Successful Treatment of Pure Aqueductal Pilomyxoid Astrocytoma and Arrested Hydrocephalus With Endoscopic Tumor Resection Followed by Chemotherapy: A Case Report and Technical Considerations

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BACKGROUND AND IMPORTANCE: Pure aqueductal tumors are extremely rare and may lead to death, regardless of histopathology. We report the first case of pure aqueductal pilomyxoid astrocytoma in an adult woman with arrested hydrocephalus. Endoscopic tumor resection for securing aqueductal patency followed by chemotherapy allowed control of both the tumor and the hydrocephalus.

CLINICAL PRESENTATION: A 20-year-old woman presented with mild cognitive dysfunction and marked ventricular dilatation. She had no preoperative symptoms of intracranial hypertension, and her head circumference was more than 2 SDs above the 98th percentile. The aqueduct of Sylvius was entirely occupied by a neoplastic lesion with 2 periventricular nodules at the wall of the third ventricle and anterior horn of the right lateral ventricle. She was treated successfully with endoscopic tumor resection followed by chemotherapy, and the patency of the aqueduct of Sylvius was secured throughout the duration of treatment.

CONCLUSION: Pure aqueductal tumors may be an appropriate indication for endoscopic tumor resection to secure the aqueductal patency of cerebrospinal fluid. This case demonstrates the surgical technique of a combined rigid-flexible endoscopic transforaminal approach, which was a useful surgical intervention for the management of this patient.

KEY WORDS: Aqueductal tumor, Arrested hydrocephalus, Cerebrospinal fluid dissemination, Endoscopic tumor resection, Long-standing overt ventriculomegaly in adults, Pilomyxoid astrocytoma

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Pure aqueductal tumors are extremely rare distinct entities that radiographically resemble tectal gliomas.[1-5] Aqueductal gliomas, once called "pencil gliomas,"[6,7] consist of a discrete aqueductal mass and principally infiltrate the periaqueductal zone, sparing the major white fiber tracts, cerebellar pathways, and the brainstem nuclei and representing a wider histopathological spectrum.[3,5,6,8,9]

Most patients with pure aqueductal tumors show symptoms of intracranial hypertension because of obstructive hydrocephalus and require tumor resection or cerebrospinal fluid (CSF) diversion.[3,5,9] The tumor may plug the lumen of the aqueduct of Sylvius (AS), frequently leading to sudden death.[6,7,9-11] In cases with disseminated lesions, ventricular peritoneal shunts may present a potential risk of intraperitoneal dissemination, and long-term extraventricular drainage has the potential for intracranial infection or ventricular collapse. In this study, we report a rare case of aqueductal pilomyxoid astrocytoma (PMA) treated successfully with endoscopic surgery followed by chemotherapy.

ABBREVIATIONS: AS, aqueduct of Sylvius; **BA,** basilar artery; **CISS,** coronal constructive interference in steady state; **ETV,** endoscopic third ventriculostomy; **MB,** mammillary body; **MI,** massa intermedia; **PMA,** pilomyxoid astrocytoma.

CLINICAL PRESENTATION

A 20-year-old female college student presented with mild cognitive impairment. Head computed tomography revealed marked

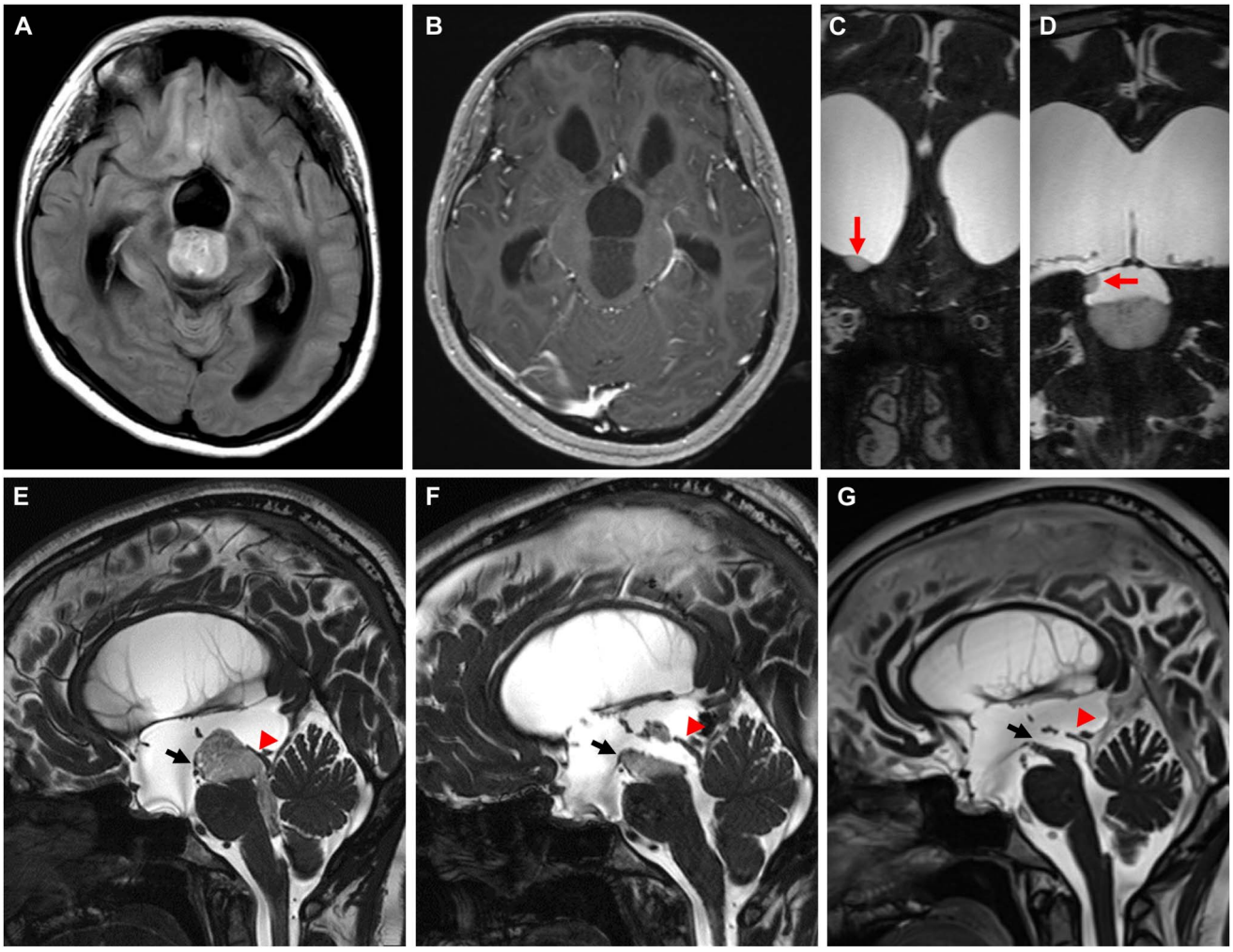

FIGURE 1. *Magnetic resonance images showing an occupying mass inside the lumen of the aqueduct.* **A**, *Fluid-attenuated inversion recovery images after gadolinium enhancement.* **B**, *T1-weighted images after gadolinium enhancement.* **C** *and* **D**, *Multiple periventricular nodules suggesting cerebrospinal fluid dissemination are seen on coronal CISS images (red arrows).* **E**, *Sagittal CISS images showing an aqueductal tumor with an incarcerated mass toward the fourth ventricle.* **F**, *Aqueductal patency was secured immediately after the surgery.* **G**, *Complete tumor response and flattening of the third ventricle floor were obtained after completion of chemotherapy (18 months after surgery). The tegmentum of the midbrain was dislocated forward (black arrows). The tectum plate did not show neoplastic changes (red arrowheads). CISS, constructive interference in steady state.*

triventricular dilatation. However, no clinical signs suggestive of intracranial hypertension, including headache and papilledema, were observed. Mild vertical gaze palsy and dyscoria in the left eye were observed. Her head circumference (58.0 cm) was more than 2 standard deviations above the 98th percentile (56.4 cm).[12] She had no familial history of neurofibromatosis type 1. Magnetic resonance imaging revealed a mass lesion (size, 50 × 25 × 23 mm) occupying the AS on a fluid-attenuated inversion recovery sequence, whereas T2-weighted images showed equal attenuation of the CSF. Gadolinium-enhanced T1-weighted images showed less-enhanced mass lesions. Coronal constructive interference in steady state magnetic resonance images revealed 2 other paraventricular lesions at the third ventricle and the right anterior horn, indicating CSF dissemination. Furthermore, sagittal constructive interference in steady state images showed incarcerated herniation of the caudal part of the tumor toward the fourth ventricle, and the floor of the third ventricle could not be detected (Figure 1).

Treatment Strategy and Surgical Technique

We planned an endoscopic tumor resection to obtain the pathological diagnosis and restore aqueductal patency (Figure 2, see Video).

The patient was positioned supine under intraoperative monitoring of motor-evoked potentials; burr holes were made 30 mm laterally astride the midline for a biportal endoscopic approach[13]; and

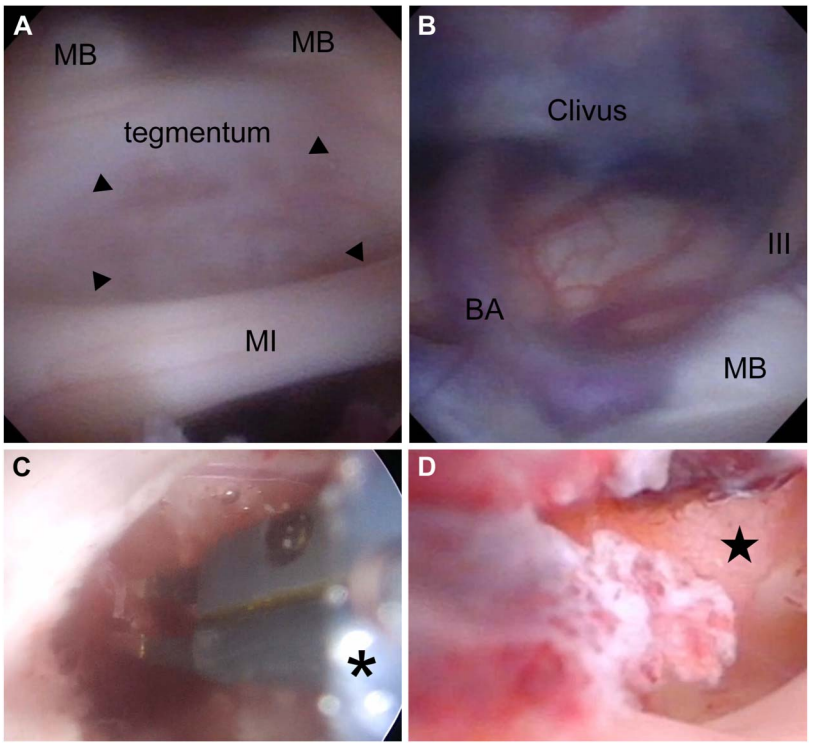

FIGURE 2. *Intraoperative endoscopic views showing the following findings:* **A**, *The entrance of the aqueduct of Sylvius is completely buried by the tumor (arrowhead).* **B**, *The floor of the third ventricle appears like a transparent veil over the prepontine space.* **C**, *Under the view with a rigid endoscope, the tumor is removed through the right-side Neuroport whose tip is located inside the third ventricle (asterisk).* **D**, *Penetration into the fourth ventricle is achieved using a flexible endoscope (star: choroid plexus). III, oculomotor nerve; BA, basilar artery; MB, mammillary body; MI, massa intermedia.*

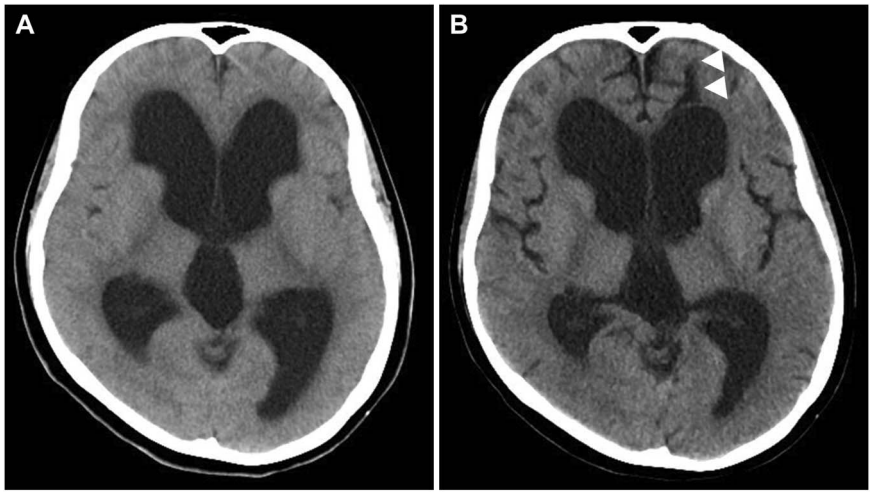

FIGURE 3. **A**, *Preoperative computed tomography scan showing marked ventricular dilatation.* **B**, *Although the ventricular size remained unchanged, the cerebral sulcus was clearly apparent postoperatively. Low-order transcortical corridor toward the aqueduct through the left anterior horn of the lateral ventricle is found (arrowhead).*

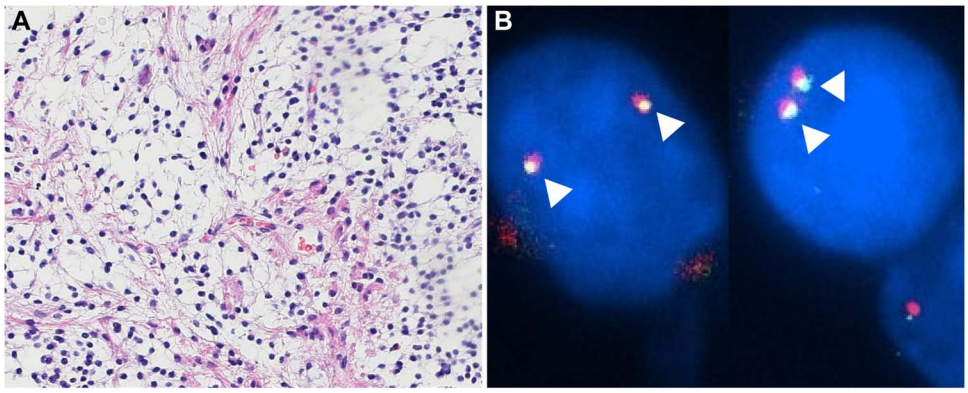

FIGURE 4. *Pathological findings:* **A**, *The tumor is composed of small monomorphic bipolar cells with delicate and thin cell processes and round oval nuclei in an abundant myxoid background. (H&E, × 100)* **B**, *FISH analysis reveals yellow signal of fusion gene (arrowheads) between the KIAA1549 (red) and the BRAF (green) oncogenes (BK fusion) of the tumor cells. FISH, fluorescence in situ hybridization.*

a lazy m-shaped skin incision was made along the hairline of the forehead. A Neuroport (Olympus Corp., inner diameter 9 mm) was used as an endoscopic sheath under the guidance of the navigation system. On the right side, the tip of the Neuroport was placed beyond the foramen of Monro to prevent damage when the equipment was inserted. The corridor on the left side was mainly used to set the rigid endoscope to visualize the intrathird ventricle surgical field. The CSF pressure was not elevated. First, using a flexible endoscope (VEF-V; Olympus Corp.), we confirmed complete occlusion of the AS and highly stretched third ventricle floor, preventing an effective third ventriculostomy (ETV). Subsequently, under the view of a 4-mm 0-degree rigid endoscope fixed with a fixation device (EndoArm; Olympus Corp.), the tumor continuing into the tegmentum inside the AS was debulked using forceps toward the fourth ventricle. Bleeding from the tumor could be managed suitably using coagulating devices. The incarcerated tumor toward the fourth ventricle was removed in a piecemeal fashion by using biopsy forceps under the flexible endoscope. An expanding balloon catheter (6 mm; Fuji Systems) was also useful for dissection between the tumor and the brainstem and for extracting the tumor mass. Finally, we successfully secured aqueductal patency (Figure 1F).

Postoperative Course

Ventricular dilatation remained unchanged, but the preoperative findings showing an unclear cerebral sulcus improved (Figure 3). New dyscoria in the right eye and strabismus superiorlaterally in the left eye occurred after surgery, indicating periaqueductal damage. Pathological examination revealed a PMA with the KIAA1549-BRAF fusion gene (Figure 4). The tumor cells showed negative results in H3K27M immunohistochemical staining, and the MIB-1 labeling index was 1%. Chemotherapy using carboplatin and vincristine was initiated,[14,15] and the tumor, including disseminated lesions, disappeared during the chemotherapeutic administration, consisting of 8 cycles of maintenance therapy (Figure 1G). The patient is

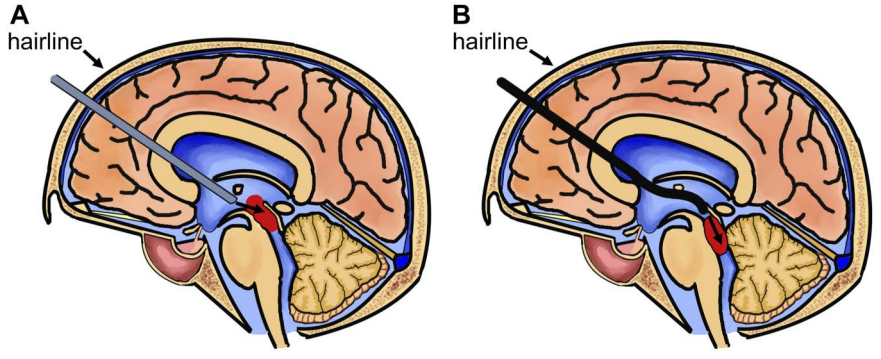

FIGURE 5. *Schematic representation of a combined rigid-flexible endoscopic approach for pure aqueductal tumors.* **A**, *Tumor at the entrance of the aqueduct can be removed by straight-line manipulations under the rigid endoscope.* **B**, *Tumor at the way out of the aqueduct must be removed by a flexible endoscope.*

currently healthy and without recurrence 8 months after the completion of all chemotherapeutic protocols.

Informed Consent

Informed consent from the patient and institutional ethics committee approval were obtained before submission.

DISCUSSION

PMAs show a more aggressive clinical course with a higher rate of recurrence and CSF dissemination than pilocytic astrocytomas, which are generally seen in young children and in the hypothalamic-chiasmatic region.[16,17] This case report presents quite rare clinical conditions, including adult onset,[18] a pure aqueductal location,[19] and compensated hydrocephalus with complete occlusion of the AS. To the best of our knowledge, this is the first report describing aqueductal PMA in an adult.[3,5,6,8,9]

We speculate that the reason why this patient presented no symptoms of intracranial hypertension, preexisted long-standing overt ventriculomegaly in adults,[20] may have hidden the occurrence of the tumor or led to its symptom-free development in early childhood. Although surgery may not always be necessary for patients with arrested hydrocephalus, long-standing ventriculomegaly may be associated with cognitive decline and even sudden death.[21,22] Patients with arrested hydrocephalus that become progressive may require surgical intervention without delay, and ETV is preferable over shunt placement.[20-22]

The field of endoscopic surgery has seen some remarkable advancements recently, especially for intraventricular tumors.[2,3,23-30] Considering the instantaneous effect of subsequent long-term chemotherapy, we could not expect favorable outcomes by ETV followed by chemotherapy for such a large tumor. Endoscopic tumor resection for ensuring aqueductal patency might have been the best way to avoid complications related to CSF diversion.[31] An adequate surgical trajectory to the AS through the foramen of Monro is critical for a safe surgery.[23,27,32] Care must be taken to avoid damaging the fornix, deep cerebral veins, and tegmentum during surgery.[33,34] After removal of tumor at the entrance of the AS, surgical procedures should be performed using a flexible endoscope that can incline the tip of the endoscope because the tumor descends downward toward the fourth ventricle (Figure 5).[35]

Limitations

Although this is a quite rare case report with marked ventriculomegaly, it may be useful for demonstrating endoscopic surgical techniques of intraventricular tumor resection.

CONCLUSION

We reported a rare case of pure aqueductal PMA in an adult. Endoscopic tumor resection using a combined rigid-flexible endoscope may be useful as a less-invasive surgical treatment option in such cases.

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Disclosures

The authors have no personal, financial, or institutional interests in any of the drugs, materials, or devices described in this article.

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VIDEO. Biportal endoscopic tumor resection of a pure aqueductal pilomyxoid astrocytoma.