary care center for further evaluation. On Digital

Subtraction Angiography (images not available) there was no direct connection with the vasculature arguing against thrombosed carotid aneurysm. On surgical resection the mass was consistent with Rathke Cleft Cyst.

DISCUSSION

With the increased use of imaging, incidental findings of RCCs have become more frequent because they are often asymptomatic in patients. As the volume and extent of compression increase, patients may become symptomatic. The cystic volume may increase by acute hemorrhage or fluid accumulation causing the cyst to compress on surrounding brain structures [4]. This is represented by the T2 hypointense and T1 hyperintense internal contents in the case presented above. Although symptoms occur in a minority of cases, when present, common symptoms include headache, endocrine abnormalities, and visual field defects [5]. One review including 143 patients described headaches as the most common symptom (27% were acute and 29% were chronic) followed by endocrine dysfunction in 16% of patients and visual defects in 6% of patients [6]. Another review with 258 patients identified RCC manifestation at diagnosis as headaches in 34.9% of patients, endocrine dysfunction in 19.4%, and visual defects in 17.1% [2]. The patient presented in this case presented for altered mental status almost a year and a half before etiologies causing hypopituitarism were explored. Some additional pathologies to include in the differential diagnosis of sellar/suprasellar masses are pituitary apoplexy, craniopharyngioma, dermoid cysts, lipomas, and thrombosed cavernous aneurysms along with arachnoid cysts and cystic pituitary adenomas [2, 7].

When patients are asymptomatic, RCCs may be found incidentally on CT or MR imaging. RCCs on CT may be low or isodense homogenous lesions, but they may also be slightly hyperdense without or with peripheral enhancement that is ring or capsular-like [8]. Although imaging assists in visualizing an RCC, it is not unusual for them to appear with various signals due to differing compositions with a small study stating that 50% of surgically proven RCCs were misdiagnosed as adenomas due to the range of MR signals that RCCs displayed [9,10]. Characteristics more common of RCCs include the presence of intracystic nodules and a midline location whereas pituitary adenomas are more likely to demonstrate fluid-fluid levels, septations, and off midline lesions [9]. RCCs that appear hypointense on T1 and hyperintense on T2 may contain CSF-like fluid while those appearing hyperintense on T1 and hypointense on T2 are more likely composed of mucoid material or blood products, but a homogenous hypointense T2 signal is more indicative of an RCC compared to other pathologies including chordomas, lipomas, and abscesses that appear hyperintense on T2 [8]. These varying signal intensities of RCCs complicate the ability to distinguish from other pathology. This was evident in the case above with initial diagnosis concerning for a possible thrombosed aneurysm.

CT imaging is beneficial for assessing the bony anatomy of the sella and is useful in evaluating osseous erosion that

may be caused by a sellar mass. MRI, however, serves as the gold standard for assessing sellar and parasellar masses [11]. Aneurysms within the internal carotid artery likely appear with a halo appearance on MRI and may also show rim-like enhancement as well as intralesional filling defects that suggest thrombosis [12,13]. The reason for initially suggesting the finding of thrombosed cavernous aneurysm was the tubular nature of the mass as well as the T2 hypointensity within the mass which mimicked a possible flow void, and which was also near the adjacent vasculature suggesting a possible direct connection. Additionally, increased T1 signal within the lesion suggested blood products/thrombus within this tubular appearing structure. The peripheral calcification was originally thought to be vascular further confusing the diagnosis. Finally, a thrombosed cavernous aneurysm might be expected to be relatively hyperdense depending on the age of the thrombus.

Treatment of RCCs varies depending on symptomatology. For asymptomatic patients, conservative management with serial imaging is a viable option. One review noted that RCCs remain stable in about half of cases and roughly 25% progress with conservative management while an equal percentage spontaneously regress on serial imaging follow-up [6]. Treatment for symptomatic patients involves surgical drainage where the cyst wall may or may not be fenestrated, however, this lowers recurrence rates but increases surgical complications [14]. Transsphenoidal surgery is the typical approach, and surgery has shown different efficacies for different symptoms. Visual impairment may improve in 90% or more of patients RCCs, but some literature suggests that anterior pituitary dysfunctions may improve in less than 30% of patients with additional literature suggesting that preoperative dysfunction is permanent and persists after surgery [3, 15]. It is therefore important to discuss the risks, benefits and expected outcomes with the patient prior to surgery.

CONCLUSIONS

Differentiating RCCs from pathologies like adenomas or thrombosed cavernous aneurysms may be difficult to elucidate on imaging given the variable appearance RCCs may have, causing overlap with the imaging findings from other pathologies. General imaging characteristics for RCCs include a midline position and intracystic nodules compared to adenomas which more likely demonstrate a fluid level, septations, and lateral positioning or thrombosed aneurysms that may have a halo appearance on post contrast imaging with the presence of a flow void. In a similar case to that presented above, investigators found what appeared to be a thrombosed aneurysm of the middle cerebral artery by MRI imaging that was later shown to have no direct vascular connection to the middle cerebral artery by DSA and was instead found to be a hemorrhagic cavernoma [16]. This underscores the complexity of neuroimaging and the importance of a broad differential diagnosis when indicated and to exclude possibilities that could lead to catastrophic bleeding at the time of surgery. As in the case presented above, DSA helped to exclude the possibility of a thrombosed aneurysm in favor of the second differential diagnosis which was RCC. This

allowed for the safe resection of this mass and return of pituitary function.

TEACHING POINT

Rathke Cleft Cysts may have a heterogenous appearance on MRI and can easily be misinterpreted as other entities such as thrombosed cavernous aneurysms or adenomas. Additional imaging modalities may be necessary to make this distinction prior to surgical management.

QUESTIONS

Question 1: A patient presents with visual field defects and has a suprasellar lesion visualized on imaging. What should be included on a reasonable differential diagnosis?

- A. Craniopharyngioma (applies)
- B. Rathke Cleft Cyst (applies)
- C. Pituitary adenoma (applies)
- D. Pineal gland tumor
- E. Thrombosed cavernous aneurysm (applies)

Explanation: Pathologies to include in the differential diagnosis of sellar/suprasellar masses are pituitary apoplexy, craniopharyngioma, dermoid cysts, lipomas, thrombosed cavernous aneurysms, arachnoid cysts, and cystic pituitary adenomas.]

A. Craniopharyngioma (applies) [Craniopharyngiomas can be suprasellar located and increase in size compressing the optic chiasm leading to visual field defects.]

B. Rathke Cleft Cyst (applies) [Although often asymptomatic, they can expand in size and cause visual field defects through compression of visual pathway structures.]

C. Pituitary adenoma (applies) [This is a common sellar mass that can extend and compress the optic chiasm.]

D. Pineal gland tumor [These are not typically located in the suprasellar region and often cause symptoms related to obstruction of cerebrospinal fluid.]

E. Thrombosed cavernous aneurysm (applies) [These can mimic sellar or suprasellar masses and cause visual symptoms through compression.]

Question 2: Which imaging characteristic is most helpful if present to make Rathke Cleft Cyst the leading diagnosis?

- A. Spiculated margins and edema
- B. Flow void on T2 imaging
- C. Isointense T1 and T2 image that is solid and enhances
- D. T1 hypointense, T2 hyperintense, and ring enhancement
- E. Heterogenous T1 hyperintense, T2 hypointense, and no enhancement (applies)

Explanation: RCCs can have various imaging findings and overlap with other pathologies. Proteinaceous filled cysts likely appear T1 hyperintense, T2 hypointense while fluid filled cysts commonly appear T1 hypointense and T2 hyperintense.

A. Spiculated margins and edema [These are findings more characteristic of malignant tumors, not benign cysts.]

B. Flow void on T2 imaging [This is indicative of a vascular lesion, not a cyst.]

C. Isointense T1 and T2 image that is solid and enhances [This is more characteristic of a solid tumor and less of a cystic lesion like a RCC.]

D. T1 hypointense, T2 hyperintense, and ring enhancement [This can describe a cystic lesion with inflammatory or neoplastic characteristics such as an abscess but not typically a RCC.]

E. T1 hyperintense, T2 hypointense, and no enhancement (applies) [Proteinaceous fluid within a RCC can produce these characteristics.]

Question 3: What region of the brain are Rathke Cleft Cysts generally located?

- A. Pituitary gland (applies)
- B. Pineal gland
- C. Cerebellopontine angle
- D. Subarachnoid space
- E. Midbrain

Explanation: [Rathke's cleft cysts (RCC) are benign cysts that develop in the region of the pituitary gland of the brain when Rathke's pouch fails to develop properly]

A. Pituitary gland (applies) [RCCs develop from remnants of Rathke's Pouch which gives rise to the anterior pituitary gland.]

B. Pineal gland [The pineal gland is located in the epithalamus, a different anatomical region from the pituitary gland.]

C. Cerebellopontine angle [This is a region located at the base of the brain between the pons and cerebellum.]

D. Subarachnoid space [RCCs are sellar lesions and do not originate from the subarachnoid space.]

E. Midbrain [This is part of the brainstem which does not contain the pituitary gland.]

Question 4: What is the most common presentation for Rathke Cleft Cysts?

- A. Visual field defects
- B. Majority are asymptomatic (applies)
- C. Headaches
- D. Endocrine dysfunctions
- E. Tinnitus

Explanation: [RCCs are usually asymptomatic, but in a minority of cases when symptoms are present, common symptoms include headache, endocrine abnormalities, and visual field defects.]

A. Visual field defects [Visual field defects are a common symptom but RCCs are usually asymptomatic.]

B. Majority are asymptomatic (applies) [RCCs are usually incidental findings on imaging and asymptomatic until their size increases.]

C. Headaches [Headaches are a common symptom but RCCs are usually asymptomatic.]

D. Endocrine dysfunctions [Endocrine dysfunctions are a symptom but RCCs are usually asymptomatic.]

E. Tinnitus [Tinnitus is usually associated with issues related to the inner ear or auditory pathway.]

Question 5: What is the typical management of a symptomatic Rathke Cleft Cyst?

- A. Endovascular coiling
- B. IV steroids
- C. Radiation
- D. Transsphenoidal surgery (applies)
- E. Serial imaging

Explanation: [Transsphenoidal surgery is the typical approach while serial imaging can be used for asymptomatic RCCs.]

A. Endovascular coiling [This is a vascular procedure used for brain aneurysms.]

B. IV steroids [Steroids can be used for some brain pathologies but do not treat the RCC.]

C. Radiation [Radiation is typically used for malignant lesions and not benign lesions like RCCs.]

D. Transsphenoidal surgery [Transsphenoidal surgery is the standard and most common approach to drain the cyst, remove the cyst, and relieve pressure from the cyst.]

E. Serial imaging [This is management that is common for asymptomatic cysts.]

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FIGURES

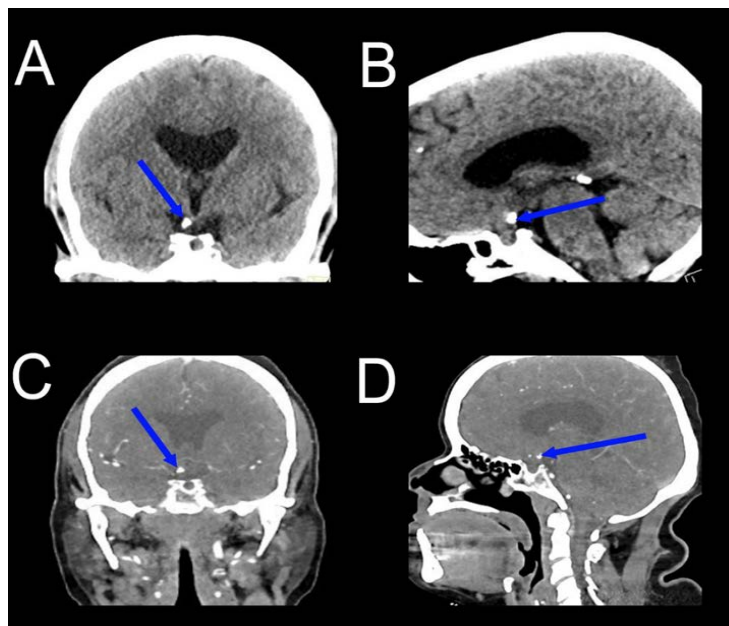


Figure 1: Coronal and Sagittal non-contrast CT of the head (A&B respectively) revealed a suprasellar mass with peripheral coarse calcification (blue arrows). Coronal and Sagittal CT head angiogram (C&D) reveals a non-enhancing sellar and suprasellar mass with peripheral coarse calcification.

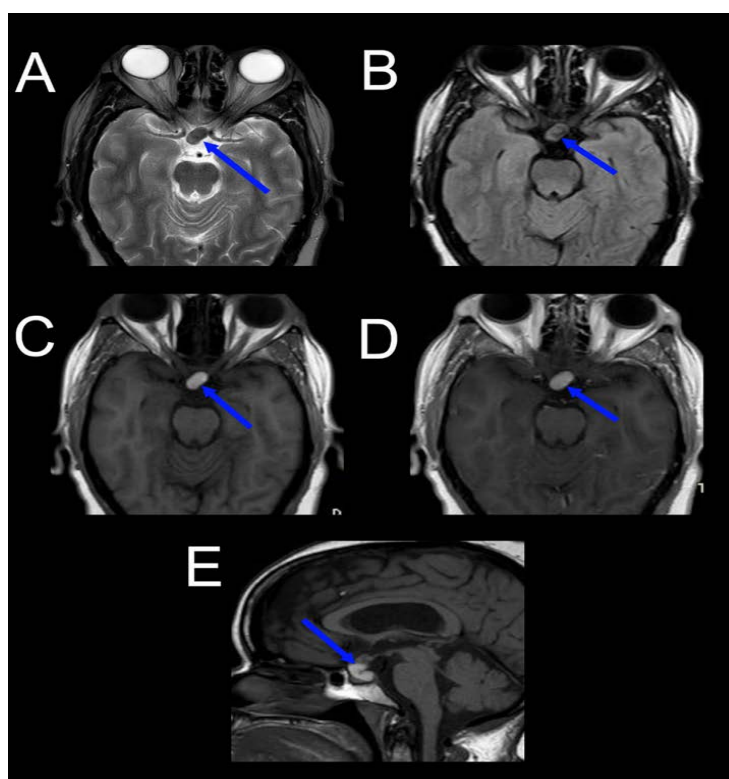


Figure 2: Axial T2 weighted image (A) showing a tubular hypointense suprasellar mass (blue arrow). T2 FLAIR weighted axial image of the mass showing heterogenous hypointense signal of the mass (B). Pre (C) and Post contrast (D) axial T1 weighted images showing mass does not enhance. Sagittal T1 weighted image (E) showing hyperintense signal within the sella/suprasellar mass. The adenohypophysis is compressed anteriorly by the mass. The Neurohypophysis is not visualized.

KEYWORDS

Rathke Cleft Cyst, Thrombosed Cavernous Carotid Aneurysm, Pituitary Macroadenoma, Suprasellar, Hypopituitarism.

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