



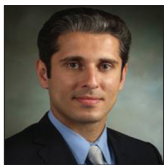
Case Report

Minimizing cortical disturbance to access ventricular subependymoma – A novel approach utilizing spinal minimally invasive tubular retractor system

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ABSTRACT

Background: Subependymomas are rare benign tumors found primarily in the lateral and fourth ventricles. Patients become symptomatic when the tumor obstructs cerebrospinal fluid pathways. We present a novel minimally invasive technique for lateral ventricular subependymoma resection.

Case Description: A 57-year-old male presented after a period of progressive ataxia, right upper extremity tremor, and syncopal events. Emergent non-contrast computed tomography of the brain demonstrated a lobulated mass in the left lateral ventricle causing moderate-to-severe obstructive hydrocephalus. Emergent ventriculostomy was placed as a temporizing measure. Subsequent magnetic resonance imaging (MRI) illustrated a large benign appearing mass causing obstruction of the left foramen of Monroe. A small craniotomy was performed utilizing previous ventriculostomy twist hole. The left lateral ventricle was accessed through sequential dilation of ventriculostomy tract using a minimally invasive spine surgery tubular system. Tumor was resected en bloc under microscopic assistance. The patient had an excellent outcome with return to baseline mental status and was discharged from the hospital postoperative day 1. Follow-up MRI demonstrated gross total resection of the mass and decreasing lateral ventricle hydrocephalus with minimal cortical disturbance.

Conclusion: A minimally invasive tubular system approach to ventricular tumors can be utilized to minimize cortical resection and brain retraction. Minimally invasive surgery also has the potential to decrease the length of stay and enhance postoperative recovery.

Keywords: Intraventricular tumors, Minimally invasive spine surgery, Minimally invasive spine tubular retractor, Subependymoma

BACKGROUND AND IMPORTANCE

Subependymomas are benign intraventricular slow-growing tumors found mostly in the lateral and fourth ventricles.^[1] These rare tumors were first described in 1945 by Scheinker^[13] and are mostly seen in middle-aged men.^[3,4] Patients become symptomatic when a tumor reaches 3–5 cm, blocking cerebrospinal fluid

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(CSF) pathways. Eliyas *et al.*^[6] presented a case series of ventricular tumor resections utilizing a specialized neuronavigation obturator for dilation through the sulcus. Here, we present a case of a left lateral ventricle pedunculated subependymoma resected through a minimally invasive spine tubular system which is readily available and does not require specialized instrumentation.

CLINICAL PRESENTATION/CASE REPORT

A 57-year-old male presented to the emergency department after 2 weeks of the right upper extremity tremor, progressive ataxia,

and a syncopal event. Neurologic examination was significant only for confusion and a resting tremor of his right upper extremity. Non-contrast brain computed tomography (CT) demonstrated a left lateral ventricle lobulated soft tissue density mass measuring 2.0 cm × 2.2 cm causing moderate-to-severe obstructive hydrocephalus at the foramen of Monroe [Figure 1a and 1b]. An emergent ventriculostomy was placed as a temporizing measure. Subsequent magnetic resonance imaging (MRI) illustrated a large benign appearing mass obstructing the left foramen of Monroe [Figure 2a-f]. The patient was taken to the operating room for mass resection.

The patient was placed under general anesthesia in a supine position with the head slightly flexed. A two-inch straight incision was made over the left frontal region incorporating the ventriculostomy puncture site [Figure 3]. A small craniotomy was completed, centered over the previous ventriculostomy twist hole. With neuronavigation assistance, bipolar electrocautery and suction were used to follow the ventriculostomy drain to the left lateral ventricle. Minimally invasive spine sequential dilators followed this trajectory to the ventricle to place a 14-mm diameter by 6-cm length minimally invasive spinal tubular retractor [Figure 4]. The operative microscope was then used to complete the operation [Video 1-4].

A small incision was made into the mass to obtain biopsy. Internal debulking was allowed for manipulation of the mass. A cottonoid covered the Foramen of Monroe to isolate the lateral ventricle in case of intraoperative bleeding. Bipolar electrocautery

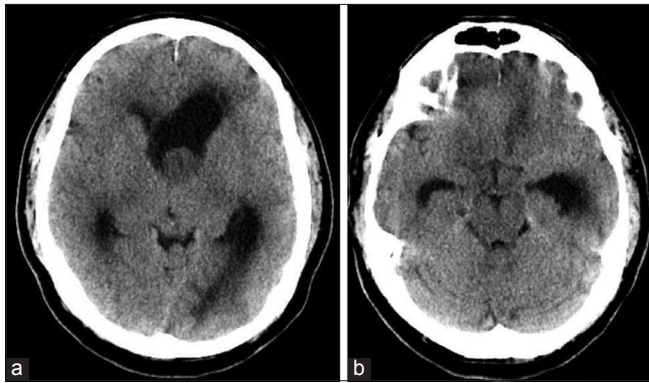


Figure 1: (a and b) Computed tomography brain w/o contrast noting lobulated soft tissue density mass left lateral ventricle measuring 2.0 cm × 2.2 cm causing severe obstructive hydrocephalus at the foramen of Monroe.

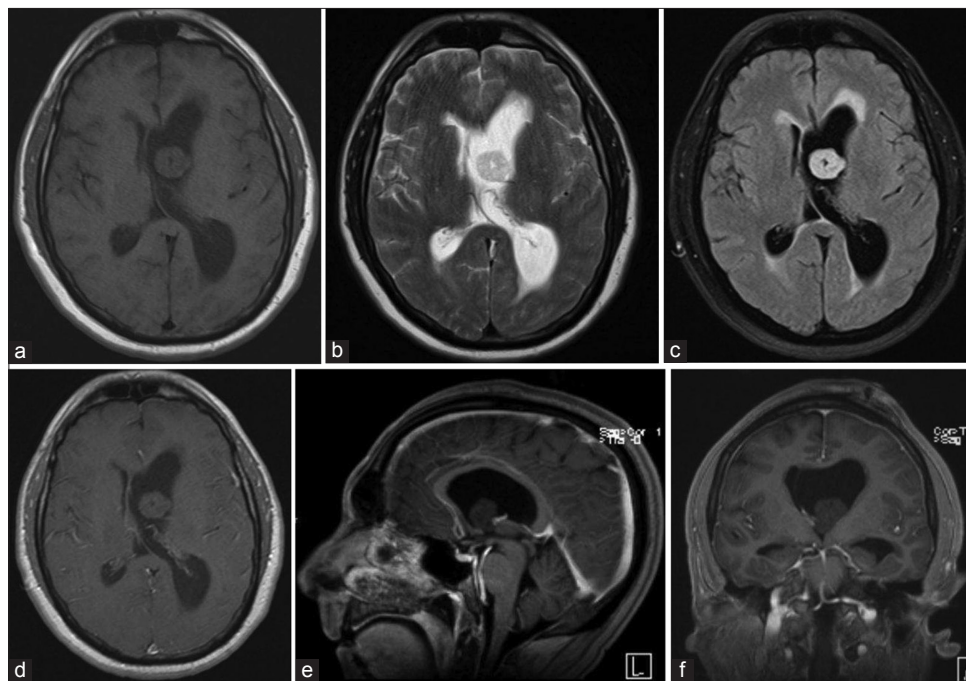


Figure 2: Magnetic resonance imaging brain with gadolinium demonstrating a large benign appearing mass causing obstruction of the left foramen of Monroe, (a) T1 hypointense mass, (b) T2 hypointense mass, (c) Flair hyperintense mass with transependymal edema, (d) Axial T1 w/gad hypointense mass without evidence of enhancement, (e) Sagittal T1 w/gad non-enhancing mass obstructing foramen of Monroe, (f) Coronal T1 w/gad non-enhancing mass obstructing lateral ventricle with left ventricle hypertrophy and rightward septal shift.

and micro scissors were used to transect the pedicle from the lateral ventricular wall. The mass was then removed en bloc. The ventricular anatomy was examined to confirm open CSF pathway before removal of the tubular retractor [Figure 5]. No ventricular catheter or drain was placed.

The patient had immediate return to baseline mental status and was discharged from the hospital postoperative day 1. Follow-up MRI demonstrated gross total resection of the mass and decreased lateral ventricle hydrocephalus with minimal cortical disturbance [Figure 6a-f]. Pathology demonstrated a subependymoma with microcystic degeneration, the WHO Grade 1 [Figure 7a-c]. Immunostains indicated that the lesional cells were positive for GFAP and S-100 [Figure 7d and 7e]. The proliferating cell fraction was <1% on KI-67 immunostain [Figure 7f]. 2 weeks later, the patient had improvement in gait and resolution of tremor.

DISCUSSION

Subependymomas are extremely rare benign and slow-growing tumors usually found in the ventricular system. Incidence has traditionally been reported as 0.5–0.7% of intracranial tumors; however, recent studies have found a much lower frequency of 0.07% of cases.^[3,11] Symptoms usually arise when the CSF is blocked causing intracranial hypertension. Previous studies have reported the most common symptom being headaches. Nausea, vomiting, dizziness, seizures, ataxia, memory loss, and vision changes have been reported as well.^[3,7,8,16] The patient reported ataxia and tremors but presented with syncope.

On CT, the mass will typically appear hypodense or isodense to the surrounding tissue and without calcification.^[3] T1-weighted MRI will demonstrate a hypointense mass and T2 will demonstrate a hyperintense mass without enhancement with gadolinium. Masses found in the supratentorial region tend to be ≥ 3 cm, located on the lateral wall or septum pellucidum, and are more likely to cause hydrocephalus by obstructing the foramen of Monroe.^[3,4] Posterior fossa tumors tend to be under 3 cm and extend into the foramina of Luschka and/or Magendie. They also tend to have calcifications and heterogeneous enhancement on CT and MRI.^[3] The differential diagnosis for subependymoma includes ependymoma, subependymal giant cell astrocytoma, choroid plexus papilloma, meningioma, metastatic lesions, and neurocytoma.

Pathologically, these tumors are most often pure subependymomas; however, they can be mixed subependymoma-ependymoma tumors as well as subependymoma-astrocytoma tumors.^[3,8] Histologically, the pathogenesis is incompletely understood but thought to originate from bipotential subependymal cells, causing a lobulated, hypocellular architecture with degenerative changes and microcalcifications. The tumor often has a sharp demarcation between normal brain parenchyma. Typical findings on immunostaining as were found in our patient are +GFAP and S-100 with minimal to no mitotic activity on KI-67.^[9]



Figure 3: 2” incision incorporating the ventriculostomy puncture site.

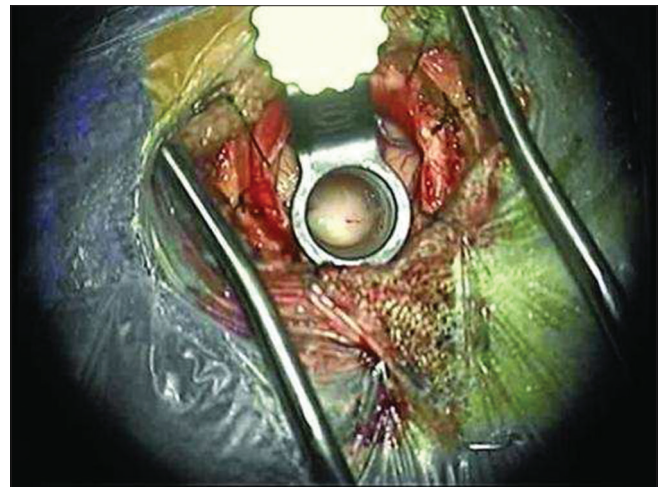


Figure 4: 14 mm × 6 cm minimally invasive spinal tubular retractor used for the transcortical exposure of the mass in the left lateral ventricle.

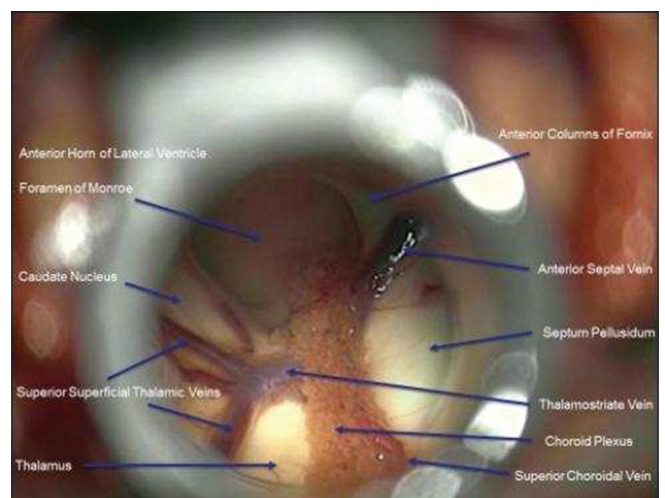


Figure 5: Ventricular anatomy, labeled.

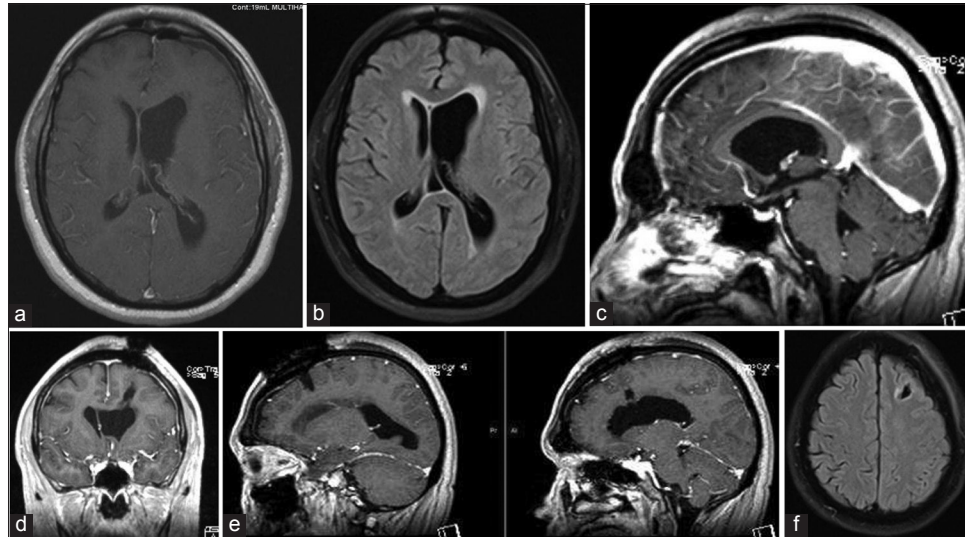


Figure 6: Postoperative magnetic resonance imaging brain w/gad demonstrating total resection of the mass and decreasing lateral ventricle hydrocephalus with minimal cortical disturbance, (a) Axial T1 w/gad with decreased left lateral ventricular hydrocephalus and gross total resection of ventricular mass, (b) Flair demonstrating reduced transependymal edema, (c) Sagittal T1 w/gad with gadolinium with patent foramen of Monroe after complete resection of intraventricular mass, (d) Coronal T1 w/gad illustrating the trajectory of tubular channel through cortex, (e) Sagittal T1 w/gad view of tubular channel through the cortex, (f) Axial flair view minimal cortical disruption.

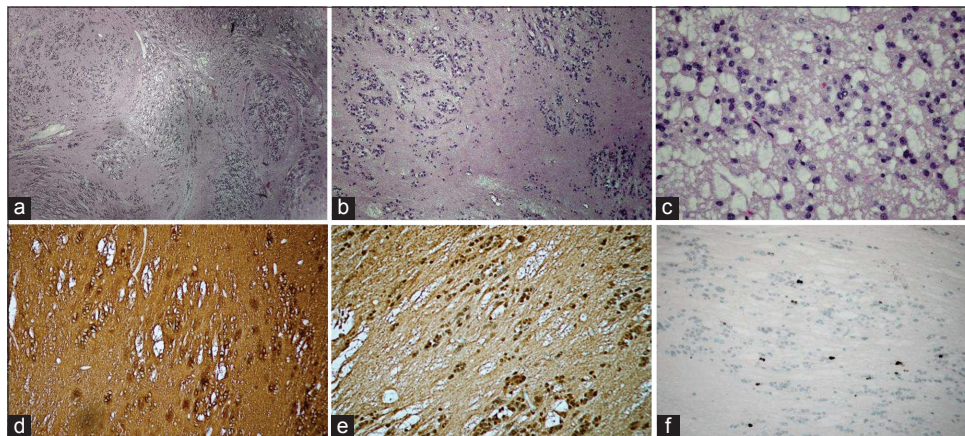


Figure 7: (a-c) Subependymoma with microcystic degeneration, the WHO Grade 1, (d and e) Positive lesional cell GFAP and S-100 immunostains, (f) Proliferating cell KI-67 immunostain fraction <1%.

Occasionally, nuclear polymorphism is seen which may portend a more aggressive tumor.^[10]

Subependymomas are a surgical disease with total resection being considered curative. There are varying opinions on the role of radiotherapy; however, there is no significant association with increasing survival to support its routine use. Authors conclude that radiotherapy may be considered if a subependymoma is a mixed tumor type, if a subtotal resection is performed, or if a patient remains symptomatic after a subtotal resection.^[3,8,13] Regardless, multiple studies evaluating long-term outcomes demonstrate almost no recurrence or progression of tumor.^[3,8,16] Previous case reports vary on their use of ventriculostomy placement which is indicated for significant pre- or postoperative hydrocephalus.

With posterior fossa tumors causing hydrocephalus, ventriculoperitoneal shunts are recommended preoperatively to prevent herniation.^[3] Before modern microneurosurgical techniques with neuronavigation, operative mortality was between 23.5 and 33%.^[12,15] Now, operative mortality rates are extraordinarily low, with only one study reporting a perioperative death in 43 cases.^[3] The only independent risk factor for shorter progression-free survival identified was a tumor with poorly defined borders.^[16]

When considering access to the tumor, traditionally a transcortical or transcallosal approach is undertaken. The former tends to have a higher rate of postoperative seizure and subdural fluid collections. Transcallosal approaches run the risk of hemiparesis, postoperative

mutism and classical disconnection syndrome. Surgical outcome depends predominantly on the patient's preoperative deficits rather than the approach taken.^[2] Although there is no universally recommended approach to access ventricular tumors, the approach that least disrupts the natural anatomy improves outcomes.^[5] Deep lesions require extensive cortical retraction to keep the channel open. A transsulcal approach utilizing a frameless stereotactic system with specialized dilators and tubular retractor had successful outcomes.^[6] However, a minimally invasive spinal tube system can be adapted as a working channel to access the lateral ventricles which allows for robust instrumentation for tumor resection with minimal cortical resection and blood loss.

CONCLUSION

Subependymomas are indolent benign tumors found primarily in the lateral and fourth ventricles and become symptomatic when CSF flow is obstructed. A minimally invasive spine tubular system approach to ventricular tumors can be utilized to minimize cortical resection and brain retraction. Minimally invasive surgery also has the potential to decrease the length of stay and enhance postoperative recovery.

Patient consent

The patient has consented to the submission of the case report for submission to the journal.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

The authors declare that they have no conflicts of interest.

REFERENCES

- Artico M, Bardella L, Ciappetta P, Raco A. Surgical treatment of subependymomas of the central nervous system. Report of 8 cases and review of the literature. *Acta Neurochir (Wien)* 1989;98:25-31.
- Asgari S, Engelhorn T, Brondics A, Sandalcioğlu IE, Stolke D. Transcortical or transcalsal approach to ventricle-associated lesions: A clinical study on the prognostic role of surgical approach. *Neurosurg Rev* 2003;26:192-7.
- Bi Z, Ren X, Zhang J, Jia W. Clinical, radiological, and pathological features in 43 cases of intracranial subependymoma. *J Neurosurg* 2015;122:49-60.
- Chiechi MV, Smirniotopoulos JG, Jones RV. Intracranial subependymomas: CT and MR imaging features in 24 cases. *AJR Am J Roentgenol* 1995;165:1245-50.
- D'angelo VA, Galarza M, Catapano D, Monte V, Bisceglia M, Carosi I. Lateral ventricle tumors: Surgical strategies according to tumor origin and development—a series of 72 cases. *Neurosurg* 2005;56:36-45.
- Eliyas JK, Glynn R, Kulwin CG, Rovin R, Young R, Alzate J, et al. Minimally invasive transsulcal resection of intraventricular and periventricular lesions through a tubular retractor system: Multicentric experience and results. *World Neurosurg* 2016;90:556-64.
- Hernández-Durán S, Yeh-Hsieh TY, Salazar-Araya C. Pedunculated intraventricular subependymoma: Review of the literature and illustration of classical presentation through a clinical case. *Surg Neurol Int* 2014;5:117.
- Jain A, Amin AG, Jain P, Burger P, Jallo GI, Lim M, et al. Subependymoma: Clinical features and surgical outcomes. *Neurol Res* 2012;34:677-84.
- Maiuri F, Gangemi M, Iaconetta G, Signorelli F, Del Basso De Caro M. Symptomatic subependymomas of the lateral ventricles. Report of eight cases. *Clin Neurol Neurosurg* 1997;99:17-22.
- Matsumura A, Ahyai A, Hori A. Symptomatic subependymoma with nuclear polymorphism. *Neurosurg Rev* 1987;10:291-3.
- Matsumura A, Ahyai A, Hori A, Schaake T. Intracerebral subependymomas. Clinical and neuropathological analyses with special reference to the possible existence of a less benign variant. *Acta Neurochir (Wien)* 1989;96:15-25.
- Ragel BT, Osborn AG, Whang K, Townsend JJ, Jensen RL, Couldwell WT, et al. Subependymomas: An analysis of clinical and imaging features. *Neurosurgery* 2006;58:881-90.
- Rath TJ, Sundgren PC, Brahma B, Lieberman AP, Chandler WF, Gebarski SS, et al. Massive symptomatic subependymoma of the lateral ventricles: Case report and review of the literature. *Neuroradiology* 2005;47:183-8.
- Scheinker M. Subependymoma: A newly recognized tumor of subependymal derivation. *J Neurosurg* 1945;2:232-240.
- Scheithauer BW. Symptomatic subependymoma. Report of 21 cases with review of the literature. *J Neurosurg* 1978;49:689-96.
- Varma A, Giraldi D, Mills S, Brodbelt AR, Jenkinson MD. Surgical management and long-term outcome of intracranial subependymoma. *Acta Neurochir (Wien)* 2018;160:1793-9.

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