Rathke cleft cyst case series

20 patients with LRCCs and 25 patients with CCPs. Both tumors had a maximal diameter of more than 20 mm. We evaluated the patients' clinical and MR imaging findings, including symptoms, management strategies, outcomes, anatomic growth patterns, and signal changes.

The age of onset for LRCCs versus CCPs was 49.0 ± 16.8 versus 34.2 ± 22.2 years (p = .022); the following outcomes were observed for LRCCs versus CCPs: (1) postoperative diabetes insipidus: 6/20 (30%) versus 17/25 (68%) (p = .006); and (2) posttreatment recurrence: 2/20 (10%) versus 10/25 (40%) (p = .025). The following MR findings were observed for LRCCs versus CCPs: (1) solid component: 7/20 (35%) versus 21/25 (84%) (p = .001); (2) thick cyst wall: 2/20 (10%) versus 12/25 (48%) (p = .009); (3) intracystic septation: 1/20 (5%) versus 8/25 (32%) (p = .030); (4) snowman shape: 18/20 (90%) versus 1/25 (4%) (p < .001); (5) off-midline extension: 0/0 (0%) versus 10/25 (40%) (p = .001); and (6) oblique angle of the sagittal long axis of the tumor: 89.9° versus 107.1° (p = .001).

LRCCs can be differentiated from CCPs based on their clinical and imaging findings, especially their specific anatomical growth patterns. They suggest using the pretreatment diagnosis to select the appropriate surgical approach and thus improve the clinical outcome [1)]

2018

Lin et al., conducted a retrospective analysis of 109 consecutive cases of pathology-confirmed RCCs treated via a transsphenoidal approach at a single center from 1995 to 2016. The majority of cases (86.2%) involved cyst fenestration, drainage, and partial wall resection.

A total of 109 surgeries in 100 patients were included, with a mean follow-up duration of 67 months (range 3-220 months). The mean patient age was 44.6 years (range 12-82 years), and 73% were women. The mean maximal cyst diameter was 14.7 mm. Eighty-eight cases (80.7%) were primary operations, and 21 (19.3%) were reoperations. Intraoperative CSF leak repair was performed in 53% of cases and was more common in reoperation cases (71% vs 48%, p < 0.001). There were no new neurological deficits or perioperative deaths. Two patients (1.8%) developed postoperative CSF leaks. Transient diabetes insipidus (DI) developed in 24 cases (22%) and permanent DI developed in 6 (5.5%). Seven cases (6.4%) developed delayed postoperative hyponatremia. Of the 66 patients with preoperative headache, 27 (44.3%) of 61 reported postoperative improvement and 31 (50.8%) reported no change. Of 31 patients with preoperative vision loss, 13 (48.1%) reported subjective improvement and 12 (44.4%) reported unchanged vision. Initial postoperative MRI showed a residual cyst in 25% of cases and no evidence of RCC in 75% of cases. Imaging revealed evidence of RCC recurrence or progression in 29 cases (26.6%), with an average latency of 28.8 months. Of these, only 10 (9.2% of the total 109 cases) were symptomatic and underwent reoperation.

Transsphenoidal fenestration and drainage of RCCs is a safe and effective intervention for symptomatic lesions, with many patients experiencing improvement of headaches and vision. RCCs show an appreciable (although usually asymptomatic) recurrence rate, thereby mandating serial follow-up. Despite this, full RCC excision is typically not recommended due to risk of hypopituitarism, DI, and CSF leakage [2)].

2011

A total of 33 patients (20 females, median age 43 years) were identified. At presentation, major visual field defects were detected in 58% of patients and gonadotrophin, ACTH and TSH deficiency in 60, 36 and 36% of patients respectively. Desmopressin treatment was required in 18% of patients. Treatment consisted of cyst evacuation combined with or without biopsy/removal of the wall. Post-operatively, visual fields improved in 83% of patients with impairment, whereas there was no reversal of ACTH or TSH deficiency or of diabetes insipidus. All but one subject had imaging follow-up during a mean period of 48 months (range 2-267). Cyst relapse was detected in 22% of patients at a mean interval of 29 months (range 3-48 months); in 57% of them, the recurrence was symptomatic. Relapse-free rates were 88% at 24-months and 52% at 48-months follow-up. At last assessment, at least quadrantanopia was reported in 19% of patients, gonadotrophin, ACTH and TSH deficiency in 50, 42 and 47% of patients respectively. Desmopressin treatment was required in 39% of patients.

In this study of patients with RCC and long-term follow-up, we showed a considerable relapse rate necessitating long-term monitoring. Surgical intervention is of major importance for the restoration of visual field defects, but it does not improve endocrine morbidity, which in the long-term affects a substantial number of patients [3].

1)
Yang CH, Wu CH, Lin TM, Chen ST, Tai WA, Yu KW, Luo CB, Lirng JF, Chang FC. Clinical and imaging findings for the evaluation of large Rathke's cleft cysts and cystic craniopharyngiomas. Pituitary. 2023 May 25. doi: 10.1007/s11102-023-01326-3. Epub ahead of print. PMID: 37227614.
2)
Lin M, Wedemeyer MA, Bradley D, Donoho DA, Fredrickson VL, Weiss MH, Carmichael JD, Zada G. Long-term surgical outcomes following transsphenoidal surgery in patients with Rathke's cleft cysts. J Neurosurg. 2018 May 18:1-7. doi: 10.3171/2017.11.JNS171498. [Epub ahead of print] PubMed PMID: 29775155.
3)
Trifanescu R, Stavrinides V, Plaha P, Cudlip S, Byrne JV, Ansorge O, Wass JA, Karavitaki N. Outcome in surgically treated Rathke's cleft cysts: long-term monitoring needed. Eur J Endocrinol. 2011 Jul;165(1):33-7. doi: 10.1530/EJE-11-0142. Epub 2011 Apr 18. PubMed PMID: 21502327.