CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 20 (2016) 49–52

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Two-stage surgical resection of an atypical teratoid rhabdoid tumor occupying the infratentorial and supratentorial compartment in children under two years: Report of two cases

Paul M. Foreman*, Casey J. Madura, James M. Johnston, Brandon G. Rocque

Department of Neurosurgery, University of Alabama at Birmingham, Birmingham, AL, United States

ARTICLE INFO

Article history:
Received 20 November 2015
Received in revised form 2 December 2015
Accepted 10 January 2016
Available online 16 January 2016

Keywords:
Atypical teratoid rhabdoid tumor
Resection
Infratentorial
Supratentorial

ABSTRACT

INTRODUCTION: Atypical teratoid rhabdoid tumors are highly malignant neoplasms that present in young children and can grow to a large size. Maximal safe surgical resection is a mainstay of treatment.
PRESENTATION OF CASES: Two cases of children under the age of two with large tumors involving the supratentorial and infratentorial compartments are presented. A two-staged operative approach combining a standard suboccipital approach to the fourth ventricle followed by an infratentorial, supracerebellar approach was utilized for resection.
DISCUSSION: Maximal safe surgical resection of large tumors in young children is challenging. A staged approach is presented that affords maximal tumor resection while minimizing perioperative morbidity.
CONCLUSION: A staged operative approach appears safe and efficacious when resecting large tumors from both the infratentorial and supratentorial compartments in children less than two years of age.

1. Introduction

Atypical teratoid rhabdoid tumors (ATRT) are rare, highly malignant embryonal tumors of the central nervous system (CNS) primarily occurring in young children with an overall age-adjusted incidence of 0.07 per 100,000 [1]. While they represent only 1.6% of all brain and CNS tumors diagnosed in the pediatric population, ATRTs represent 10.1% of tumors diagnosed in patients less than 1 year of age [1]. Although the overall survival remains poor, with most patients living less than 1 year, aggressive multi-modality therapy has improved survival [1–4]. Extent of surgical resection has been identified as a significant prognostic factor in the treatment of these aggressive tumors [2,4–6]. However, maximal safe resection of large tumors in very young children can be challenging due to technical factors, blood loss, and prolonged anesthesia.

Two cases of large ATRTs occupying both the infratentorial and supratentorial compartments in children less than two years of age are presented.

2. Presentation of cases

2.1. Case 1

An 18 month-old girl presents to an outside hospital with a history of vomiting and acute onset altered mental status. On exam, she was lethargic and unresponsive, but withdrew to painful stimuli. A computed tomography (CT) was obtained demonstrating a large posterior fossa mass with hemorrhage and hydrocephalus (Fig. 1). She was then intubated and transferred to Children's of Alabama (COA).

On arrival, she was a Glasgow Coma Scale (GCS) 4 with a right gaze preference, reactive pupils, absent left corneal reflex, and absent cough and gag reflex. An external ventricular drain (EVD) was placed at bedside and the patient taken emergently to the operating room (OR).

Patient was positioned prone on the operating table for a suboccipital craniotomy for hematoma evacuation and tumor debulking. A skin and fascial incision was performed, exposing the occiput, posterior ring of C1, and the top of the C2 lamina. A high-speed drill was then used to perform the occipital craniotomy and C1 laminectomy. Dura and arachnoid were then opened and tumor immediately encountered (Fig. 2A). Tumor resection proceeded rostrally through the fourth ventricle and into the third ventricle (Fig. 2B). Blood loss at that time was approximately 150 cc; the patient began to experience episodes of hypotension and the decision was made to stop the operation. Dura was closed using an expansile patch and bone was replaced. The wound was

* Corresponding author at: Faculty Office Tower 1005, 510 20th Street South, Birmingham, AL 35294, United States. Fax: +1 205 996 4208.
E-mail address: pforeman@uabmc.edu (P.M. Foreman).

http://dx.doi.org/10.1016/j.ijscr.2016.01.007

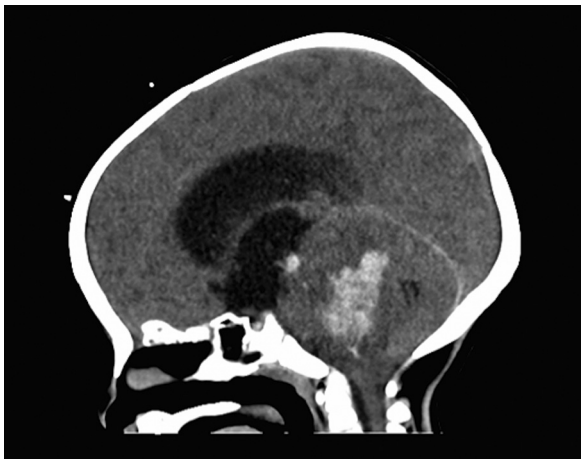

Fig. 1. Sagittal CT head demonstrating a large, hemorrhagic posterior fossa mass filling the fourth ventricle with associated hydrocephalus and brainstem compression.

closed and she was transferred to the intensive care unit (ICU) in stable condition. Postoperative magnetic resonance imaging (MRI) demonstrated subtotal resection of a very large infratentorial tumor with extension through the tentorial incisura into the supratentorial compartment. Histopathologic examination of tissue from this procedure revealed atypical teratoid rhabdoid tumor (ATRT).

The patient made an excellent recovery and was extubated on postoperative day 5. Given the large remaining tumor burden, plans were made for a repeat craniotomy for completion of the posterior fossa tumor resection combined with a supracerebellar, infratentorial approach to residual tumor mass. Incision was opened, but this time the bone removal was extended superiorly exposing the transverse sinus and confluence of sinuses. The dura was again opened and additional tumor removed from the fourth ventricle and cerebellopontine angle (CPA). Attention was then turned to the supracerebellar surface. After coagulating and dividing bridging veins between the tentorium and cerebellum, the space was opened revealing extensive amounts of tumor. Resection continued rostrally, dissecting tumor from the deep venous structures, skeletonizing the vein of Galen, and eventually exposing the atrium of the lateral ventricle. Tumor resection continued until pia and choroidal fissure were identified. Navigation was then used to confirm that all accessible tumor had been removed. Once again, an expansile duraplasty was performed with replacement of the craniotomy flap. Patient was extubated at the end of the procedure and returned to the ICU at her neurologic baseline. Postoperative MRI demonstrated near complete resection of residual tumor.

The patient's post-operative course was significant for hydrocephalus, treated with ventriculoperitoneal shunting. She made a full recovery, and underwent ablative chemotherapy with autologous stem cell rescue followed by radiation.

2.2. Case 2

A 22-month old male presents to an outside hospital with gait instability, nausea, and vomiting. On exam he was neurologically intact except for an ataxic gait. A CT was obtained demonstrating a large posterior fossa mass and hydrocephalus. He was transferred to COA. On initial evaluation, he was alert and interactive but developed lethargy, bradycardia and hypertension over the subsequent 24 h. An EVD was placed at bedside with clinical improvement. A MRI was obtained the following day revealing a large posterior fossa mass with extension through the tentorial incisura into the supratentorial compartment with involvement of the tectum (Fig. 3A). Plans were made for a two-staged surgical resection.

For the first stage, patient was positioned prone on the operating table and incision, dissection, and bone removal was similar to that previously described. Tumor was evident between the cerebellar tonsils upon opening of the dura and arachnoid. Tumor resection proceeded rostrally to the level of the aqueduct. At this point the tectum was noted to be engulfed in tumor. During microdissection of tumor off the dorsal mesencephalon, bleeding was encountered from a traversing vein. Additionally, the patient experienced a bout of hyperthermia coincident with an intraoperative blood transfusion and the decision was made to stop the operation. Hyperthermia resolved with acetaminophen and ice packs. Patient was closed with an expansile duraplasty with replacement of the craniotomy flap. Temperature normalized by the conclusion of the case and patient awoke at his neurologic baseline. Postoperative MRI demonstrated a large amount of residual tumor in the quadrigeminal cistern and extending superiorly through the tentorial incisura with near complete resection of the fourth ventricular aspect (Fig. 3B). Histopathologic examination again revealed ATRT.

As planned, the patient was brought back to the operating room on postoperative day 6 for a redo craniotomy for a supracerebellar, infratentorial approach. The supracerebellar space was entered as previously described (Fig. 4A and B). Tumor was apparent encasing the deep venous structures. Using microsurgical dissection, soft tumor was removed from the venous complex. After sacrifice of the precentral cerebellar vein to improve access to the midline, the resection was carried down to and stopping short of the tectum. At this point, near-complete resection had been accomplished and the decision was made to stop. The case was closed as previously described. Patient awoke moving all extremities and returned to the ICU. As the anesthetic effects cleared, he was noted to have right cranial nerve VI and VII palsies in addition to truncal ataxia and

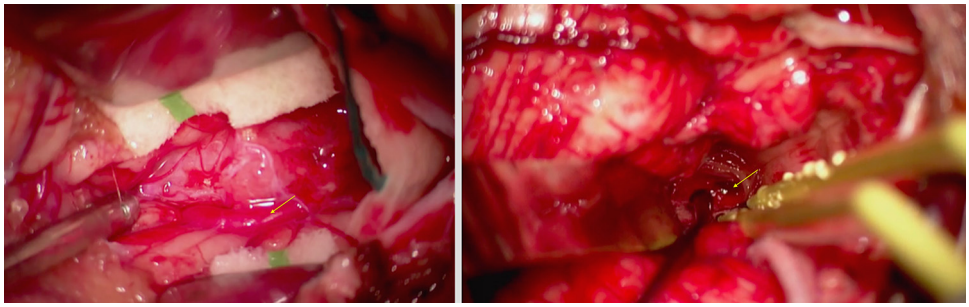

Fig. 2. Intraoperative photographs from *Case* 1—suboccipital, infracerebellar approach for tumor resection and hematoma evacuation. (A) Tumor extruding from the fourth ventricle upon opening of the dura and arachnoid (Arrow notes left PICA). (B) Following tumor resection and hematoma evacuation, a rostrally oriented view of the fourth ventricle and aqueduct (Arrow at cerebral aqueduct).

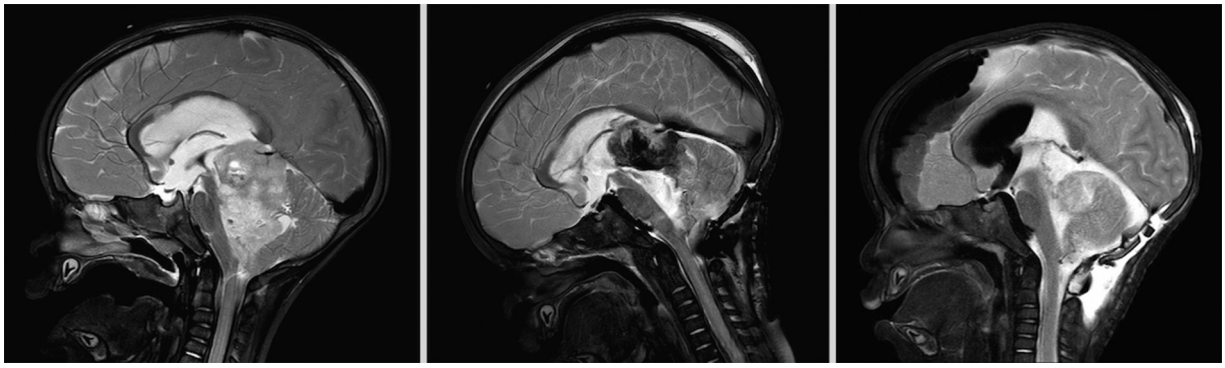

Fig. 3. Preoperative and postoperative sagittal T2 MRIs from *Case 2*. (A) Preoperative MRI demonstrating a large posterior fossa tumor with extension through the tentorial incisura into the supratentorial compartment. (B) Postoperative MRI following a suboccipital, infracerebellar approach for resection of tumor within the posterior fossa. Note the large residual, hemorrhagic tumor extending into the supratentorial compartment. (C) Postoperative MRI following a supracerebellar, infratentorial approach for residual tumor. Note the near complete resection with restoration of normal anatomic relationships.

cerebellar mutism. Postoperative MRI demonstrated near complete resection of tumor (Fig. 3C).

This patient also developed hydrocephalus, requiring surgical treatment with an endoscopic third ventriculostomy (ETV). The cranial nerve VI and VII palsies and truncal significantly improved, however, his symptoms of cerebellar mutism persisted. A similar oncologic strategy was implemented.

3. Discussion

Atypical teratoid rhabdoid tumors are malignant embryonal neoplasms most commonly diagnosed in young children. Large tumors occupying both the infratentorial and supratentorial compartments are thought to arise from the superior medullary velum (SMV), a thin lamina of white matter connecting the superior cerebellar peduncle and the inferior colliculi to form the roof of the rostral fourth ventricle [7]. With its center at the SMV, the tumor grows caudally into the fourth ventricle and rostrally into the quadrigeminal cistern, displacing the brainstem ventrally, the vermis posteriorly, and the fourth ventricle caudally [7]. Because of its growth within the ventricular and cisternal space, the tumor can achieve a large size prior to clinical detection [7].

Given the goal of maximal safe resection in a young child, the operation can be staged in an effort to minimize operative morbidity. A suboccipital craniotomy provides direct access to tumor within the fourth ventricle up to the level of the aqueduct, allowing for brainstem decompression. The telovelar variation of this approach provides additional lateral access for additional tumor resection. However, the rostral extent of resection is limited by the cerebellar vermis, superior cerebellar peduncles, and the SMV. This restriction is overcome by the supracerebellar infratentorial approach, popularized by Stein [8], that permits access to the tectal and pineal regions. The combination of these approaches allows for resection of large tumors spanning the infratentorial and supratentorial compartments without traversing intact neural structures. Cerebrospinal fluid diversion is another important technical aspect of posterior fossa tumor resection. Despite both patients being at high risk for postresection permanent CSF diversion [9], external ventricular drainage was selected over preoperative shunting or ETV. An EVD allows for continuous pressure monitoring, output recording, and can be easily removed if hydrocephalus resolves following tumor resection—advantages not provided by a shunt or ETV.

Atypical teratoid rhabdoid tumors harbor hallmark histologic and molecular features associated with the SMARCB1 tumor suppressor locus on chromosome 22q11.23 [6,10,11]. These hallmarks distinguish ATRTs as a distinct clinical entity separate from the more common primitive neuroectodermal tumors (PNET) [12]. Clinically, these tumors are characterized by aggressive behavior and propensity to occur in young children [1,13]. Despite a lack of consensus on the optimal treatment strategy, aggressive multimodal treatment is often pursued based on experience with other embryonal brain tumors [6].

Extent of surgical resection has been recognized as a positive prognostic predictor in the treatment of ATRTs [2,4–6]. Prolonged resections lead to increased blood loss, physiologic stress on the patient, and surgeon fatigue. The emergent first surgery of *Case 1* was complicated by blood loss and episodes of hypotension. The

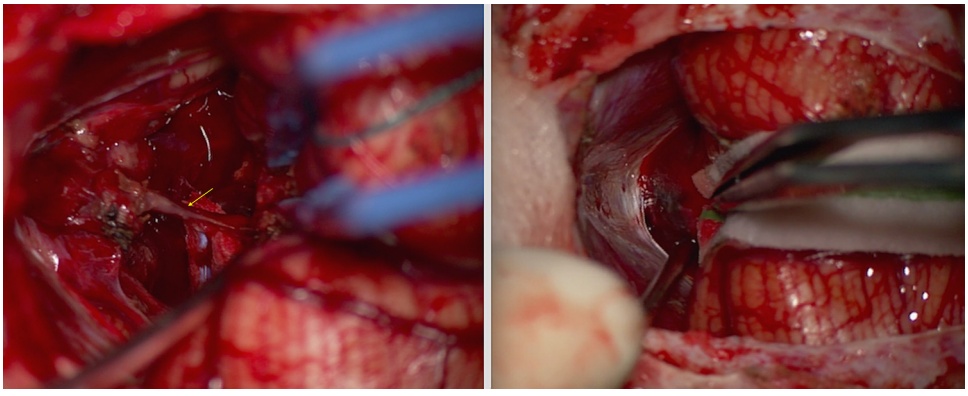

Fig. 4. Intraoperative photographs from *Case 2*—supracerebellar, infratentorial approach for residual supratentorial tumor. (A) Supracerebellar view of the precentral cerebellar vein (arrow). (B) Unimpeded view of the supracerebellar, infratentorial space following coagulation and division of the precentral cerebellar vein.

first surgery of *Case 2* was also complicated by blood loss, leading to a transfusion that produced hyperthermia. Although each situation was handled appropriately and the patient was not harmed, it emphasizes the delicate nature of complex tumor resection in young children.

4. Conclusion

Atypical teratoid rhabdoid tumors are highly malignant tumors that present in young children. A staged operative approach appears safe and efficacious when resecting large tumors from both the infratentorial and supratentorial compartments in children less than two years of age.

Conflict of interest

None.

Funding

We have acknowledged funding from a T32 in the manuscript.

Ethical approval

IRB approval is not required for case reports at the University of Alabama at Birmingham.

Consent

Not required per University of Alabama at Birmingham IRB.

Authors contribution

Paul M. Foreman—data collection, manuscript writing, critically revising article, reviewed final version of article, creation of figures.

Casey Madura—data collection, critically revising article, reviewed final version of article.

James Johnston—study concept, patient contribution, critically revising article, reviewed final version of article, study oversight.

Brandon Rocque—study concept, patient contribution, critically revising article, reviewed final version of article, creation of figures, study oversight.

Guarantor

Paul Michael Foreman.

Acknowledgements

We would like to thank Kirby I. Bland and his T32 NIH training grant for their support.

References

[1] Q.T. Ostrom, Y. Chen, P.M. de Blank, A. Ondracek, P. Farah, H. Gittleman, et al., The descriptive epidemiology of atypical teratoid/rhabdoid tumors in the United States, 2001–2010, Neuro Oncol. 16 (2014) 1392–1399.

[2] S.N. Chi, M.A. Zimmerman, X. Yao, K.J. Cohen, P. Burger, J.A. Biegel, et al., Intensive multimodality treatment for children with newly diagnosed CNS atypical teratoid rhabdoid tumor, J. Clin. Oncol. 27 (2009) 385–389.

[3] I. Slavc, M. Chocholous, U. Leiss, C. Haberler, A. Peyrl, A.A. Azizi, et al., Atypical teratoid rhabdoid tumor: improved long-term survival with an intensive multimodal therapy and delayed radiotherapy. The Medical University of Vienna experience 1992–2012, Cancer Med. 3 (2014) 91–100.

[4] L. Lafay-Cousin, C. Hawkins, A.S. Carret, D. Johnston, S. Zelcer, B. Wilson, et al., Central nervous system atypical teratoid rhabdoid tumours: the Canadian paediatric brain tumour consortium experience, Eur. J. Cancer 48 (2012) 353–359.

[5] A. Biswas, P.K. Julka, S. Bakhshi, A. Suri, G.K. Rath, Intracranial atypical teratoid rhabdoid tumor: current management and a single institute experience of 15 patients from north India, Acta Neurochir. (Wien.) 157 (2015) 589–596.

[6] J. Torchia, D. Picard, L. Lafay-Cousin, C.E. Hawkins, S.K. Kim, L. Letourneau, et al., Molecular subgroups of atypical teratoid rhabdoid tumours in children: an integrated genomic and clinicopathological analysis, Lancet Oncol. 16 (2015) 569–582.

[7] T. Tomita, P. Frassanito, Tumors of the superior medullary velum in infancy and childhood: report of 6 cases, J. Neurosurg. Pediatr. 11 (2013) 52–59.

[8] B.M. Stein, The infratentorial supracerebellar approach to pineal lesions, J. Neurosurg. 35 (1971) 197–202.

[9] P. Foreman, S. McClugage 3rd, R. Naftel, C.J. Griessenauer, B.J. Ditty, B.S. Agee, et al., Validation and modification of a predictive model of postresection hydrocephalus in pediatric patients with posterior fossa tumors, J. Neurosurg. Pediatr. 12 (2013) 220–226.

[10] I. Versteege, N. Sevenet, J. Lange, M.F. Rousseau-Merck, P. Ambros, R. Handgretinger, et al., Truncating mutations of hSNF5/INI1 in aggressive paediatric cancer, Nature 394 (1998) 203–206.

[11] J.A. Biegel, L. Tan, F. Zhang, L. Wainwright, P. Russo, L.B. Rorke, Alterations of the hSNF5/INI1 gene in central nervous system atypical teratoid/rhabdoid tumors and renal and extrarenal rhabdoid tumors, Clin. Cancer Res. 8 (2002) 3461–3467.

[12] L.B. Rorke, R.J. Packer, J.A. Biegel, Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood: definition of an entity, J. Neurosurg. 85 (1996) 56–65.

[13] K.F. Ginn, A. Gajjar, Atypical teratoid rhabdoid tumor: current therapy and future directions, Front. Oncol. 2 (2012) 114.