Surgical Management of Rathke Cleft Cysts

Vishwaraj Ratha[1], Subodh Patil[1], Vikram S. Karmarkar[1], Nishit J. Shah[2], Chandrashekhar E. Deopujari[1]

BACKGROUND: **The diagnosis of Rathke cleft cysts (RCC) has increased in recent times as a result of improvements in imaging techniques; however, symptomatic patients are uncommon and accurate preoperative diagnosis may sometimes be difficult. The indications of surgical management protocol are evolving. We aim to provide a comprehensive review of clinical, imaging, and histopathologic features with operative management strategies along with outcome and prognosis in RCC.**

METHODS: **A retrospective analysis (2003–2015) was performed of 58 consecutive cases of RCC seen in a surgical unit. Twenty-seven surgically treated symptomatic RCCs were further evaluated for their clinical presentation, imaging characteristics, surgical approaches, and intraoperative findings.**

RESULTS: **Headache was the most common presenting complaint followed by visual deficit. Hormonal abnormality was observed in 13 patients. On magnetic resonance imaging, the characteristic intracystic nodule was identified in 6 patients. Transsphenoidal surgery for cyst excision was performed in all 27 patients with an endoscopic route in 25 patients and radical excision was performed in 17 patients. The pituitary stalk and the normal gland were preserved in all patients. Headache improved in 96% of patients and visual field defect resolved in all. Around 46% had improvement of the anterior pituitary axis. New permanent hormone deficiency was not observed. The recurrence rate was 3.7% after a minimum of 18 months follow-up.**

CONCLUSIONS: **RCCs are an uncommon disease with a wide spectrum of clinical and radiologic features. Endonasal endoscopic transsphenoidal surgery provides excellent clinical and endocrinologic improvement. We believe that radical excision does not necessarily result in endocrinologic impairment and may have a better impact on recurrence and cyst resolution.**

INTRODUCTION

Rathke cleft cyst (RCC) is a benign, nonneoplastic cystic lesion that originates from congenital remnants of the Rathke pouch.[1] These are predominantly sellar lesions with or without suprasellar extension and occasionally are exclusively suprasellar. Most are asymptomatic, encountered in normal pituitary glands in routine autopsies.[2,3] However, they are occasionally large enough to produce visual or endocrinologic disorders by compressing visual apparatus, pituitary gland, or rarely, hypothalamus. They are clinically significant but uncommon lesions. Various case reports have described their coexistence along with pituitary tumors.[4]

Over the last few decades, improvement in neuroimaging techniques has increased the diagnosis of RCC, leading to its better understanding. It is thus imperative to differentiate clinically significant from incidental and other cystic lesions of this region, such as craniopharyngioma, cystic pituitary adenoma, and so on. A proper preoperative analysis is essential, especially in incidentally detected asymptomatic patients, because it alters the management protocol.[5]

The literature has shown that surgery in symptomatic RCCs improves both endocrine and visual function.[6] A thorough understanding of the surgical anatomy and pathophysiology is paramount to plan out an optimum management strategy. The

Key words
- Endocrine recovery
- Endoscopic endonasal transsphenoidal surgery
- Rathke cleft cyst
- Surgical outcome

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
DI: Diabetes insipidus
EETS: Endonasal endoscopic transphenoidal surgery
MRI: Magnetic resonance imaging
RCC: Rathke cleft cyst
TSS: Transsphenoidal surgery

From the Departments of [1]Neurosurgery and [2]Otorhinolaryngology, Bombay Hospital Institute of Medical Sciences, Mumbai, India

To whom correspondence should be addressed: Vishwaraj Ratha, M.Ch Neurosurgery. [E-mail: thinkvishwaraj@gmail.com]

Citation: World Neurosurg. (2017) 107:276-284.
http://dx.doi.org/10.1016/j.wneu.2017.07.164

Journal homepage: www.WORLDNEUROSURGERY.org

Available online: www.sciencedirect.com

aim of our study is to provide a comprehensive review of clinical, imaging, and histopathologic features, as well as operative and nonoperative management strategies in patients with RCCs.

We also attempt to evaluate our evolving surgical techniques over the last decade for symptomatic RCCs and understand the outcome and prognosis of such lesions because the data available for RCC from India are restricted to a few case reports and case series.[7-10]

METHODS

Patient Population

This is a retrospective study of patients with RCC, treated at Bombay Hospital from 2003 to 2015 by the 2 senior authors. The medical records were retrieved from a database of 884 patients with sellar-suprasellar lesions who were either under observation or had surgical treatment. Only cases histologically proved or with neuroimaging features strongly suggestive of RCC were included. The present study was approved by the institutional review board. Data collected included demographics, clinical charts, visual examination, neuroimaging, operation notes, and pathologic reports. All patients studied consented to the chart review and, in addition, were contacted by telephone for the follow-up.

Study Population and Design

A total of 58 cases were retrieved; 27 patients underwent surgical intervention whereas the remaining 31 were managed conservatively and observed for progression of size and/or symptoms. Twelve patients in the conserved group were lost to follow-up and in the remaining 19 patients, a minimum follow-up of 3 years did not result in crossover to the surgical group. Neuroimaging reviews of all 27 surgical patients were available, and in the conserved group, we relied on the original radiologist's report for 15 patients whose films could not be obtained for review. Neuroimaging included gadolinium-enhanced magnetic resonance imaging (MRI) scans (pituitary protocol) for most of the patients except for the patients diagnosed in the earlier part of decade, when contrast-enhanced computed tomography was used. Endocrine evaluation was carried out using standardized basal and dynamic stimulation tests.[11] These tests were later categorized according to each identified pituitary hormonal abnormality. Headache was assessed according to its subjective quality, site, and severity. Visual status examination included a formal perimetry evaluation and acuity testing before and after surgery.

Statistical analysis of the data was performed using SPSS 15.0 software (SPSS Inc., Chicago, Illinois, USA).

Imaging, Hormonal Assessments, and Outcomes

RCCs were characterized based on preoperative and postoperative MRI findings and categorized as 1) purely intrasellar, 2) intrasellar/suprasellar, or 3) purely suprasellar. Hormonal data included preoperative and postoperative levels of serum prolactin, morning serum cortisol and adrenocorticotropic hormone, thyroid-stimulating hormone and free T4, free T3, insulinlike growth factor 1, total and free testosterone, serum sodium, and urine specific gravity. All patients with endocrinologic dysfunction (preoperatively and postoperatively) were referred to the endocrinologist for a thorough assessment and opinion. The medical management was formed in conjunction with their assessment and after a discussion. Clinic notes were reviewed for preoperative and postoperative visual status (visual acuity and visual field tests), report of headaches, menstrual/sexual history, and implementation of hormone replacement. Sexual dysfunction was defined as either low sexual libido and/or erectile dysfunction. The RCC diagnosis was confirmed by histologic examination of the surgical material in each case. Postoperative outcomes were determined from the clinical notes. The first follow-up after surgery was performed at 6 weeks followed by 3 months, 6 months, and 1 year.

The biochemical assessment was performed at 6 weeks and then at 6 months and later at yearly intervals for 3 years, and the radiologic follow-up was performed at 6 weeks followed by 6 months and yearly depending on 1) clinical symptoms or 2) radiologic findings on first postoperative imaging.

Surgical Technique

All patients underwent RCC removal via a direct endonasal endoscopic transphenoidal surgery (EETS) approach except for the first 2 cases, which were performed as endoscope-assisted microsurgery, as described by Zada et al.[12] When the cyst was located behind the anterior pituitary gland, occasionally its removal involved a direct approach through the anterior inferior pituitary gland via a low midline vertical glandular incision (i.e., a transpituitary approach). At the completion of cyst removal, the cavity was carefully inspected for residual cyst contents and cyst lining. An angled endoscope was used for assisted visualization in all endoscopic cases. Radical excision was attempted only when it was deemed safe and feasible. Small weeping cerebrospinal fluid (CSF) leaks when identified after cyst removal were repaired with collagen sponge; larger CSF leaks were repaired with fat and fascia lata and collagen sponge; and large diaphragmatic leaks had a lumbar drain placed for 48–72 hours of CSF diversion. For patients without an intraoperative CSF leak, only a collagen sponge was placed.

RESULTS

Patient Population

Of 884 consecutive patients diagnosed with sellar-suprasellar lesions from 2000 to 2015, 58 (6.56%) had RCC, 27 of whom required intervention. Most patients in the surgical arm were female (63%), with a median age of 37 years (range, 18–58 years) and most of whom were premenopausal. The median duration of follow-up was 38 months, with a minimum follow-up of 18 months.

Presenting Symptoms

Headache was the most common presenting symptom in the surgical group, seen in 25 of 27 patients (92.6%). It was chronic in 19 patients and 6 had sudden acute episodes. Visual field impairments were identified in 6 patients (22.2%) and all had bitemporal hemianopia on perimetry testing. Only 1 patient, who presented with visual acuity disturbance and double vision, had suspected hemorrhage in RCC. Endocrinologic symptoms and signs at presentation were noted in 13 patients (48.1%). Table 1 describes the demographic details and presenting complaints of the patients.