CASE REPORT

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Surgical strategy for symptomatic pineal cyst: is endoscopit third ventriculostomy necessary in addition to cyst fenestration?

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ABSTRACT

Symptomatic large pineal cyst (PC) remains a rare entity. The stable natural course of asymptomatic PCs is well established. However, large cysts may cause pressure-related symptoms necessitating surgical intervention. The surgical strategy for symptomatic PCs is still controversial. Regardless of the approach, total resection of the cyst is not mandatory. The endoscopic approach allows cyst fenestration in patients with associated obstructive hydrocephalus. On the other hand, the necessity of simultaneous endoscopic third ventriculostomy (ETV) is still debatable. Here, we report a case of a woman who underwent endoscopic cyst fenestration, biopsy, and third ventriculostomy for a large symptomatic PC and discuss the surgical strategy. A 30-year-old woman presented with headache and diplopia, MRI showed a large PC and accompanying obstructive hydrocephalus. Simultaneous cyst fenestration, biopsy and ETV with endoscopy was successfully completed. She had an uneventful recovery period with immediate relief of symptoms. Although, the aqueduct was communicated due to cyst shrinkage, the patency of the third ventricular stoma was demonstrated in long-term follow-up scans. Based on clinical course of the present case, we concluded that ETV in addition to cyst fenestration should be considered necessary and beneficial in cases of large symptomatic PC with associated hydrocephalus whenever an endoscopic intraventricular approach is considered.

Keywords: pineal cyst, cyst fenestration, hydrocephalus, neuroendoscopy, endoscopic third ventriculostomy

Abbreviations:
PC: pineal cyst
ETV: endoscopic third ventriculostomy
MRI: magnetic resonance imaging
CSF: cerebrospinal fluid

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INTRODUCTION

Despite the high prevalence rate of up to 23% of pineal cyst (PC) in healthy adults,[1] symptomatic PC remains a rare entity.[2,3] Most PCs are found incidentally and remain stable over months or years.[4,5,6] However, larger cysts are more likely to be associated with symptoms due to pressure on the quadrigeminal plate and obstruction of the cerebral aqueduct causing hydrocephalus. Symptomatic cysts vary in size between 7 mm and 45 mm,[7] and surgical intervention may be necessary.[8,9]

The surgical strategy for symptomatic PC is still controversial. Recently, there has been a trend toward endoscopic approaches because of their reduced invasiveness as a surgical strategy for symptomatic PC. In addition to the reduced invasiveness of the procedure, endoscopic approaches provides an avenue for endoscopic third ventriculostomy (ETV) in addition to cyst fenestration.[10,11] However, the necessity of ETV has not yet been established because cyst fenestration causes the aqueduct to reopen. Here, we report a case of successful endoscopic cyst fenestration, biopsy, and ETV for a large symptomatic PC, and discuss the significance of simultaneous ETV.

CASE PRESENTATION

History and examination

A 30-year-old woman presented with a 6-day history of headache associated with diplopia for 2 days. The patient showed Parinaud's syndrome. Magnetic resonance imaging (MRI) of the brain revealed a large well circumscribed cystic mass, which dimensions and volume were 30×20×22 mm and 55.3 cm^3, respectively, with obstruction of the aqueduct and associated obstructive hydrocephalus (Fig. 1A,B). Serum alpha fetoprotein and β-human chorionic gonadotrophin levels were 5.0 ng/mL and < 0.50 mIU/mL, respectively. She was scheduled for biopsy and fenestration of the cystic tumor with simultaneous ETV.

Surgical Technique

An Institutional Review Board approval was not required for this study. Patient consent was obtained for this procedure. Under general anesthesia, the patient was placed in the neutral-supine position with the head fixed in anteflexion in a Sugita head holder. An endoscopic sheath (Neuroport™; Olympus Corporation, Tokyo, Japan) was introduced through a right frontal burr hole, and the lateral ventricle was reached under navigation guidance. A flexible endoscope (VEF-V; Olympus Corp., Tokyo, Japan) was then introduced into the third ventricle without scratching the fornix. The structures of the third ventricular floor and pineal region tumor were identified. The aqueduct was covered and obstructed by the tumor (Fig. 2A). The tumor was entirely cystic with very thin walls. Fenestration of the cyst using forceps and balloon yielded xanthochromic serous cyst contents (Fig. 2B), which was suggestive of a PC. Specimens of the cyst wall were obtained for histopathological evaluation. During this procedure, there was slight bleeding, which was easily controlled by irrigation. The aqueduct became patent with reestablishment of cerebrospinal fluid flow after the collapse of the tumor (Fig. 2C). Perforation of the floor of the already pulsating third ventricle to the interpeduncular cistern was performed with biopsy forceps (Fig. 2D,E). A balloon catheter was used to enlarge the perforation. The basilar artery, perforators, and posterior cerebral artery were confirmed through the fenestration (Fig. 2F). After confirmation of communication of the aqueduct, third ventricular floor pulsation, and hemostasis, the wound was closed in the usual manner using burr hole cap.

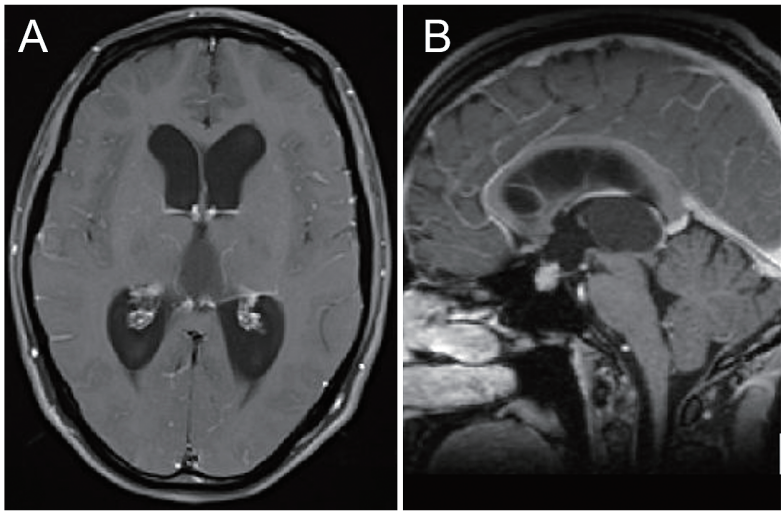

Fig. 1 Preoperative MRI
Axial (**A**) and sagittal (**B**) T1-weighted MRI with gadolinium enhancement demonstrated a large non-enhanced well circumscribed mass in the pineal region, which dimensions and volume were 30×20×22 mm and 55.3 cm^3, respectively. The mass was seen to obstruct the aqueduct with pressure on the quadrigeminal plate and associated hydrocephalus.

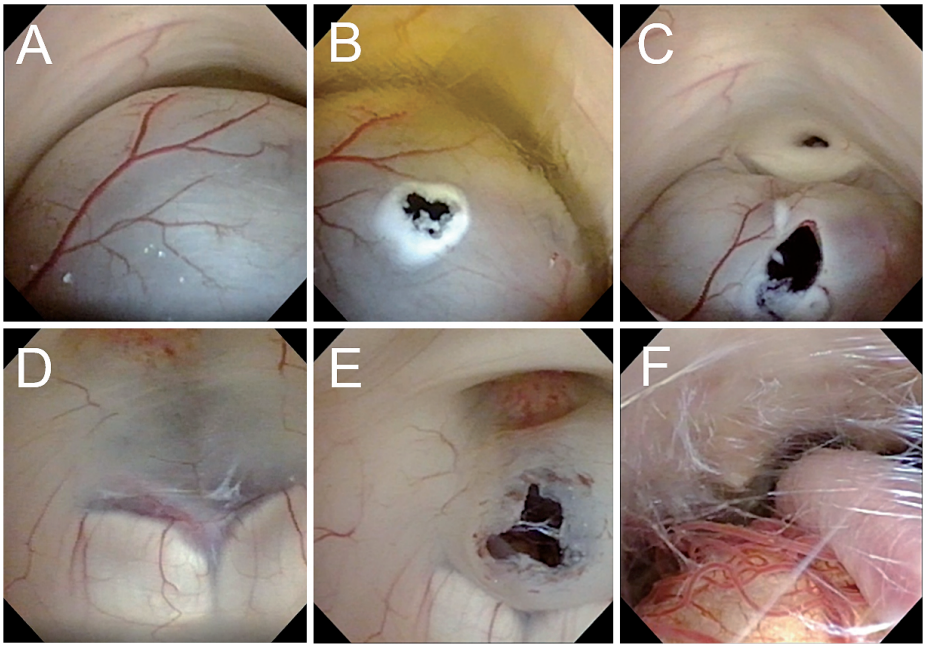

Fig. 2 Intraoperative endoscopic view
Intraoperative endoscopic images showing obstruction of the aqueduct by the large cyst (**A**). Fenestration of the cyst was performed with drainage of xanthochromic serous content (**B**) with collapse of the cyst and opening of the aqueduct (**C**). Perforation of the third ventricular floor into the interpeduncular cistern was done through the thin tuber cinereum (**D,** E) with visualization of the vessels through the fenestration (**F**).

Postoperative course

The patient had an uneventful recovery period with immediate relief of symptoms. There were no postoperative complications. There was no evidence of malignancy such as nuclear atypia, cellular pleomorphism and foci of hemosiderin pigment with histopathological examination, which confirmed the lesion as a PC. Follow-up brain MRI at 18 months after surgery showed progressive regression of the cyst size with normal ventricular sizes (Fig. 3 A,B). The patency of the third ventricular stoma was also demonstrated in long-term follow-up scans (Fig. 3 C,D).

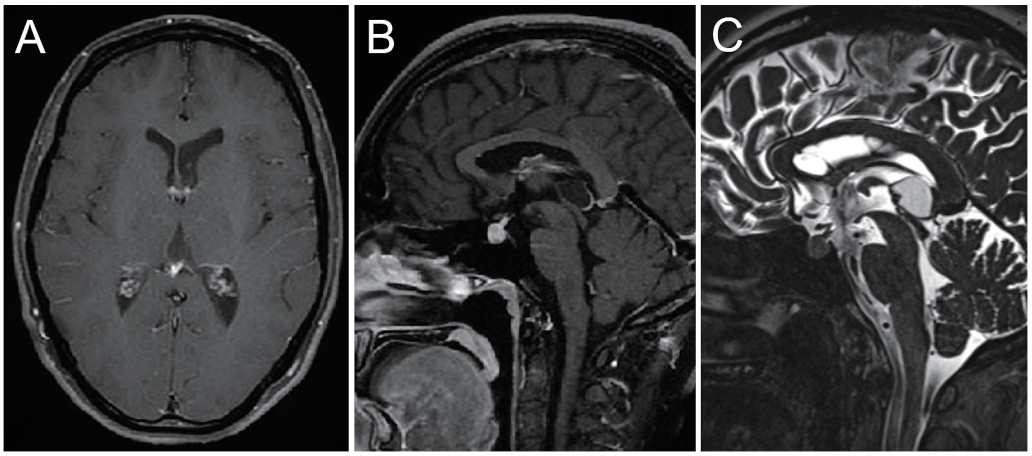

Fig. 3 Postoperative MRI
Postoperative MRI at 18-month (**A, B**) follow-up demonstrated progressive regression of the cyst and absence of hydrocephalus. The third ventricular stoma was not occluded (**C**), and the CSF patency of the third ventricular stoma is indicated (arrow, **D**).

DISCUSSION

The natural course of PCs in asymptomatic individuals is usually benign[12] and may not require long-term follow-up,[6,13,14] although a recent study by Starke et al[15] suggested that lesions showing increases in size, contrast enhancement, or hemorrhage on MRI were predictors of malignancy and therefore required serial MRI follow-up with a lower threshold for neurosurgical intervention. Surgical treatment may be required for symptomatic large PCs.[10]

The symptoms are mostly due to mass effects on the quadrigeminal plate leading to compression of the superior colliculi with resultant Parinaud's syndrome. Furthermore, occlusion of the cerebral aqueduct results in obstructive hydrocephalus. Headache is the most common symptom experienced by patients.[12,13,16,17,18] This was also present in our patient and may have been attributable to intracranial pressure from hydrocephalus. However, in the absence of hydrocephalus, headache could result from compression of veins in the pineal region.[19] These symptoms related to mass effects are strong indications for surgical treatment of PC.[4,20]

The surgical strategy for symptomatic PC, which is an extremely rare entity, is controversial. However, total resection of the cyst is not mandatory.[11] Surgical approaches reported in the literature include microsurgical fenestration/resection via the infratentorial supracerebellar[2,12,16,20] or occipital-transtentorial approach,[8,20] stereotactic aspiration,[16,21,22] and the endoscopic transventricular

approach.[3,16,20] The endoscopic infratentorial supracerebellar approach has also been reported.[23] There has been an increase in utilization of endoscopic surgery especially in cases with the presence of hydrocephalus[16] because it allows both fenestration with biopsy under direct visualization of the cyst and ETV.[12,24,25,26] However, the clinical experience of the surgeon in endoscopic procedures is a major factor in guiding the application of this approach.[16]

The various endoscopic intraventricular approaches, including ETV, cyst fenestration or both, were considered in this case due to associated hydrocephalus and the reduced invasiveness of the procedure. ETV is effective for obstructive hydrocephalus associated with PCs. David et al reported that 14 patients of symptomatic PC were treated with only ETV. Among them, 3 patients (21.4%) recurred, which suggested that only ETV might not be satisfactory for treatment of symptomatic PCs.[27] On the other hand, cyst fenestration with drainage of its contents resulted in reopening of the cerebral aqueduct. Histopathological confirmation was also essential as neuroradiological analysis alone cannot confirm the benign nature of the lesion.[8] However, the necessity of ETV as performed in this case is not yet established. Regression of the cyst following fenestration was thought to reopen the cerebral aqueduct and was therefore considered adequate. Furthermore, the third ventricular stoma could be occluded when CSF drainage through the cerebral aqueduct is established.

Recurrence of symptomatic PC after cyst fenestration is possible although rare.[11] Tirakotai et al reported one case of recurrence among nine patients following endoscopic transventricular cyst fenestration.[28] Furthermore, Tamura et al[17] reported a case of occluded aqueduct by the membrane structure of the PC 1 year after endoscopic cyst fenestration. Cerebrospinal fluid (CSF) flow may remain obstructed even after radical removal of a pineal lesion due to deformation of the rostral part of the aqueduct.[29] Therefore, ETV is considered effective in addition to cyst fenestration. There have been reports of satisfactory results following simultaneous cyst fenestration and ETV in cases of symptomatic PC.[3,26] Spontaneous regression of PC after ETV has also been reported.[30] Although it might be technically risky to perform both ETV and cyst fenestration simultaneously, these procedures could be completed safely by using a flexible endoscope instead of a rigid endoscope.

The patency of the third ventricular stoma was obvious on follow-up MRI, and there was no hydrocephalus, which confirmed long-term CSF communication through the third ventricular stoma despite aqueduct opening. Although the cyst was quite large with marked reduction in size and opening of the aqueduct following cyst fenestration, recurrence of the cyst is also possible. Therefore, we believe that ETV was necessary in this case. It is worth noting that the success of ETV as a highly specialized technique is largely dependent on the experience and training of the surgeon due to its high risk of complications, including bleeding,[17] and is therefore better performed by a surgeon with a great deal of experience in endoscopic intracranial procedures.

CONCLUSIONS

ETV in addition to cyst fenestration should be considered necessary and beneficial in cases of large symptomatic PC with associated obstructive hydrocephalus as it serves as a double check for non-recurrence of symptoms. In this era of increasing expertise in less invasive intracranial endoscopic procedures, this could be a better solution for patients with associated hydrocephalus.

ETHICAL APPROVAL

All procedures in studies involving human participants were performed in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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