Devastating complications from alcohol cauterization of recurrent Rathke cleft cyst

Case report

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Rathke cleft cysts are commonly found on autopsy but are seldom symptomatic. Conventional treatment of these lesions consists of transsphenoidal drainage with partial excision of the cyst, and recurrence is rare. Alternatively, the instillation of absolute alcohol into the cyst has been described in several reports, with no complications. The authors report on a woman with Rathke cleft cyst that recurred three times after the initial treatment; the lesion was treated with alcohol cauterization on the final recurrence with devastating complications. This 51-year-old woman presented in 1992 with headaches and visual disturbances. Admission magnetic resonance imaging revealed a sellar/suprasellar lesion that was treated with conventional surgery and was subsequently confirmed to be a Rathke cleft cyst. The patient again presented with recurrence of the cyst at 22, 26, and 31 months after the initial presentation. On the final recurrence the cyst was treated with alcohol cauterization. Postoperatively, the patient awoke blind and suffered a seizure from leakage of the alcohol. The patient remains completely blind in both eyes and is also anosmic and has left lid ptosis and exotropia, which signify damage to the first through third cranial nerves.

Rathke cleft cysts have been known to recur after primary surgery; however, this is the first report of a single cyst recurring three times despite conventional surgical treatment. Additionally, this is the first report in which devastating complications from alcohol cauterization of the cyst have been described. The authors therefore advocate caution when attempting alcohol cauterization and advise that meticulous care be taken to ensure the patency of the cyst.

KEY WORDS • Rathke cleft cyst • transsphenoidal surgery • alcohol cauterization • blindness • magnetic resonance imaging

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Case Report

History and Initial Operation. This 51-year-old woman presented to our ophthalmology clinic at the Doheny Eye Institute in April 1995 for evaluation of bilateral blindness. As detailed in her medical records, in early 1992, 3 years before our initial evaluation, the patient had presented to another hospital with symptoms of headaches of approximate.
ly 5 months’ duration; subjective decreases in visual acuity, predominantly in the left eye; and an increased left-sided blind spot. At that time, the patient reported normal menstrual periods, and a laboratory endocrine workup revealed a normal T4 (7.4 μg/dl), a T3 uptake of 27%, and a normal prolactin level of 20.9 ng/ml. An MR image obtained in June 1992 had revealed a 2-cm sellar/suprasellar mixed cystic and solid mass compressing the optic chiasm with upward displacement of the chiasm (Fig. 1). The initial diagnosis was pituitary adenoma rather than craniopharyngioma, and the patient underwent a transsphenoidal hypophysecytomy in July 1992 at the other hospital. Intraoperatively, a large cyst was seen and 5 ml of milky white fluid was expressed from it; subsequently, biopsy samples of the tissues surrounding the cyst were obtained. The pathology report on the cyst fluid revealed predominantly acellular, proteinaceous material with rare white cells and rare ciliated columnar epithelial cells, and the tissue biopsy sample showed acidophilic and basophilic cells from the adenohypophysis that had normal architecture. The patient did well and was discharged on postoperative Day 3. Results of a follow-up CT scan obtained on postoperative Day 21 were normal.

Second Operation. In May 1994, the patient again presented to the same hospital with symptoms of headache and visual disturbances. A repeated MR image revealed a recurrence of the cystic lesion in the sella turcica measuring 1.7 cm, with suprasellar extension and compression/displacement of the optic chiasm toward the right. In a repeated transsphenoidal hypophysectomy a cyst with gray-yellow, proteinaceous material was resected, and biopsy samples of the surrounding tissues were obtained. Histological examination demonstrated fragments of ciliated columnar epithelium and cellular crush artifact in otherwise normal pituitary tissue.

Third Operation. The patient returned in September 1994 with symptoms of headache and visual disturbances and was admitted for recurrence of the sellar cyst as demonstrated on MR imaging. The cyst measured 2 cm in diameter and there was significant compression of the optic chiasm. The visual field examination performed during this admission showed bitemporal field deficits. A third transsphenoidal hypophysectomy was performed, and 4 ml of green-yellow fluid was drained. Instillation of absolute alcohol was planned but abandoned because of the development of a CSF leak. The defect in the arachnoid was patched with a muscle graft, and the patient recovered uneventfully and was discharged. One month after discharge, a repeated MR image demonstrated a recurrence of the cyst, which measured 1.5 cm in diameter and displayed continuing suprasellar extension and compression of the optic chiasm. Nevertheless, the signal intensity of the cystic contents had increased on both T1 and T2-weighted images, which was indicative of subacute hemorrhage into the cyst.

Fourth Operation. The patient was readmitted in February 1995 for intractable headaches and nausea; she did not report any new visual problems. An MR image demonstrated resolution of the subacute hemorrhage in the cyst, but we observed interval enlargement of the cyst diameter from 1.5 to 2 cm and compression of the optic chiasm. A post-Gd MR image demonstrated no enhancement of the sellar mass. The patient was brought to the operating room for repeated transsphenoidal hypophysectomy and instillation of absolute alcohol. Six to 7 ml of green fluid was initially aspirated from the cyst, and on a subsequent attempt to aspirate more cystic fluid, CSF was obtained. Nonetheless, 6 ml of absolute alcohol was instilled into the cyst space and 5 ml was withdrawn 5 minutes later. No CSF leak was noted immediately postoperatively. Nevertheless, the patient awoke in the recovery room completely blind in both eyes, and she suffered a seizure that night. She began a regimen of high-dose steroid drugs and phenytoin. An MR image obtained with and without Gd on postoperative Day 2 demonstrated resolution of the sellar mass and a normal optic chiasm; however, there was evidence of edema in both frontal lobes and in the left temporal lobe accompanied by meningeal enhancement consistent with chemical meningitis (Fig. 2). A follow-up MR image obtained 2 months later demonstrated continued resolution of the cyst.
Complications in recurrent Rathke cleft cyst

Outcome. When the patient presented to our neuroophthalmology clinic 2 months after her last surgery, she was found to be anosmic bilaterally and she also had no light perception bilaterally, even with the brightest indirect ophthalmoscope. Her pupils were dilated and fixed at 7 and 6.5 mm OD and OS, respectively, and she had a left lid ptosis of 2.5 mm relative to the right eyelid. Additionally, her left eye showed a 35 diopter exotropia reflecting a paresis of adduction, and her optic discs showed devastating optic nerve atrophy. In summary, the patient demonstrated damage to the first and second cranial nerves bilaterally, and a partial third cranial nerve deficit on the left side. Her condition has remained the same to this day.

Discussion

Rathke cleft cysts are commonly believed to be derived from the remnants of the Rathke pouch, which is a dorsal diverticulum of the stomodeum.2,9,23 Other theories for the genesis of Rathke cleft cysts include derivation from the neuroepithelium,5,25 derivation from endoderm,10 reverse metaplasia of pituitary precursor cells,19 and a shared epithelial origin with craniopharyngiomas at differing historical stages of maturation.14,28 Rathke cleft cyst is a common finding at autopsy, with a reported prevalence of 13 to 33%.8,15,23,24 These cysts, however, are rarely symptomatic. In one report the authors estimate that, among symptomatic pituitary masses, Rathke cleft cysts account for only 1%,18 and in a second report 2% is indicated.22 A review of the literature revealed only 35 reported cases of symptomatic Rathke cleft cyst by 1977,28 after the advent of CT and MR imaging, the number of reported cases increased to 155 cases by 1991.26 Since 1991, there have been several additional case series,6,12,17,18,20,22 the increase in the number of reported cases is largely attributed to the advent and increasing popularity of modern neuroimaging modalities, particularly MR imaging.6,26,27

When the disease is symptomatic, patients present with complaints related to the expanding mass of the Rathke cleft cyst. Even though the exact incidences of the most common presenting signs and symptoms vary from report to report, they universally include headaches, visual disturbances, and endocrine abnormalities.1,6,9,17,18,20,26 Nevertheless, despite modern neuroimaging methods the preoperative diagnosis of Rathke cleft cyst is still problematic. In one report an MR imaging finding known as the "posterior ledge sign" has been described as pathognomonic for Rathke cleft cyst by 1977;28 after the advent of CT and MR imaging, the number of reported cases increased to 155 cases by 1991.26 Since 1991, there have been several additional case series,6,12,17,18,20,22 the increase in the number of reported cases is largely attributed to the advent and increasing popularity of modern neuroimaging modalities, particularly MR imaging.6,26,27

Intraoperative diagnosis based on frozen-section prepara-
tions is important because of the relatively conservative relationship to craniopharyngiomas.11,14,18,22,28 Although in this report is unclear, the authors postulate that the true recurrence rate of these lesions may be underestimated or that Rathke cleft cyst may, in fact, be misdiagnosed as craniopharyngiomas because of confusion regarding the diagnosis of the two entities.

In this report we present a woman with Rathke cleft cyst whose history and postoperative course are unique in several regards. This patient experienced three recurrences of the Rathke cleft cyst despite conventional transsphenoidal drainage and cyst biopsy/excision. On the final recurrence, absolute alcohol was instilled into the cystic space and it entered the CSF, resulting in bilateral anosmia, blindness, and partial left third cranial nerve damage.

This case history is the first in which multiple Rathke cleft cyst recurrences are reported following transsphenoidal cyst drainage and attempted cyst wall excision after each recurrence. It is important to note that the recurrences may be linked to the unsuccessful attempts to excise part of the cyst wall. Some authors have suggested that Rathke cleft cysts are more likely to recur after drainage of the cyst alone without partial excision of its wall,26 although Rathke cleft cysts have been known to recur despite successful partial wall resection and even after absolute alcohol cautery.12,22 It has also been suggested that these lesions may be prone to recurrence if there is a solid component of the cyst that is composed of stratified squamous epithelium, which may indicate a more aggressive lesion with a closer relationship to craniopharyngiomas.11,14,18,22,28 Although in our patient there was evidence of a solid component of the Rathke cleft cyst on MR imaging, it was never verified through histological examination of surgical specimens.

Relatively conservative treatment of Rathke cleft cyst with drainage and partial cyst excision is recommended because of the relatively benign nature of the lesion and the fear of iatrogenic damage to the pituitary.5,17,18,22,26 Nevertheless, it has been suggested that in cases of more aggressive, recurrent lesions, wider excision and postoperative radia-

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tion treatment may be beneficial. In two studies the use of absolute alcohol to cauterize the cyst has been reported to lessen the likelihood of recurrence. Alcohol leads to cell death by cell-membrane lysis, protein denaturation, and vascular occlusion; it has been used extensively elsewhere to sclerose and obliterate other kinds of tumorous or infectious lesions, including hepatic, renal, bone, and thyroid cysts. In the 59 patients included in the two aforementioned reports, there was only one recurrence of the Rathke cleft cyst. In both reports the authors advocate alcohol instillation if the cyst is patent and if there is no direct communication with the CSF. Additionally, in the report by Kleinschmidt-DeMasters, et al., those authors said that they routinely repair the sellar floor with a bone strut and Gelfoam prior to closure.

Absolute alcohol was instilled into our patient on her third cyst’s recurrence. Although a CSF leak was not noted after the operation, a communication had been created with attempted aspiration of the cyst that yielded CSF, and this communication likely persisted. It is also possible that the absolute alcohol itself eroded through the cyst lining into the CSF. In any event, the postoperative MR image and the clinical picture demonstrate leakage of the alcohol into the CSF space, resulting in chemical meningeval irritation and irreversible damage to three cranial nerves. This is the first report of such complications after this procedure.

Even though the treatment of recurrent Rathke cleft cysts remains controversial, given the devastating complication suffered by our patient, we advocate extreme caution when attempting absolute alcohol ablation of a cyst. In particular, meticulous attention must be paid to ensure that there is no communication between the cyst and the CSF. If there are any signs or suspicions of a CSF leak, this should be a contraindication to the instillation of absolute alcohol.

References


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