An isolated primary Rathke’s cleft cyst in the cerebellopontine angle

Case report

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Rathke’s cleft cysts (RCCs) are benign cysts typically located in the sellar or suprasellar region; ectopic isolated lesions are extremely rare. The authors describe the case of a 25-year-old man with a giant symptomatic RCC arising primarily at the cerebellopontine angle (CPA), only the second case reported thus far. The patient presented with a 2-year history of right hearing impairment and tinnitus accompanied by vertigo and headache and a 2-week history of right facial numbness. Subsequently, he underwent total cyst removal via retrosigmoid craniotomy with a good recovery. He experienced no recurrence during a 64-month follow-up period. The possible pathogenesis, differential diagnosis, and surgical treatment of such cysts are discussed in this article. Isolated ectopic RCCs can arise from the ectopic migration of Rathke’s pouch cells during the embryonic period. It is still difficult to distinguish ectopic RCCs from other cystic lesions of the CPA given the lack of specific imaging features. Aggressive resection of the cyst wall is not recommended, except when lesions do not closely adhere to adjacent structures.

KeY WorDS • cerebellopontine angle • Rathke’s cleft cyst • Rathke’s pouch • suboccipital retrosigmoid craniotomy • oncology

Rathke’s cleft cysts (RCCs) are believed to develop in the infundibulohypophyseal axis and account for 13%–33% of incidentally encountered pituitary lesions. These cysts frequently occupy the sella or suprasellar space; occasionally they extend to the sphenoid sinus, planum sphenoidale, or clival region. However, isolated ectopic RCCs are extremely rare, and only one case at the cerebellopontine angle (CPA) has been reported in the literature. Herein, we present another case of RCC arising primarily from the CPA and discuss its pathogenesis, clinical features, and surgical treatment.

Case Report

Clinical Presentation. A 25-year-old man was referred to our institution with a 2-year history of progressive hearing impairment and tinnitus on his right side, accompanied by intermittent vertigo and headache. These symptoms worsened over 6 months, and the patient had experienced right-sided facial numbness for 2 weeks. He did not demonstrate any other neurological deficits. Neurological examination revealed that he had hearing loss in the right ear (Gardner-Robertson Class IV), a decreased pinprick sensation in the V2 and V3 distributions of the right trigeminal nerve, and a positive Romberg sign.

Radiological Aspects. Computed tomography scanning revealed a hypodense regular lesion at the right CPA. No enlargement or erosion of the internal auditory canal was detected (Fig. 1A). Magnetic resonance imaging revealed a 38 × 30 × 25-mm homogeneous sharp-edged ovoid cyst without contrast enhancement at the right CPA, extending from the tentorium downward to the foramen magnum (Fig. 1D–F). The cyst had CSF-like signal intensity on both T1- and T2-weighted images (Fig. 1B and C). It exerted marked compression on the adjacent brainstem and cerebellum, but there was no sign of peritumoral edema or hydrocephalus. The differential diagnosis included cystic vestibular schwannoma, epidermoid cyst, and arachnoid cyst.

Abbreviations used in this paper: CN = cranial nerve; CPA = cerebellopontine angle; RCC = Rathke’s cleft cyst.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
Rathke’s cleft cyst in the cerebellopontine angle

**Operation and Pathological Findings.** The patient underwent cyst resection via a right suboccipital retrosigmoid craniotomy. A huge translucent cystic lesion containing milky white fluid was observed during the operation. The cyst loosely adhered to cranial nerves (CNs) V, VIII, and IX as well as the brainstem and cerebellum (Fig. 2A and B). The cyst contents were completely evacuated, and total removal of the cyst wall was achieved after careful dissection (Fig. 2C and D). Pathological examination revealed that the cyst was lined partially by single-layer columnar epithelium and partially by stratified squamous epithelium without keratinization (Fig. 3). These findings confirmed the histological diagnosis of RCC.

**Postoperative Course.** Postoperatively, the patient recovered well without any complication. The preoperative tinnitus, vertigo, headache, and facial numbness disappeared within 2 weeks. His hearing improved to Gardner-Robertson Class II at 3 months after surgery. The most recent 64-month follow-up MRI study did not show any evidence of cyst recurrence (Fig. 4).

**Discussion**

Rathke’s cleft cysts are benign, epithelium-lined cysts filled with mucous or caseous material. These cysts are thought to arise from the remnants of Rathke’s pouch during the embryological migration. They are regarded as the most common incidentally found lesions in the sellar region, followed by pituitary adenomas. Sporadic RCCs located in the nasopharynx, sphenoid sinus, or clivus have been described by other authors. However, it seems more appropriate to consider these lesions as an extension of intrasellar or suprasellar RCCs because of their close relationships with the originating site of Rathke’s pouch. Khalatbari et al. also suggested that most of the ectopic craniopharyngiomas described in past publications are essentially various extensions of large suprasellar lesions. In this regard, ectopic RCCs primarily developed beyond the craniopharyngeal canal are rather rarely reported in the literature. To our knowledge, only Zhou et al. have reported an isolated case in the CPA.

The origin and pathogenesis of ectopic RCC remains unclear, but some authors have pointed out the possible continuum between RCCs and craniopharyngiomas. Multiple cases of the isolated CPA craniopharyngioma have been reported. Therefore, it is reasonable to assume that the formation mechanism of ectopic RCC is similar to that of craniopharyngioma. Rathke’s pouch arises as an invagination of the primitive oral cavity and fuses with the developing neural tube at the early embryological period. The migration path of Rathke’s pouch forms the craniopharyngeal duct; accordingly, the cells of Rathke’s pouch proliferate and rotate to form the anterior pituitary gland. Because of their different rates during the rotation, most Rathke’s pouch cells spread throughout the sellar region. However, a few cells may migrate outside the sellar region. Under such circumstances, the failure of these ectopic cells to regress during development may lead to the formation of an ectopic RCC.

**Fig. 1.** Preoperative imaging of the ectopic RCC. A: Axial CT scan showing a hypodense mass in the right CPA without enlargement or erosion of the internal auditory canal. B and C: Homogeneous sharp-edged cystic mass with hypointense signal on axial T1-weighted MR image and hyperintensity on axial T2-weighted MR image. D–F: Axial, coronal, and sagittal Gd-enhanced MR images showing the lesion extending from the tentorium downward to the foramen magnum without contrast enhancement.
Our pathological examination showed that the cyst was lined partially by columnar epithelium and partially by stratified squamous epithelium, which overlapped histological features with the craniopharyngioma. Ikeda and Yoshimoto also reviewed 42 patients with RCCs and found that 3 cases had squamous epithelium in the cyst wall similar to that observed in craniopharyngiomas. These findings may support the hypothesis of a continuum between RCCs and craniopharyngiomas. Additionally, some authors have considered the squamous metaplasia as one of the factors affecting the recurrence of RCC. Aho et al. reported 118 surgical cases of RCC with a recurrence rate of 18%. They found that 32% of the patients with squamous metaplasia had a recurrence, whereas only 11% of those without metaplasia experienced recurrent events. We did not find the association between the recurrence and squamous metaplasia in this case, but more case studies are needed to clarify this association.

The variable clinical manifestations of RCCs mainly depend on the anatomical location of the cysts that may cause mass effect on adjacent structures. For lesions of the sellar region, specific symptoms typically present as visual dysfunctions, pituitary hormone deficits, or diabetes insipidus. Likewise, patients with ectopic lesions can present with other symptoms specific to cyst locations. For example, our patient had hearing impairment, tinnitus, and vertigo, symptoms consistent with those for other CPA lesions.

The combination of CT and MRI studies may improve...
the preoperative diagnosis of RCC. However, the differential diagnosis of RCC from other cystic lesions in the CPA remains difficult. Typically, the RCC has an oval or dumbbell shape, a variable intensity on MRI depending on the cystic content, and little or no cyst wall enhancement without any signs of invasion. The vestibular schwannoma is the most common tumor of the CPA and usually has a characteristic CT manifestation of enlargement or erosion of the internal auditory canal. In addition, enhanced tumor components are frequently found on MRI. Epidermoid cysts often present as a lobulated mass similar to CSF on CT or MRI, and calcification can sometimes be detected. They do not enhance but can exhibit restricted diffusion on diffusion-weighted imaging. The arachnoid cyst has a clearly outlined border, with signal intensity similar to that of CSF on MRI. No calcification and postcontrast enhancement can be observed in such cysts.

Most authors recommend surgical removal for the treatment of symptomatic RCCs. However, the extent of resection of the cyst wall remains controversial. Although our earlier experience showed that radical removal would not necessarily reduce recurrence and in fact could increase the risk of postoperative complications, we still performed a total resection in the featured case. The reason is that the cyst wall in this case could be easily dissected without inflicting unnecessary damage to surrounding structures. In contrast, for cyst walls closely adhered to adjacent structures, a conservative surgical strategy is recommended for better functional outcomes.

Conclusions

We have described an isolated primary RCC at the CPA, the second such case in the literature. Lesions such as this can arise from ectopic migration of Rathke’s pouch cells during the embryonic period. Although the combination of CT and MRI is helpful, it is still difficult to distinguish between ectopic RCCs and other cystic lesions of the CPA. Aggressive resection of the cyst wall is not recommended except for lesions that are not closely adhered to adjacent structures.

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Disclosure

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