Enlarging pediatric ectopic Rathke's cleft cyst in the prepontine cistern: case report

Takamasa Kamei, MD,¹ Masahiro Nonaka, MD, PhD,¹ Yoshiko Uemura, MD, PhD,²
Yasuo Yamanouchi, MD, PhD,¹ Yumiko Komori, MD,¹ Ryoichi Iwata, MD,¹ Junichi Takeda, MD,¹
Tetsuo Hashiba, MD, PhD,¹ Kunikazu Yoshimura, MD, PhD,¹ and Akio Asai, MD, PhD¹

Departments of ¹Neurosurgery and ²Pathology and Laboratory Medicine, Kansai Medical University, Hirakata, Osaka, Japan

Rathke's cleft cyst is a cystic disease that occurs in the sella turcica or, occasionally, in the suprasellar area. An ectopic Rathke's cleft cyst is extremely rare, and its nature is less well understood. The authors report the case of a 14-year-old girl who presented with a growing cystic lesion in the prepontine cistern, immediately behind the dorsum sellae. Preoperative imaging and intraoperative investigation showed part of the cyst wall continuing into the dorsum sellae, to the pituitary gland. The cisternal portion of the cyst wall was totally resected via a right subtemporal approach. Histopathological examination of the cyst wall showed a monolayer of ciliated cells, identical to those of Rathke's cleft cyst. To the best of the authors' knowledge, this represents the first pediatric case of Rathke's cleft cyst occurring in the prepontine cistern.

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RATHKE's cleft cyst mostly occurs in the sella turcica and suprasellar areas. Occurrence in other areas is rare, representing ectopic Rathke's cleft cyst. Normally, Rathke's pouch closes in early embryogenesis, and Rathke's cleft cyst arises when a remnant of the pouch grows between the pars distalis and pars nervosa of the pituitary.¹³ We encountered a case of enlarging pediatric ectopic Rathke's cleft cyst in the prepontine cistern.

Case Report

The patient was a 14-year-old girl with no history of disease. MRI, performed because she was experiencing headaches and dizziness, revealed a circular cystic mass measuring 14 × 10 × 9 mm in the prepontine cistern behind the dorsum sellae and a spinous structure extending

to the pituitary gland. The cyst was located in the upper half of the prepontine cistern. It appeared hypointense on T1-weighted images and isointense on T2-weighted images, with no areas of high intensity on diffusion-weighted images and no contrast enhancement (Fig. 1). We carefully followed this patient, and another MRI study 10 months later revealed enlargement of the cystic lesion to 18 × 13 × 10 mm, as well deformation of the original shape. FLAIR imaging showed trabecular structures reaching the cyst from the pars intermedia of the pituitary (Fig. 2). DICOM (Digital Imaging and Communications in Medicine) data of the MRI studies were transferred to iPlan 2.5 software (Brainlab). Several trajectories for surgical approaches were then determined, including transsylvian, suboccipital, transclival, and subtemporal routes. We decided to

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FIG. 1. Imaging at first visit. **A:** Axial T1-weighted image showing a homogeneous hyperintense round mass in the prepontine cistern (arrow). **B:** Axial T2-weighted image demonstrating a low-intensity mass (arrow). **C:** Diffusion-weighted image showing no signal-hyperintense content in the cyst (arrow). **D:** Sagittal T1-weighted image revealing a "spur" pointing to the dorsum sellae (arrow).

resect the cyst via a right subtemporal approach. After gently retracting the temporal lobe, the arachnoid of the prepontine cistern was incised. The yellowish-white cyst was detected in the prepontine cistern, which was filled with milky-white viscous fluid. After removing the cyst contents by suction, we totally resected the cisternal portion of the cyst wall (Fig. 3).

Histopathological examination showed the cyst wall to be lined by ciliated epithelium. The histopathological diagnosis was Rathke's cleft cyst (Fig. 4). The patient's postoperative course was uneventful. Her preoperative symptoms resolved after surgery, and as of the time of writing,

FIG. 3. Intraoperative photographs. **A:** A yellowish-white cyst is observed in the prepontine cistern. **B:** Collapsed cyst wall showing attachment to the dorsum sellae. Asterisk shows the cyst; white circle, the posterior cerebral artery; white square, the temporal lobe; black square, the collapsed cyst wall; and white triangle, the dorsum sellae. Figure is available in color online only.

she has remained symptom free for a year. Postoperative MRI performed 3 months after surgery suggested total resection of the lesion in the cisternal portion (Fig. 5).

Discussion

Among the various types of nonneoplastic intracranial cystic disease, Rathke's cleft cyst is one of the most frequently encountered clinically. Most are located in the sella turcica and suprasellar regions. The authors of most previous reports of ectopic Rathke's cleft cyst have described extracranial lesions, located in the nasopharynx or sphenoid sinus.³ To date, only 3 reports appear to have described intracranial ectopic Rathke's cleft cyst. Two lesions were located at the cerebellopontine angle,^{6,16} including one pediatric case,¹⁶ and in one adult case the lesion was located in the prepontine cistern¹² (Table 1).

The cyst in our case was located in the upper half of the prepontine cistern. MRI showed a trabecula-like structure reaching the cyst from the interbulbar cleft on FLAIR imaging. We considered this finding as suggestive that the cyst originated in Rathke's cleft and grew into the prepontine cistern. Extension of the suprasellar Rathke's cleft

FIG. 2. **A:** Preoperative sagittal FLAIR image showing enlargement and distortion of the cyst shape compared with the image obtained on the first visit. Note the part of the cyst hanging over the dorsum sellae (arrow). **B:** Preoperative coronal heavily weighted T2 image revealing the location of the cyst (arrow) in the prepontine cistern.

FIG. 4. Photomicrograph showing the cyst wall lined by ciliated epithelium (arrows). H & E, original magnification $\times 100$. Figure is available in color online only.

cyst to the prepontine cistern has been reported previously.⁵ However, the cystic mass in our case was not found in either the supra- or intrasellar regions.

The natural course of ectopic Rathke’s cleft cyst has not been described. Furthermore, there are no cases in which enlargement of the cyst was captured on images. Our report offers the first documentation of enlargement of the prepontine cyst in a pediatric case approximately 1 year after initial MRI.

Other cystic diseases, such as colloid cyst,¹¹ epithelial cyst,^{8,10} and neurenteric cyst, show similar histopathological characteristics.⁹ Neurenteric cysts are known to occur on the ventral side of the spinal cord, along the course of the notochord. Some reports have described very rare neurenteric cysts occurring at the cerebellopontine angle, craniovertebral junction, and lower prepontine regions.^{1,2,7} The cyst in our case was located only in the upper prepon-

FIG. 5. Axial (A) and sagittal (B) T2-weighted images obtained 3 months after surgery, showing no residual lesion in the prepontine cistern.

tine region, where neurenteric cysts have not previously been reported.

Various surgical approaches have been applied to access prepontine lesions, including lateral suboccipital and transsylvian routes.^{8,10–12} Since the cyst in our case was located in the upper half of the prepontine cistern, we carefully selected the surgical approaches. We applied iPlan surgical planning software and used the “trajectory view” to determine the surgical approach (Fig. 6).¹⁵ If the transsylvian approach is to be selected, the posterior clinoid process and dorsum sellae would need to be drilled off to allow visualization of the cyst. Drilling of the posterior clinoid process needs to be performed with extreme care to avoid injuring the posterior communicating artery and oculomotor nerve. Furthermore, opening of the venous plexus in the posterior clinoid process risks venous hemorrhage,⁴ which is not easy to control once it occurs. If a lateral suboccipital approach is used, brainstem retraction is required to reach the lesion, since the cyst is located behind the brainstem. Retraction of the brainstem carries a risk of damaging this structure, with disastrous consequences. Another approach is to reach the skull base where the cyst is located via the nasal cavity, using an endoscope. The transclival endoscopic endonasal approach may be useful, but it requires a high degree of technical skill to drill the thick clival bone appropriately, avoid damage to the normal pituitary gland, and prevent cerebrospinal fluid leakage.¹⁴

We therefore considered the subtemporal approach as useful for resection of the cyst located in the upper half

TABLE 1. Summary of intracranial ectopic Rathke’s cleft cysts described in the literature

Authors & Year	Age (yrs), Sex	Symptoms	Lesion Size (mm)	MRI Features	Lesion Location	Histopathological Finding
Fan et al., 2014	25, M	HA, vertigo, tinnitus, hearing impairment	38 × 30 × 25	T1: low intensity; T2: high intensity; CE	Cerebellopontine angle	Single-layer columnar epithelium
Kim, 2012	41, F	HA	25	T1: isointense; T2: hypointense; CE	Upper half of the prepontine cistern	Ciliated columnar epithelium
Zhou et al., 2010	12, F	HA, vertigo, nausea, vomiting	Not available	T1: hyperintense; T2: hypointense; CE	Cerebellopontine angle	Simple ciliated columnar epithelium
Present case	14, F	HA, dizziness	14 × 10 × 9 (→ 18 × 13 × 10)*	T1: low intensity; T2: high intensity; CE	Upper half of the prepontine cistern	Lined by ciliated epithelium

CE = indication that there was no contrast enhancement of the cyst wall; HA = headache.

* Parenthetical value indicates the size the lesion grew to 10 months after initial imaging.

FIG. 6. Trajectory view of the reconstructed MR images. **A:** Trajectory of the transsylvian approach (white solid arrow). Dotted arrows show the posterior clinoid process. **B:** Trajectory of the lateral suboccipital approach (white solid arrow). **C:** Trajectory of the endonasal transclival approach (white solid arrow). **D:** Trajectory of the subtemporal approach (white solid arrow).

of the prepontine cistern. This approach does not require drilling of any deep-seated bones or retraction of the brainstem. Although retraction of the temporal lobe is necessary with the subtemporal approach, this approach allows observation of the entire cyst. In addition, it is important to prevent contusion of the temporal lobe by shortening retraction time, gently retracting the temporal lobe, and achieving sufficient drainage of cerebrospinal fluid from the lateral ventricle.

Conclusions

We have reported a pediatric case of enlarging ectopic Rathke's cleft cyst in the prepontine cistern. To our knowledge, no pediatric case of Rathke's cleft cyst enlarging only in the prepontine cistern has been reported previously. Total resection of the cisternal portion of the cyst was successfully achieved using a subtemporal approach. Since cyst remnants may be present, we have recommended annual follow-up imaging for this patient.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Nonaka, Yamanouchi. Acquisition of data: Nonaka, Kamei, Uemura, Yamanouchi. Analysis and interpretation of data: Nonaka, Kamei, Komori, Asai. Drafting the article: Nonaka, Kamei, Asai. Critically revising the article: Nonaka, Uemura, Yamanouchi, Iwata, Takeda, Hashiba, Yoshimura, Asai. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Nonaka. Study supervision: Nonaka, Asai.

Correspondence

Masahiro Nonaka, Department of Neurosurgery, Kansai Medical University, 2-3-1, Shinmachi, Hirakata City, Osaka 573-1191, Japan. email: nonakamasa65@gmail.com.