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Case Report

Combined endoscopic endonasal transtubercular and transclival approaches for large neurenteric cyst in posterior cranial fossa: A case report and literature review

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ABSTRACT

Background: Intracranial neurenteric cysts (NCs) are extremely rare tumors that more commonly involve the posterior fossa than any other cranial part. While transcranial skull base surgery has been the mainstay of treatment, the utility of endoscopic transnasal surgery (ETS) remains to be established.

Case Description: We report a case of a large posterior fossa NC extensively involving the suprasellar region, cerebellopontine angle, and prepontine cistern, which we successfully resected with ETS through a combination of transtubercular and transclival routes. Before surgery, the patient presented with abducens nerve and pseudobulbar palsies, which resolved within 2 weeks postoperatively. The patient remained free from recurrence for 3 years postoperatively.

Conclusion: Extended ETS may offer a minimally invasive option for the posterior fossa NC, extensively occupying the ventral space of the brainstem.

Keywords: Endoscopic transnasal surgery, Extended endoscopic transnasal surgery, Neurenteric cyst, Posterior fossa, Skull base tumor

INTRODUCTION

Neurenteric cysts (NCs) are rare benign congenital lesions that most commonly arise in the spinal canal, especially at the cervical and upper thoracic levels.[13,22,25,31] Intracranial NCs are extremely rare, with the most common locations being the posterior fossa and craniovertebral junction,[13] and typically present with headache and location-specific symptoms.[25,37] Radiographic characteristics are an oblong, sharply demarcated cyst with smooth or lobulated margins, usually appearing hyperintense on both T1- and T2-weighted images of magnetic resonance imaging (MRI) with possible contrast enhancement.[6,7,24,25,29] Signal intensity varies because of differences in the protein content of cyst fluid.[5,6]

Pathologically, the cyst is lined with nonciliated gastrointestinal-type epithelium and ciliated respiratory-type epithelium, which is poor in mucin-producing cells, or a mixture of both.

[10,24,26,29] As the posterior fossa NC is usually soft and easily removable, surgical resection is the first choice of treatment. [1,10,18,32,34,37] While the previous literature on NCs favors transcranial approaches, techniques of endoscopic skull base surgery have advanced over the past few decades, enabling minimally invasive approaches to tumors located ventral to the brainstem. However, to date, no report has described extended endoscopic transnasal surgery (eETS) for large NCs, thus, it remains unclear whether ETS is safely applicable for posterior fossa NCs, especially if it involves a wide range of skull base regions. To address this issue, we report a case of a large posterior fossa NC that was successfully treated using eETS.

CASE PRESENTATION

A 46-year-old male patient who suffered from progressive diplopia, dysphagia, and hoarseness was referred for the management of a 35 × 32 × 51 mm lobulated cystic lesion located ventral to the brainstem, extending from the suprasellar region to the pontomedullary junction, involving the oculomotor, trochlear, trigeminal, abducens, facial, and acoustic nerves [Figure 1a]. The mass appeared isointense on T1- and T2-weighted images without contrast enhancement, containing small areas showing hyperintensity on T1-weighted images and hypointensity on T2-weighted images [Figure 1b-d]. Given the absence of direct compression of the lower cranial nerves, patient's dysphagia and hoarseness were considered signs of pseudobulbar palsy due to brainstem compression. To achieve total resection under direct vision in a single surgical intervention, we selected eETS as the first choice of treatment.

ETS

Under standard general anesthesia, the patient was secured in the supine position. After a 3-point pin head holder was applied, the head was slightly rotated to the right and tilted to the left so that the nostrils were directed toward the surgeon. A surgical navigation system (StealthStation S7; Medtronic, Minneapolis, MN, USA) and electrophysiological monitoring of the extraocular muscles, cranial nerves (facial nerves and lower cranial nerves), auditory brainstem response, motor-evoked potential, and sensory evoked potential for all the extremities were established. During the surgery, a 4 mm diameter, 0° and 30° rigid neuroendoscope (Karl Storz Endoscopy Japan, Tokyo, Japan) and a robotic arm holder (Point Setter; Mitaka Kohki, Tokyo, Japan) were used.

After lateralizing the bilateral middle turbinates, vertical mucosal incisions were made on both sides of the nasal septum, and the submucosal dissection was advanced until the anterior bony wall of the sphenoid sinus was exposed. Next, a nasal speculum designed for ETS (Fujita Medical Instruments, Tokyo, Japan) was deployed in the submucosal space to obtain a wide surgical corridor without being disturbed by redundant nasal anatomical structures. The posterior edge of the bony septum was temporarily displaced during the surgery, and the sphenoid sinus was opened wide. The purpose of this approach was to ensure the smooth delivery of the surgical instruments and endoscopes without performing middle turbinectomy and posterior septostomy.[33] Then, the optic canal, tuberculum sellae, planum sphenoidale, carotid prominence, and clival recess were observed, and the skull base bones were carefully removed using a drill with a diamond bur. The ventral duramater was exposed to facilitate both transtubercular and transclival approaches [Figure 2a]. The suprasellar part of the mass was removed through the transtubercular route. Next, we approached the sellar floor and the lesion in the posterior cranial fossa was resected through the transclival route.

The cyst comprised a soft, grayish capsule filled with yellow mucinous and caseous components [Figure 2b]. During the procedure, we identified the optic chiasma, bilateral oculomotor, trigeminal, abducens, and facial nerves, and the posterior cerebral arteries, superior cerebellar arteries, and basilar artery [Figure 2c-e]. Maximum surgical resection was safely performed under direct vision [Figure 2f]. The skull base defects were reconstructed using gelfoam, with

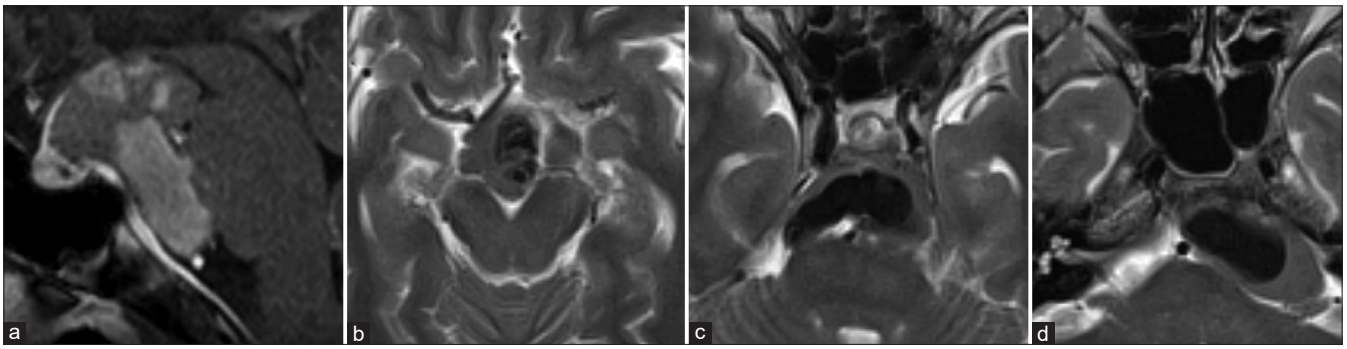

Figure 1: Magnetic resonance imaging reveals a large mass located ventral to the brainstem extending from the suprasellar region to the pontomedullary junction (a, contrast-enhanced T1-weighted image, sagittal plane; b-d, T2-weighted image, axial plane).

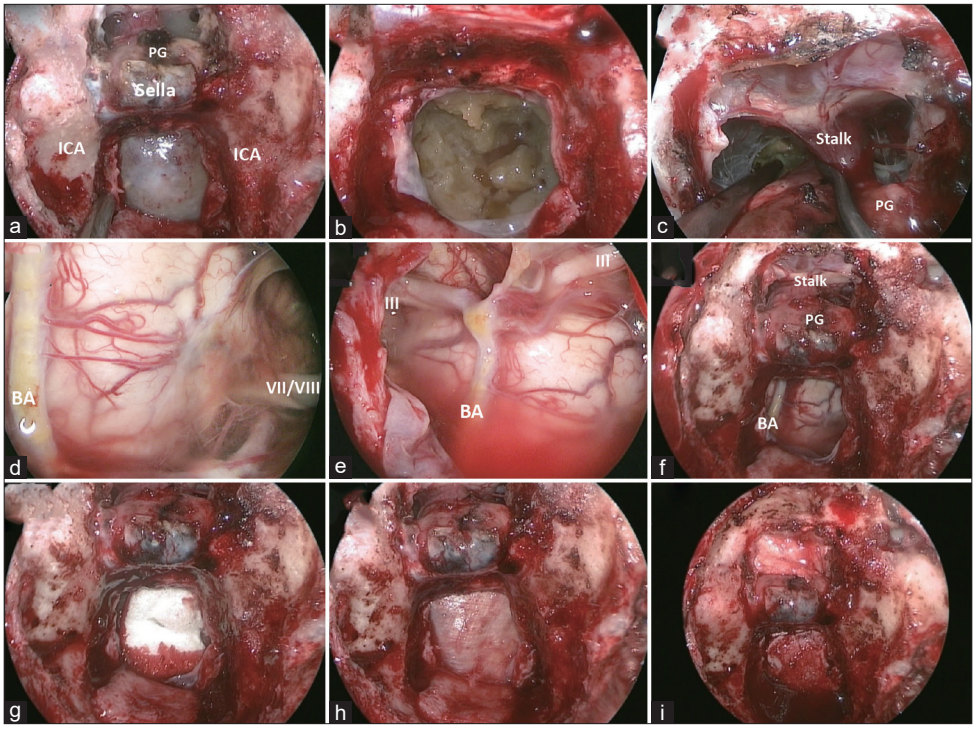

Figure 2: Suprasellar part of the mass is removed through the transtubercular route, and the lesion in the posterior cranial fossa is resected through the transclival route (a). The cyst is composed of soft grayish capsule, and yellow mucinous and caseous components are observed (b). During the procedure, we identified the stalk, pituitary gland (c), basilar artery, facial nerve, vestibulocochlear nerve (d), and oculomotor nerve (e). Surgical resection was enabled under direct vision (f). The skull base defects are reconstructed using gelfoam, in-lay and over-lay fascial grafts with abdominal fat pieces in a multilayer fashion (g-i).

in-lay and over-lay fascial grafts with abdominal fat pieces in a multilayered fashion [Figure 2g-i].[17] Lumbar drainage was performed before the procedure to facilitate healing of the dural defects. Cerebrospinal fluid (CSF) drainage was sustained using a pressure-control valve (Acty Valve II; Kaneka Medix Corporation, Osaka, Japan) for daily output within a range of 100–200 mL for 5 days.

The postoperative course was uneventful, and diplopia, dysphagia, and hoarseness resolved completely within 2 weeks. Histological examination revealed colloidal material and fragments of ciliated columnar epithelium, consistent with the diagnosis of NC [Figure 3a], and follow-up MRI demonstrated gross total resection with no sign of recurrence at the most recent follow-up, 3 years postoperatively [Figure 3b-d].

DISCUSSION

Herein, we report a case of a large posterior fossa NC extending from the suprasellar region to the pontomedullary junction. Localized NCs can be successfully treated using transcranial surgery. Lateral suboccipital approaches have been most frequently used to tackle posterior fossa NCs.[2,3,10,12,23,33,34] As NCs are commonly located in the midline, ventral to the brainstem, far lateral transcondylar/condylar fossa approaches are appropriate for optimizing the trajectory and minimizing intraoperative compression of the cerebellum. However, extensive NCs involving a wide range of ventral skull base regions may require more complicated skull base techniques, including labyrinthectomy and ligation of the sigmoid sinus, and a combination of multiple invasive approaches, which carries a significant risk of adverse events.[1,3,20,28,32,35] Despite the invasiveness of skull base approaches, complete removal of the cyst wall is difficult and is associated with high recurrence rates.[8,19,27,37] The risk of postoperative cranial nerve complications is not negligible. In addition, insufficient resection may lead to cyst leakage and potentially cause meningeal irritation, which may ultimately lead to symptomatic hydrocephalus. Sinus thrombosis has also been reported.[2,3,15,19,23,36]

In the past decade, the application of eETS has been further expanded to intradural lesions in the posterior cranial fossa, such as posterior fossa meningiomas, ventrally located brainstem cavernous malformations, schwannomas, chordomas, chondrosarcomas, and even vertebrobasilar aneurysms.[4,12,14,16,21] Nevertheless, regarding the posterior fossa NC, only three case reports are available, as shown in [Table 1].[9,11,30] All of these NCs were relatively small, localized lesions, and thus, a purely transclival approach was

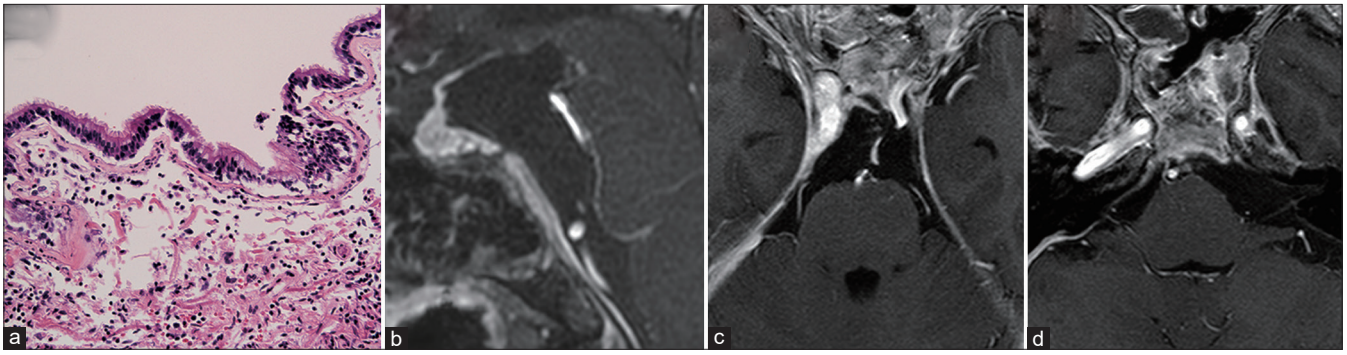

Figure 3: Histological examination demonstrates colloidal material and fragments of ciliated columnar epithelium, which indicates the diagnosis of neurenteric cyst (a, hematoxylin-eosin stain, ×40). Follow-up magnetic resonance imaging demonstrates gross total resection (b-d), with no sign of recurrence at 3 years after surgery.

Table 1: Previously published cases treated by endoscopic approach.

Author, year	Age (years)	Sex	Location	Diameter (mm)	Involved CNs	Trajectory	EOR	Complications	FU period	Recurrence
Cobb, 2010	37	Male	Prepontine cistern	20	None	Trans-clival	GTR	None	N/A	N/A
Prevedello, 2010	42	Male	Prepontine cistern	N/A	VI	Trans-clival	GTR	None	3 months	No
	47	Male	Prepontine cistern	22	None	Trans-clival	GTR	None	24 months	No
Fomichev, 2016	27	N/A	Intra-axial (pons)	20	None	Trans-clival	GTR	None	N/A	N/A
Present case	46	Male	Suprasellar cistern to prepontine cistern	46	III, VI, VII–X	Combined transtubercular and trans-clival	GTR	None	3 years	No

CN: Cranial nerve, EOR: Extent of resection, FU: Follow-up, GTR: Gross total resection, N/A: Not available

utilized. In contrast, the cyst in our case was much larger and extensive, involving a wide range of skull base territories, and thus, a combination of endoscopic transtubercular and transclival approaches was employed, resulting in successful removal without neurological complications. This demonstrated that eETS represents a reasonable option for posterior fossa NCs with wide accessibility and minimal invasiveness, although a larger case series is needed to determine its efficacy.

Although extensive skull base defects may increase the risk of postoperative CSF leak, our multilayered reconstruction method seems applicable to any size of the dural defect and can successfully prevent CSF leakage.[17] The patient remained free from tumor recurrence as of 3 years after surgery, and we are continuing clinical observation, as recurrence may occur even thereafter.[2,3,6,12,16,23]

CONCLUSION

For the posterior fossa NC located ventral to the brainstem, eETS can offer a useful option, especially when a wide range of skull base regions and associated cranial nerves is involved. The surgical field can be easily extended with a combination of transtubercular and transclival approaches. A larger case series is needed to determine the efficacy of eETS for extensive posterior fossa NCs.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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