Stereotactic endoscopic resection and surgical management of a subependymal giant cell astrocytoma

Case report

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Subependymal giant cell astrocytomas (SEGAs) are benign tumors, most commonly associated with tuberous sclerosis complex (TSC). Arising from the lateral ependymal surface, these tumors may obstruct one or both foramina of Monro, resulting in hydrocephalus and often requiring treatment. Although interhemispheric-transcallosal and transcortical-transventricular approaches have been the standard methods for resecting these tumors, advances in neuroendoscopic techniques have expanded SEGAs as a potential target for endoscopic resection.

The authors present a case of an endoscopically resected SEGA with stereotactic guidance in a 4-year-old girl with TSC. A gross-total resection of an enlarging SEGA was achieved. This represents one of the early case reports of endoscopically resected SEGAs. Although recent advances in medical treatment for SEGAs with mammalian target of rapamycin (mTOR) pathway inhibitors have shown promising initial results, the long-term safety and efficacy of mTOR inhibitors has yet to be determined. The propensity of these tumors to cause obstructive hydrocephalus requires that a surgical option remain. Neuroendoscopic approaches may allow a safe and effective technique.

Key words • stereotactic radiosurgery • endoscopic tumor removal • subependymal giant cell astrocytoma • tuberous sclerosis

Tuberous sclerosis complex is a rare, autosomal dominant, multisystem syndrome with multiple CNS manifestations. Gene mutations causing tuberous sclerosis include TSC1 and TSC2.1,5,9,13 These genes encode proteins involved in the mTOR signaling pathway. The mTOR pathway is the target of the chemotherapeutic agent rapamycin.3,4,8,14 One manifestation of TSC is a SEGA, with or without hydrocephalus. The tumor is almost exclusively intraventricular and is attached to the lateral ependymal surface. Hydrocephalus can develop insidiously as a SEGA grows, or acutely after tumoral hemorrhage,17 usually due to obstruction of the foramen of Monro.3,7,10 Other manifestations of tuberous sclerosis include subependymal nodules and tubers, which do not cause clinically significant mass effect.5,10,12,13,15,16

At our institution, SEGAs have been resected by an interhemispheric-transcallosal approach or a transcortical-transventricular approach. Endoscopic approach for resection of intraventricular tumors has been reported, including a small number of SEGAs.2,6,11 This case represents our initial experience with endoscopic resection of a SEGA.

Case Report

History. This 4-year-old, globally delayed girl presented with an enlarging SEGA in the right lateral ventricle. She had been diagnosed at 4 months of age, after the onset of generalized tonic-clonic seizures. Prior to the current report, she had undergone bilateral craniotomies

Abbreviations used in this paper: mTOR = mammalian target of rapamycin; SEGA = subependymal giant cell astrocytoma; TSC = tuberous sclerosis complex.
for subdural electrode placement and resection seizure foci. Her SEGA was monitored with serial imaging. The tumor was 7 mm in diameter at the age of 2 years. Two years later it was 25 mm and was causing moderate hydrocephalus (Fig. 1). A small SEGA was noted on the left side, and remained stable over the period of observation. Subependymal nodules were also noted. The patient’s family elected resection of the right-sided tumor due to the increasing size and hydrocephalus.

**Operation.** The patient was positioned supine with the head placed in rigid fixation. BrainLab stereotactic registration was performed for tumor localization, entry point and trajectory planning, and intraoperative localization throughout the operation (Fig. 2). A rigid, high-resolution, 2-mm working channel, rod-lens MINOP endoscope (Aesculap) was introduced via a right frontal approach. The vascular, ependymally based tumor was identified (Fig. 3 left). The outer capsule was cauterized using a combination of monopolar and bipolar diathermy (Fig. 3 right). Grasping forceps were used to obtain a tumor specimen (Fig. 4 left). Further resection was performed using a 5-Fr pediatric endotracheal suction cannula and pediatric feeding tube with a 10-ml syringe. The tubes were cut in a beveled fashion (Fig. 4 right). Transparency of the catheter allows for direct visualization during tumor removal and helps to prevent inadvertent capture of vessels, choroid plexus, or ependyma. Hemostasis was maintained with irrigation and bipolar/monopolar cauterization. An external ventricular catheter was placed and tunneled in the subgaleal space for CSF drainage postoperatively. It was removed on postoperative Day 3 (see Video 1 for footage of the operation).

**Video 1.** Video showing endoscopic resection of the SEGA. Demonstration of suction technique and tumor visualization. Click here to view with Media Player. Click here to view with Quicktime.

**Postoperative Course.** The patient’s postoperative course was notable for 2 days of apathy and abulia. Postoperative MRI studies confirmed gross-total resection of tumor and edema of the ipsilateral caudate nucleus, a possible correlation of her transient impairment (Fig. 5).
Endoscopically resected subependymal giant cell astrocytoma

subsequently returned to her neurological baseline. Her ventricular size has remained stable; no CSF diversion has been required.

Discussion

The natural history of SEGAs in the setting of TSC is generally of slow growth during the first 2 decades of life. The most common clinical presentation is of elevated intracranial pressure due to CSF obstruction at one or both of the foramina of Monro. For many patients, particularly those without hydrocephalus or significant tumor growth, this disease is most appropriately managed with serial clinical and radiographic follow-up.\(^1,3\)

Craniotomy for resection of an enlarging SEGA has been the favored approach at our institution. Interhemispheric-transcallosal and transcortical-transventricular approaches have been used. With the emergence of mTOR inhibitors, the paradigm of management for SEGAs may evolve to rely less on resection.\(^3,4,8,14\) The limitations of mTOR inhibitors include the following: lack of long-term follow-up, unclear length of required treatment, and limited side effect profile. Additional possibilities include the use of mTOR inhibitors preoperatively to facilitate endoscopic resection, or in cases of residual or recurrent tumor. However, for the tumors that show enlargement on serial MRI studies, endoscopic resection may be a feasible minimally invasive alternative.\(^2,3,11\) The advantages of an endoscopic approach include a smaller skin incision and cortical exposure, and clear visualization of the tumor and its attachment. The technical advancements include stereotaxy with planning of trajectory and entry point, rod lens with video capability, and the development of instruments for use in the working channel. Disadvantages include anatomical limitations and possible extensive lateral or deep attachments with proximity to the genu of the internal capsule that preclude gross-total resection.

A neuroendoscopic approach for resection of a SEGA has been reported.\(^2\) Our group has not previously used this technique. Our initial reluctance to attempt an endoscopic resection included the following reasons: the broad-based attachment of SEGAs to the caudate nucleus; significant calcifications and vascularity of these tumors; and the fact that a trajectory tangential to rather than perpendicular to the tumor favored a microsurgical approach. After gaining experience treating other tumors and disorders endoscopically, our indications have expanded. We agree with Cai and Di\(^6\) that the endoscopic technique is limited by calcification, tumor size, and vascularity. These factors may considerably lengthen the operating time. These lesions represent a formidable endoscopic target, and careful consideration should be given. The postoperative changes in the caudate demonstrate the potential for complications. If these changes had been observed in the internal capsule, a postoperative deficit could have been significantly worse and potentially permanent.

Conclusions

The treatment of TSC remains challenging. Rapamycin represents a potential paradigm shift for the treatment of SEGAs, although its long-term efficacy for tumor control has not been elucidated. By necessity, resection for enlarging tumors or tumors presenting with hydrocephalus will remain in the neurosurgeon’s armamentarium. The endoscopic approach provides a minimally invasive option for select tumors.

Disclosure

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

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References


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