Suprasellar Mature Cystic Teratoma Mimicking Rathke’s Cleft Cyst: A Case Report and Systematic Review of the Literature

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In this article, we present a 31-year-old female who presented with intermittent headache and oligomenorrhea of over 10 years’ duration. Imaging revealed a large suprasellar mass with sellar extension. The patient underwent an endoscopic endonasal trans-sphenoidal surgery to resection of the mass. Clinical, radiological, and operative findings from this patient were initially considered to be Rathke’s cleft cyst (RCC). However, postoperative histological examinations revealed a mature cystic teratoma. No radiotherapy was performed after surgery. At the most recent follow-up, approximately 1 year later, the patient is doing well with no headache and no recurrence of the teratoma.

Keywords: mature cystic teratomas, sellar region, rare lesion, neuropathology, case

INTRODUCTION

Teratomas are a type of germ cell tumor (GCT) differentiating from three germ layers. Central nervous system teratomas are very rare, accounting for 0.2%–0.9% of all intracranial tumors (1). According to The 2016 WHO Classification of Tumors of the Central Nervous System, teratomas can be classified into three types: mature, immature, and teratomas with malignant transformation (2). Mature teratomas are benign tumors that contain well-differentiated tissues from at least two germinal layers which can be divided into two subtypes: mature solid teratomas and mature cystic teratomas (MCT); the former is exceedingly rare. The latter accounts for about 0.04% to 0.7% of all intracranial tumors (3, 4). Mature teratoma recurrence rate is extremely low in cases of complete resection and usually occurs within 1 year after treatment (5), and the 10-year survival rate is 93% (6). Most of the intracranial MCTs have been found to occur in the midline structures, and the pineal area is the most frequent site (7, 8). Suprasellar MCTs have rarely been reported. Here, we describe an unusual case of a large suprasellar MCT mimicking Rathke’s cleft cyst, and conduct a systematic review of eight cases of MCTs in the sellar region (Tables 2–4). We hope to shed new light for physicians on the diagnosis and treatment of this rare disease.
CASE PRESENTATION

History and Examination
A 31-year-old female was admitted to our hospital complaining of oligomenorrhea and increasing headaches. She reported an 11-year history of intermittent headache (visual analog scale, with 10 as the worst pain, of 4/10 points), which used to be precipitated by fatigue and were alleviated by rest or non-steroidal anti-inflammatory drugs (NSAIDs). When the headaches increased in frequency and intensity and were accompanied by mild nausea, culminating in a headache lasting for 1 week with no relief from NSAIDs, the patient sought medical attention. She denied vision loss, visual field defects, polyuria, lactation, central obesity, or acromegaly during the course. The general physical examination was completely normal, and the neurologic examination showed no focal signs. A brain magnetic resonance imaging (MRI) scan with contrast was performed, demonstrating a 19 mm × 24 mm × 23 mm irregular suprasellar lesion with slight intrasellar extension. The lesion signal characteristics were isointense on T1-weighted imaging and hyperintense on T2-weighted imaging. No obvious gadolinium enhancement was noted (Figures 1A–C). Endocrine workup showed that the levels of pituitary hormones were within normal limits (Table 1).

Surgical Biopsy and Histological Findings
Endoscopic trans-sphenoidal surgery was performed. In the procedure, the cyst was observed to be predominantly suprasellar in location. It contained ivory-whitish viscous material and was resected. Hematoxylin–eosin staining is as follows: on a background of abundant myxoid stroma, we can see the following components: fibrous cyst walls lined with simple cuboidal and short columnar epithelium (H&E ×100, Figure 1D), a mass of mucous acinous cells (Figure 1E), and some chondroid tissue (Figure 1F).

Postoperative Course
The postoperative course was uneventful, with the headaches completely resolving after surgery. During the 1-year follow-up, our patient is well and there is no evidence of recurrence.

DISCUSSION
In the case report, we present a unique and rare case of MCT mimicking Rathke’s cleft cyst (RCC) of the sellar region in terms of clinical manifestations and neuroimaging.

According to The 2016 WHO Classification of Tumors of the Central Nervous System (2), teratomas are a subset of intracranial germ cell tumors and rarely present as pure teratomas (rather than mixed germ cell tumors). Teratomas can be classified into three types: mature, immature, and teratomas with malignant transformation. Intracranial teratomas are rare space-occupying lesions that account for about 0.5% of all intracranial tumors. MCTs are a subset of these neoplasms, and their occurrence in the brain is even rarer. They are benign tumors that contain well-differentiated tissues from at least two germinal layers. MCTs occur more frequently during the first or second decade of life, and there is a clear male predominance (4:1). Most intracranial MCTs occur in midline structures, most frequently in the pineal region (7).

MRI is the first choice of neuroimaging in the diagnosis of RCC. On MRI, RCCs often appear as well-demarcated, centrally located spherical or ovoid lesions of the sellar region with...
nodules inside the cyst occasionally. The majority of these smooth contoured cysts are unilobar with a diameter ranging between 5 and 40 mm (mean approximately 17 mm) (9). MRI signal intensity varies and is highly dependent on the biochemical nature of intracystic contents, which can range from clear, CSF-like fluid to thick, mucoid material (10, 11).

In the present case, RCC was suspected prior to the histological examinations of the tumor because the gender, age, clinical presentations, and neuroimaging characteristics aligned with a diagnosis of RCC.

Suprasellar MCTs are relatively rare. MCTs occur more frequently during the first or second decade of life. Rarely, reported cases have occurred on the third or fourth decade of life (5). Overall, these tumors appear to be more common in men, with a finding of 79.7% in men versus 20.3% in women (6). Moreover, the tumor mimicking RCC is a further peculiarity of the case.

The published case reports and series written in English that focus on suprasellar MCTs are limited. Therefore, we performed a comprehensive literature review of related articles and identified eight patients with a diagnosis of MCT, summarizing the data of clinical manifestations (Table 2), pituitary function (Table 3), MRI signal features (Table 3), and treatment (Table 4) of this lesion.

The most prominent symptoms at diagnosis are neurological defects (six of eight patients), particularly visual disturbance (five of eight). Headache (three of eight) and diabetes insipidus (three of eight) were also commonly seen. One patient reported amenorrhea. Regarding MRI appearance, signal intensities on T1WI and T2WI vary from case to case. In some cases, inclusions like teeth, fat, and calcification can be detected (13, 15). Variable enhancement with contrast was reported in three patients. Liu et al. (19) suggest that mature teratoma on MRI is an ovoid or irregular mass with or without multilocularity and has mixed signals derived from different tissues. The presence of fatty tissue or multilocularity is a characteristic feature of teratoma. The tumor usually presents with heterogeneous hyperintensity on T1W images and non-enhanced or moderate

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**TABLE 1 | Results of endocrine examinations before and after surgery.**

| Test          | Reference range         | Before surgery Value | 3 months after surgery Value |
|---------------|-------------------------|----------------------|------------------------------|
| LH            | 1.20–103.03 IU/L        | 10.08                | 19.36                        |
| FSH           | <30.34 IU/L             | 6.57                 | 10.33                        |
| E2            | 27–433 pg/ml            | 60                   | 130                          |
| P             | 0.38–29.26 ng/ml        | 0.211                | 1.73                         |
| PRL           | <30 ng/ml               | 16.72                | 4.92                         |
| ACTH related  |                         |                      |                              |
| ACTH          | 0–46 pg/ml              | 18.9                 | 10.5                         |
| F             | 4.0–22.3 µg/dl          | 17.28                | 12.35                        |
| FT3           | 1.80–4.10 pg/ml         | 3.17                 | 2.67                         |
| FT4           | 0.81–1.89 ng/dl         | 1.081                | 1.222                        |
| T3            | 0.66–1.92 ng/ml         | 1.179                | 0.824                        |
| T4            | 4.30–12.50 µg/dl        | 7.6                  | 8.06                         |
| TSH           | 0.36–4.34 µIU/mL        | 3.154                | 2.335                        |

LH, luteinizing hormone; FSH, follicle-stimulating hormone; E2, estradiol; P, progesterone; PRL, prolactin; β-HCG, β-human chorionic gonadotropin; ACTH, adrenocorticotropin hormone; F, cortisol; FT3, free triiodothyronine; FT4, free thyroxine; T3, triiodothyronine; T4, thyroxine; TSH, thyroid-stimulating hormone.

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**TABLE 2 | Demographic data and clinical presentation from published reports.**

| Patient | Author          | Year | Country | Age/sex | Presentation                                      | Other manifestation                                      |
|---------|-----------------|------|---------|---------|--------------------------------------------------|---------------------------------------------------------|
| 1 (12)  | Li et al.       | 2015 | China   | 13/F    | Polyuria, polydipsia, and amenorrhea              | Headache, blurred vision, short stature                  |
| 2 (8)   | Sweiss et al.   | 2013 | USA     | 57/M    | Vision impairment                                 | Left-sided facial weakness, ataxia, and short-term memory loss, seizure |
| 3 (13)  | Vendrell et al. | 2010 | France  | 18 months/M | Bilateral decreased visual acuity and hyperphagia | Bilateral decreased visual acuity and hyperphagia       |
| 4 (14)  | Kim et al.      | 2010 | Korea   | 17/M    | Polyuria and polydipsia with severe thirst, headache, and diplopia | Polyuria and polydipsia with severe thirst, headache, and diplopia |
| 5 (15)  | Muzumdar et al. | 2001 | India   | 26/M    | Headache, vision impairment                       | Headache, vision impairment                              |
| 6 (16)  | Araki et al.    | 2000 | Japan   | 3 months/M | Fontanelle bossing                               | Fontanelle bossing                                      |
| 7 (17)  | Narayanam et al.| 2012 | India   | 7/F     | Seizures, precocious puberty, headache, and vomiting | Seizures, precocious puberty, headache, and vomiting   |
| 8 (18)  | Tobo et al.     | 1981 | Japan   | 14/M    | Diabetes insipidus/panhypopituitarism              | Diabetes insipidus/panhypopituitarism                     |
| 9 (Current case) | Tobo et al. | 2021 | China   | 31/F    | Headache, oligomenorrhea                          | Headache, oligomenorrhea                                |

F, female; M, male.
enhanced multilocularity on T1W images with contrast. However, Chiloiro et al. (5) suggest that teratomas appear as low-intensity heterogeneous mass in T1- and T2-weighted magnetic resonance imaging, with variable enhancement after contrast administration. Surgery was performed in all patients, two of which were followed by radiotherapy or chemotherapy. Only one patient reported hydrocephalus and blindness during follow-up.

Neuroimaging characteristics of teratomas are not of high specificity, which make it difficult to distinguish mature teratomas from other intracranial neoplasms located in the suprasellar region that include other GCTs (germinoma, choriocarcinoma, embryonal carcinoma, and endodermal sinus tumor), craniopharyngioma, and RCC. Therefore, our case highlights the importance of obtaining a histological diagnosis to differentiate teratomas from other lesions.

### TABLE 3 | Pituitary function and pituitary magnetic resonance imaging data from published reports.

| Patient | Pituitary function | Size | Location | T1 | T2 | T1 contrast | CT |
|---------|--------------------|------|----------|----|----|------------|----|
| 1       | T4↓, TSH↑, PRL↑    | Large | Sellar and suprasellar | Cystic: hypointense | Heterogeneous intensity | Cystic: rim enhancement; solid: evidently enhanced | Hypodense |
| 2       | FSH↑, LH↑          | Large | Intra- and suprasellar | Mixed intensity | Mixed intensity | Intense enhancement | Hyperdense due to calcification |
| 3       | PRL↑, TSH↑, T3↑    | Large | Endosuprasellar | NA | NA | Partial enhancement | NA |
| 4       | ADH↓              | Large | Sellar and suprasellar | Hyperintense | NA | NA | Hypodense with peripheral rim of calcification |
| 5       | Panhypopituitarism | Large | Suprasellar | Hypointense | NA | NA | Hypodense |
| 6       | Normal             | Large | Suprasellar | Hypointense | NA | NA | Hypodense |
| 7       | Normal             | Large | Sellar and suprasellar | Isointense | Isointense | No enhancement | A mass in the suprasellar region with contrast enhancement |
| 8       | Panhypopituitarism | Large | Sellar and suprasellar | Iso/ hyperintensity | Hyperintensity with a hypointense nodule | No enhancement | – |

Size: large = large tumor size (>1 cm). 
T3, triiodothyronine; T4, thyroxine; TSH, thyroid-stimulating hormone; LH, luteinizing hormone; FSH, follicle-stimulating hormone; PRL, prolactin; ADH, anti-diuretic hormone; NA, not available.

### TABLE 4 | Treatment and outcome of patients from published reports.

| Patient | Surgery | Tumor contents | Pathology | Outcome | Complication |
|---------|---------|----------------|-----------|---------|--------------|
| 1       | Right pterional approach | Dark yellow fluid; hair and whitish fat material | Mature teratoma | Total resection/vision improved | Transient DI |
| 2       | Right pterional craniotomy/ trans-sylvian approach | Thick and yellow oil-like fluid, yellow clumps of hair embedded within fatty deposit | Mature cystic teratoma | Incomplete resection followed by external beam radiotherapy and stereotactic radiosurgery/significantly improved neurological status and vision | No |
| 3       | TSS | Teeth | Mature teratoma | Normal neurological examination except loss of visual acuity in the left eye | No |
| 4       | TSS | Fat, bony septation, keratinaceous flakes | Mature teratoma | Followed by chemotherapy and radiotherapy | NA |
| 5       | Sublabial trans-sphenoidal approach | White structure | Mature teratoma | Total resection/vision improved, normal visual field, headache gone | No |
| 6       | Surgery | Whitish structure | Mature teratoma | Total resection/panhypopituitarism and diabetes insipidus | Hydrocephalus/ complete blindness |
| 7       | Left pterional approach | Bone, cartilage, and several hairs | Mature teratoma | Total resection/headache gone, seizure-free, regression of precocious puberty | No |
| 8       | Craniotomy | Ivory-whitish viscous materials | Mature cystic teratoma | Total resection/headache resolved | No |

TSS, trans-sphenoidal surgery; NA, not available.
Histologically, MCTs are commonly multicystic, contain sebaceous fluid, and are identified by the presence of differentiated ectodermal (skin, hair, brain), mesodermal (muscle, fat, teeth, bone, cartilage), and/or endodermal elements (mucinous and ciliated epithelium). All three layers may not be seen in every case of teratoma. The differential diagnosis includes dermoid cysts, epidermoid cysts, colloid cysts, immature teratomas, and teratomas with malignant transformation.

For this case, our preoperative diagnosis was Rathke's cleft cyst, and given the absence of hair, skin, or teeth, the intraoperative findings seemed to confirm our primary diagnosis. However, MCTs were confirmed by the histological examination of the specimen when cyst walls lined with simple cuboidal and columnar epithelium, a mass of mucous acinous cells (salivary glands), and cartilage were identified. Our case highlights the importance of obtaining a histological diagnosis to differentiate MCTs from other lesions. It would also be important to exclude the presence of additional germ cell components, which would require additional treatment postresection.

The typical treatment for mature teratomas is neurosurgical excision because of their benign behavior (20), which was successfully done in this case. It is well advised to perform radical excision as the long-term outcome is excellent. Mature teratoma recurrence rate is extremely low in cases of complete resection and usually occurs within 1 year after treatment (5). Sano (6) reported that the 10-year survival rate for mature teratomas is 93%. Whether to perform radiotherapy for mature teratomas after surgery remains controversial. Sano (6) points out that radiotherapy should be conducted after surgery to suppress further growth of tumor cells. Jakacki (21) suggests that it is advocated to perform radiotherapy to immature teratomas and teratomas with malignant transformation; while mature teratomas are not typically responsive to radiation therapy, surgery is the only proven treatment modality. Therefore, the clinical experience from physicians really matters in the postoperative treatment choices for patients with mature teratomas.

CONCLUSION

MCTs in the sellar region are extremely rare, and their imaging usually lacks specificity. Therefore, it is important to obtain a thorough histological diagnosis. MCTs are benign, and complete surgical excision is the first-line treatment. In selected cases, radiation therapy was conducted in some cases but is not recommended as routine treatment. Whether to perform radiotherapy depends on the physician as there is a lack of evidence on this aspect. Close follow-up is indispensable for patients with MCTs.

DATA AVAILABILITY STATEMENT

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

ETHICS STATEMENT

Written informed consent was obtained from the participant for the publication of any potentially identifiable images or data included in this article. The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

AUTHOR CONTRIBUTIONS

SJ drafted the manuscript. SJ and ZW analyzed the data. YY made the pathological diagnosis and drafted the article of pathological findings. All authors contributed to the article and approved the submitted version.

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