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MR Imaging Appearance of Ruptured Rathke Cleft Cyst and Associated Bone Marrow Enhancement

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ABSTRACT

SUMMARY: Rathke cleft cysts are common cystic pituitary lesions seen on MR imaging. A subset of Rathke cleft cysts can rupture within the sella and are uncommon. The imaging appearance of a ruptured Rathke cleft cyst has been previously described with nonspecific imaging findings. We present 7 cases of ruptured Rathke cleft cysts and basisphenoid bone marrow enhancement below the sella that could be used to potentially distinguish a ruptured Rathke cleft cyst from other cystic lesions.

ABBREVIATIONS: RCC = Rathke cleft cyst; rRCC = ruptured Rathke cleft cyst

Rathke cleft cysts (RCC) are benign epithelium-lined remnants originating from the craniopharyngeal duct between the anterior and posterior pituitary lobes.¹ RCCs are common, having been found in 11.3% of patients in a large cadaveric study² and are often asymptomatic. When symptomatic, RCCs manifest most frequently with headaches, visual deficits, or hormonal abnormalities,¹ often leading to a contrast-enhanced brain MR imaging. A subset of RCCs can rupture within the sella, and several prior case reports described nonspecific findings on MR imaging in this setting.^{3–8} We present 7 cases of pathology-proved ruptured RCC (rRCC), each of which had basisphenoid bone marrow enhancement.

MATERIALS AND METHODS

An internal database was reviewed from 2016 to 2022 for pathology-proved rRCC. Inclusion criteria were adults (18 years of age or older) with preoperative MR imaging that included fat-saturated postcontrast T1-weighted imaging. Pertinent patient history was reviewed, which included the following: sex, age, presenting symptoms, serum prolactin levels, presumed preoperative diagnosis, surgical approach, and postoperative follow-up.

Preoperative MR imaging was evaluated for lesion size and enhancement of the basisphenoid bone marrow directly below the sella. Additionally, the images were evaluated for lesion location (midline versus off-midline), T2 signal, T1 signal, and the

presence of a T2 dark nodule. The images were reviewed by a neuroradiology instructor (I.T.M.) and a neuroradiologist with 20 years of experience (C.M.G.). Sphenoid sinus pneumatization patterns were evaluated and graded as the following: type 1 conchal, type 2 presellar, type 3 sellar, and type 4 postsellar.⁹

Post hoc analysis was conducted of 7 consecutive patients (2019–2022) with pathology-proved cystic adenoma for enhancing basisphenoid bone marrow. Inclusion criteria matched those of the patients with rRCC; adults (18 years of age or older) with preoperative MRI, including fat-saturated postcontrast T1-weighted MR imaging.

RESULTS

Patients with rRCC

Seven patients with pathology-proved rRCC were included. Patient-specific demographics are listed in Table 1. All 7 patients were women and had an average age of 44.3 years (range, 30–72 years). The median serum prolactin level was 34.9 $\mu\text{g/L}$ (range, 2.7–113.2 $\mu\text{g/L}$). Pituitary adenoma was the presumed preoperative diagnosis in 5 (71.4%) cases. The average maximum lesion diameter was 11.2 mm (range, 8.2–14.0 mm). Three patients had type 2 (presellar) pneumatization of the sphenoid sinus, and 4 patients had type 3 (sellar) pneumatization.⁹ Three patients had focal basisphenoid bone enhancement below the sella (Fig 1), while 4 patients had enhancement that extended diffusely in the axial plane (Figs 2 and 3). Patient-specific imaging findings are listed in Table 2.

Patients with Cystic Pituitary Adenoma

The average age of our patients was 40.9 years (range, 28–62 years). The average maximum adenoma diameter was 20.1 mm (range, 9–44 mm). The median serum prolactin level was

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Table 1: Demographic information of the patients with rRCC proved at surgical resection

Patient	Age (yr)	Sex	Prolactin ($\mu\text{g/L}$)	Presenting Symptom	Preop Diagnosis	Follow-Up
1	51	Female	33.2	HA	Adenoma	No recurrence at 6 mo
2	47	Female	n/a	HA, panhypopituitary	RCC	Recurrence at 3 yr
3	38	Female	2.7	HA	Adenoma	No recurrence at 4 yr
4	39	Female	22.4	Enlarging lesion	RCC	No recurrence at 1 mo
5	30	Female	36.5	Diabetes insipidus, vision changes	Adenoma	No recurrence at 2 mo
6	72	Female	42.4	Fatigue	Adenoma	No recurrence at 2 mo
7	33	Female	113.2	Hyperprolactinemia	Adenoma	No recurrence at 2 mo

Note:—HA indicates headache; Preop, preoperative; n/a, not available.

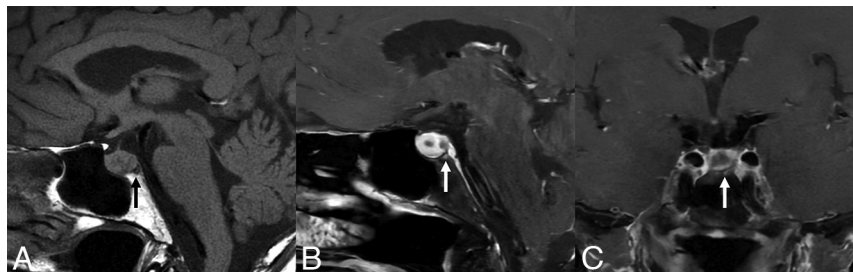


FIG 1. A 72-year-old woman who presented with fatigue and was found to have a bilobed pituitary lesion, preoperatively favored to be a pituitary adenoma. A, Precontrast T1-weighted imaging shows focal hypointense signal in the posterior basisphenoid bone marrow (arrows). Postcontrast imaging shows corresponding enhancement (B and C). Pathology confirmed an rRCC.

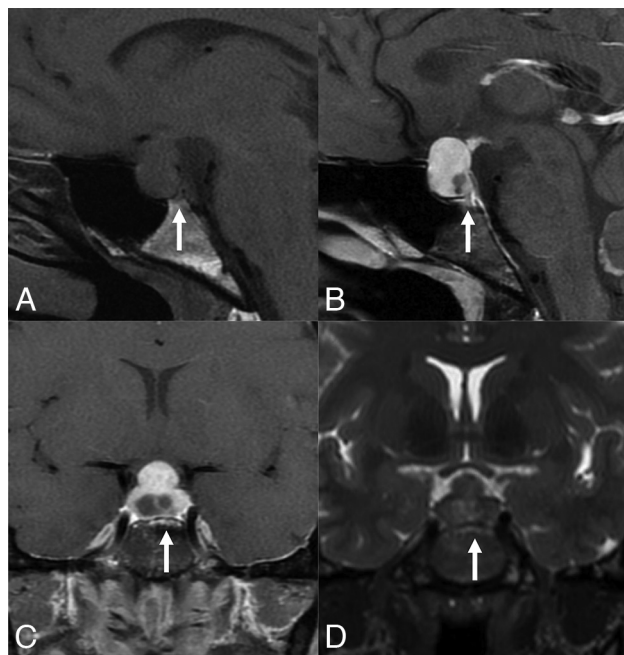


FIG 2. A 38-year-old woman presented with headaches for 6 months. MR imaging shows a sellar/suprasellar mass with a septated cystic lesion in the posterior aspect of the sella, preoperatively favored to represent a pituitary adenoma. Postcontrast MR imaging shows basisphenoid bone marrow (arrows) enhancement (B and C) with corresponding edema (D). There was no intrinsic basisphenoid T1-hyperintense signal on the precontrast imaging (A). Pathology confirmed the cystic lesion to be an rRCC. The enlarged suprasellar lesion was biopsied and found to be mixed inflammatory infiltrate and fibrosis, thought to represent inflammatory hypophysitis secondary to the rRCC.

30.8 $\mu\text{g/L}$ (range, 12.6–167.1 $\mu\text{g/L}$). Six patients had type 3 (sellar) pneumatization of the sphenoid sinus, and 1 patient had type 2 (presellar) pneumatization. None of the 7 patients had enhancing basisphenoid bone marrow below the sella.

DISCUSSION

Our study is the first to describe enhancing basisphenoid bone marrow below the sella as an imaging finding of rRCC. We present 7 cases of rRCC, all of which showed bone marrow enhancement. This finding is potentially important because the leading differential for

an rRCC is cystic pituitary adenoma, which, in our limited experience reported here, does not have basisphenoid bone marrow enhancement.

RCCs arising from failed regression of the cleft between the adeno- and neurohypophysis are frequently incidental and asymptomatic findings.¹⁰ RCCs can become symptomatic and present with headaches, visual deficits, or hormonal abnormalities and have an overlapping imaging appearance with cystic adenoma.¹¹ Park et al¹² created a diagnostic model using MR imaging to differentiate cystic pituitary adenoma and RCC, but the model does not specifically assess rRCC. Prior case reports of rRCCs have not described specific findings to differentiate them from a cystic adenoma.

Intrasellar rupture of an RCC was described in 1988,¹⁰ with associated marked inflammation of the pituitary gland thought to be secondary to the spillage of RCC contents. Numerous additional case reports have since been presented in the literature of symptomatic patients with xanthogranulomatous changes of the pituitary gland.^{3–8,13} It is thought that mucin leaking out of the rRCC can trigger a cascade of surrounding inflammation.¹⁴ The abnormal bone marrow enhancement in our cases, therefore, likely represents an imaging manifestation of the adjacent inflammatory changes. This is most evident in the patient presented in Fig 2, in which the native pituitary was markedly enlarged with suprasellar extension, preoperatively thought to be an adenoma. In this case, the RCC was found to be ruptured, and the pituitary biopsy findings were thought to represent reactive hypophysitis. Further testing was not compatible with lymphocytic hypophysitis or immunoglobulin G4-related disease. Given that the bone marrow enhancement is likely secondary to the inflammatory cascade set

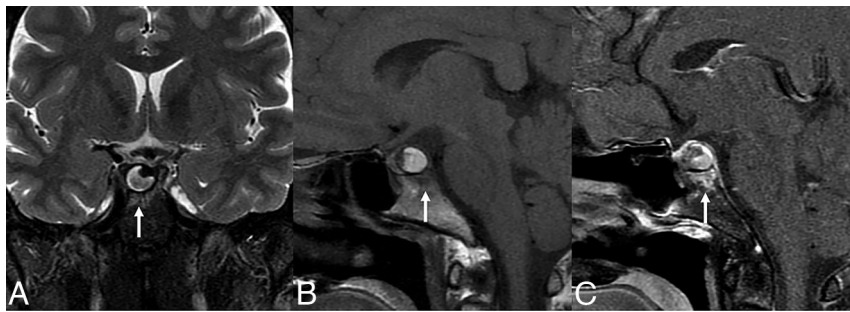


FIG 3. A 39-year-old woman with a history of gamma knife therapy to the sella for an undiagnosed lesion at another institution presented with an enlarging cystic pituitary lesion. T2-weighted imaging (A) shows a hypointense nodule and edema of the basisphenoid bone marrow (arrows). This area has corresponding T1-hypointense signal on precontrast imaging (B) and enhancement (C). Pathology confirmed an rRCC.

Table 2. MR imaging characteristics of patients with pathologic rRCCs

Patient	Max Dimension (mm)	Bone Marrow Enhancement	T2 Signal	T2 Dark Nodule	T1 Hyperintense Signal	Location
1	11.5	Focal	Hyperintense	No	No	Midline
2	14.0	Focal	Hypointense	No	Yes	Midline
3	10.8	Diffuse	Heterogeneous	No	No	Midline
4	10.8	Diffuse	Hyperintense	Yes	Yes	Off-midline
5	11.1	Diffuse	Hyperintense	Yes	No	Midline
6	8.2	Focal	Heterogeneous	No	No	Replaces entire pituitary
7	12.2	Diffuse	Hyperintense	Yes	No	Replaces entire pituitary

Note:—Max indicates maximum.

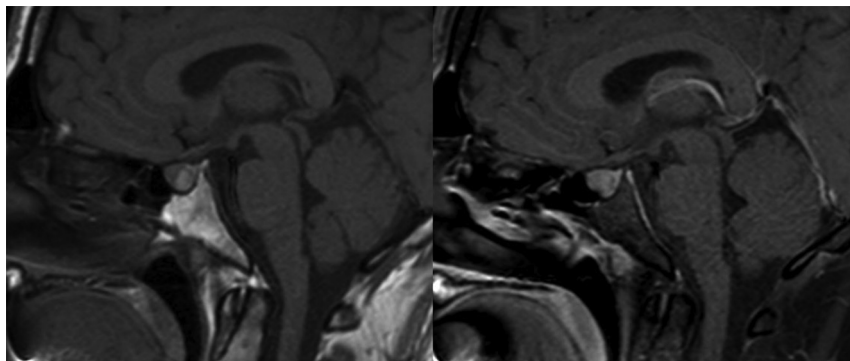


FIG 4. Example of a pathology-proved nonruptured RCC. The precontrast T1-weighted (left) images show normal signal of the basisphenoid bone marrow edema below the T1-hyperintense RCC. The fat-saturated postcontrast image (right) shows normal basisphenoid bone marrow without abnormal enhancement.

off by the RCC rupture, we would not expect to see abnormal enhancement with nonruptured RCCs (Fig 4).

Similar bone marrow imaging findings have been recently described with granulomatous hypophysitis,¹⁵ which is a noncystic inflammatory process of the pituitary gland. In that article, 100 pituitary adenomas were evaluated, none of which had basisphenoid bone marrow enhancement. Therefore, bone marrow enhancement is reflective of an inflammatory process, regardless of the cystic or

solid nature of the sellar process. The current study is the first to describe bone marrow enhancement associated with an rRCC.

Differentiating between a RCC and a cystic pituitary adenoma can be challenging because they often have an overlapping imaging appearance. Pituitary adenomas have been described as off-midline in position and as having fluid levels, a hypointense T2 rim, and septations,¹² while RCCs are associated with T2-hypointense nodules and are typically midline in position.¹¹ Our study is limited by the small number of patients; however, none of our cystic adenomas had basisphenoid bone marrow enhancement, nor has this previously been described in the literature. The differential for cystic lesions of the pituitary gland extends beyond RCC and cystic adenoma, but such lesions are expected to demonstrate different imaging findings and clinical presentations. Adamantinomatous craniopharyngiomas occur more frequently in children and are typically suprasellar and calcified.¹⁶ Pituitary abscess could also appear as a cystic lesion and, due to marked surrounding inflammatory changes, could also have bone marrow signal changes; however, an abscess should also present with restricted diffusion,¹⁷ whereas an RCC would not.¹⁸ Our patients did not have DWI for us to analyze. The broad category of hypophysitis has many primary and secondary causes, ranging from immunoglobulin G4-related disease to infectious causes such as tuberculosis.¹⁹ While these entities, outside of granulomatous hypophysitis,¹⁵ have not been previously described as having bone marrow enhancement, it is certainly possible, given the associated inflammation. These should present as solidly enhancing enlargement of the pituitary gland, however, rather than the cystic appearance expected for an RCC.

Diagnosing basisphenoid bone marrow inflammatory changes on MR imaging is predicated on having bone marrow below the sella (non-type 4 sphenoid pneumatization) and the correct MR imaging protocol, and this requirement limits generalization of this finding to all pituitary cases. The findings are most easily seen when the T2-weighted and postcontrast T1-weighted images are fat-saturated. Even in the absence of fat saturation, however, precontrast T1-weighted images can show dark bone marrow

signal (Fig 3). We believe that the presence of MR signal abnormalities consistent with basisphenoid inflammation is an important finding that can potentially help to identify an rRCC in the appropriate clinical setting. Although the small number of patients included in this report limits its generalizability, it can serve as a starting point for future studies that include more patients.

CONCLUSIONS

RCCs are commonly seen on MR imaging as cystic pituitary lesions. Intracellular rupture of an RCC is uncommon and without previously described imaging findings to differentiate it from other pathologies. We present 7 cases of basisphenoid bone marrow enhancement below the sella that could be used to potentially identify an rRCC before surgical exploration.

Disclosure forms provided by the authors are available with the full text and PDF of this article at www.ajnr.org.

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