

Clinical diagnostic and radiographic features of primary spinal atypical teratoid rhabdoid tumors tumor in a pediatric patient: A case report and review of the literature

Journal of Central Nervous System Disease
Volume 15: 1–12
© The Author(s) 2023
Article reuse guidelines:
sagepub.com/journals-permissions
DOI: 10.1177/11795735231209199



Hashim Syed¹ , Nahom Teferi¹, Alec Hanson², Meron Challa², Kathryn Eschbacher³ and Patrick Hitchon¹

¹Department of Neurosurgery, University of Iowa Hospital and Clinics, Iowa City, IA, USA. ²University of Iowa, Carver College of Medicine, Iowa City, IA, USA. ³Department of Pathology, University of Iowa Hospital and Clinics, Iowa City, IA, USA.

ABSTRACT

Atypical teratoid rhabdoid tumors (ATRTs) are rare embryonal tumors comprising 1–2% of all pediatric CNS neoplasms. Spinal ATRTs are even more uncommon, accounting for 2% of all reported ATRT cases. Despite their rarity, ATRTs affect young children disproportionately and are characterized by a high malignant potential due to a heterogeneous cellular composition and inactivating mutations in the *SMARCB1* (90%) and *SMARCA4* (10%) genes. A 15-month-old female presented with a 2-week history of decreased lower extremity movement and new-onset need for assistance with ambulation. MRI lumbar spine revealed a contrast-enhancing intradural mass at the L3-L4 level with iso-intensity on T1 and T2 sequences. The patient subsequently underwent subtotal tumor resection (~80%) given concerns for maintaining neurological function. Final pathology was consistent with spinal ATRT, and she later underwent adjuvant chemoradiation therapy per ACNS0333 protocol. She has since remained in remission with age-appropriate developmental milestones over the past 2 years. ATRTs should be considered in the differential diagnosis of intradural spinal lesions, especially in the pediatric patient population. Clinical course, presentation, and diagnosis is often delayed due to the rarity of these tumors, but contrasted craniospinal MRI is key for diagnosis and histopathology with IHC staining showing loss of INI is confirmatory. While gross total resection is the goal, maximal safe tumor resection should be prioritized in order to preserve neurological function. Adjuvant chemoradiation following gross total/subtotal resection has been shown to significantly improve overall survival.

KEYWORDS: Atypical teratoid rhabdoid tumor, primary spinal tumors, intradural extramedullary

RECEIVED: July 27, 2023. **ACCEPTED:** October 5, 2023.

TYPE: Case Report

FUNDING: The author(s) received no financial support for the research, authorship, and/or publication of this article.

DECLARATION OF CONFLICTING INTERESTS: The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

SUPPLEMENTAL MATERIAL: Supplemental material for this article is available online.

CORRESPONDING AUTHOR: Patrick Hitchon, Department of Neurosurgery and Biomedical Engineering, University of Iowa Hospitals and Clinics, 200 Hawkins Dr, Iowa City, IA 52242, USA. Email: patrick-hitchon@uiowa.edu

Introduction

Atypical teratoid rhabdoid tumors (ATRT) are rare embryonal central nervous system (CNS) tumors that primarily occur in children under 3 years old, and account for 1–2% of all pediatric CNS cancers and 4.4% of CNS tumors in the age group 0–5 years.¹ Prior to 3 years of age, ATRT has a slight male preponderance and recently, it was shown that ATRT is the most common CNS malignancy in children younger than 6 months of age.^{2,3} Most of these tumors occur intracranially, with 55–75% of these lesions encountered in infratentorial locations and 25–40% located supratentorially, even rarely occurring in the ventricles.^{1,4} Spinal location for ATRTs is exceedingly rare and has an estimated prevalence of 2% among all CNS ATRTs.^{1,5}

Primary spinal ATRT is a rare subset of ATRT, with very few cases documented in the English-language literature. Often, pediatric patients with spinal ATRT present with rapidly

progressive symptoms of pain and myelopathy due to spinal cord compression and early diagnosis and management is key to improve overall outcome.^{2,6,7} Though the radiological appearance of these rare tumors is non-diagnostic, histological examination of the tissue with the use of immunohistochemical staining is key for making the diagnosis.^{6,7}

ATRTs are characterized by their heterogeneous histology and cellular composition, and consist of varying proportions of rhabdoid, epithelial, mesenchymal, spindled, and primitive neuroectodermal tumor (PNET) cells.^{8,9} Despite this morphologic heterogeneity, all ATRTs are associated with inactivating bi-allelic mutations in one of two genes, *SMARCB1* (90% of reported cases) or *SMARCA4* (10% of reported cases), resulting in a loss-of-function mutation in the chromatin-remodeling complex responsible for cell growth inhibition.¹⁰ Together, these mutations and cellular makeup confer a highly malignant potential, with 20–40% of all ATRTs reported to



Creative Commons Non Commercial CC BY-NC: This article is distributed under the terms of the Creative Commons Attribution-NonCommercial 4.0 License (<https://creativecommons.org/licenses/by-nc/4.0/>) which permits non-commercial use, reproduction and distribution of the work without further permission provided the original work is attributed as specified on the SAGE and Open Access pages (<https://us.sagepub.com/en-us/ham/open-access-at-sage>).

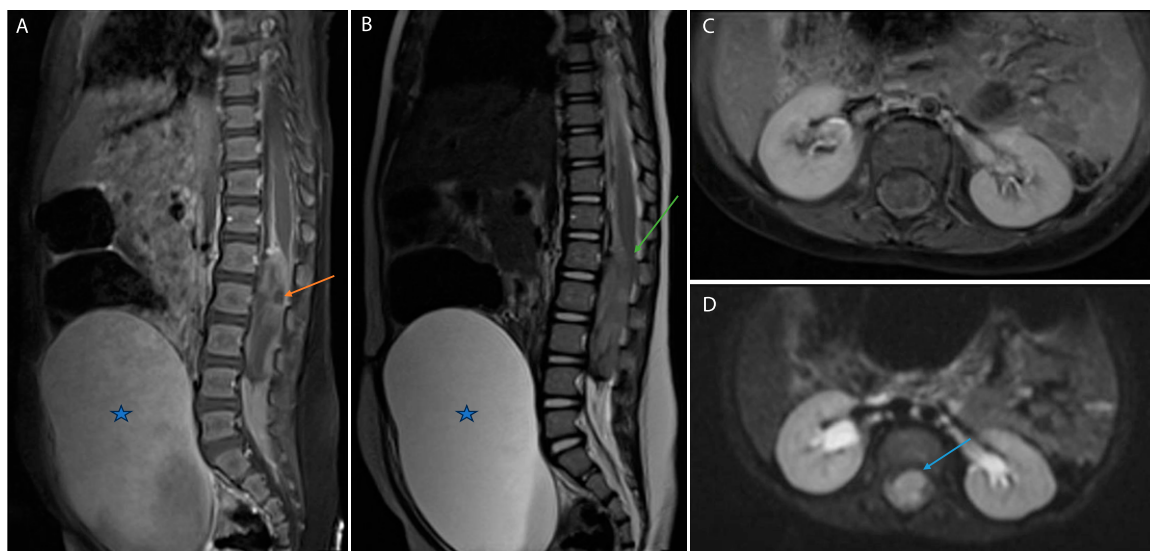


Figure 1. Pre-operative MRI of the spine: Sagittal T1 post-gadolinium contrast (A), sagittal T2 (B), axial T1 post-contrast (C), and axial DWI (D) demonstrating an intradural extramedullary mass (orange arrow) caudal and ventral to the conus medullaris (green arrow) at L1-L4 with T1 and T2 Iso-intensity, heterogeneous enhancement, and diffusion restriction (blue arrow). Note the largely distended bladder due to the mass effect and compression on the conus medullaris affecting bladder emptying (blue star).

present with leptomeningeal metastasis from their primary source on initial presentation.^{11,12}

Here we report the case of an intradural, extramedullary spinal ATRT found in a 15-month-old girl that responded well to subtotal resection followed by adjuvant chemoradiation therapy with ACNS0333 protocol.

Case presentation

A 15-month-old female presented to an outside hospital with a 2-week history of fussiness, decreased lower extremity movement, and new-onset need for assistance with walking. The patient's prenatal history was unremarkable, with a normal spontaneous vaginal delivery at 37 weeks term without any post-natal complications or history of congenital conditions in the family. Additionally, family history was negative for history of malignancy. On presentation, she was awake and alert, interactive, and demonstrated adequate tone and reflexes. However, she would not stand independently and cried upon lying prone. The patient was diagnosed with herpangina after oral ulcers were noted on physical exam, and work-up for osteomyelitis vs transient synovitis vs septic joint was initiated. Incidentally, an MRI of the pelvis obtained for further workup showed an intradural mass at the L3-L4 level with iso-intensity on T1 and T2 and mild enhancement on post-gadolinium contrast. Urinary retention was also noted on MRI, and a foley catheter was placed for suspected neurogenic bladder before the patient was transferred to the University of Iowa Hospitals and Clinics.

Subsequent total-spine MRI the following day revealed a large, intradural extramedullary mass caudal and ventral to the cauda equina spanning from L1-L4, measuring $13 \times 17 \times 53 \text{ mm}^3$ (Figure 1A-D). The mass showed heterogeneous enhancement and diffusion restriction. On gradient images, the mass exhibited

heterogeneous foci of blooming, suggestive of hemorrhagic foci. Given these findings, the initial differential consisted of small round blue cell tumors, such as embryonal tumors or Burkitt Lymphoma. Myxopapillary ependymoma was also considered but thought to be less likely on the differential given the patient's age. Following preoperative optimization, the patient underwent a posterior approach L1-L4 laminoplasty for intradural mass resection. Following bilateral L1-L4 laminectomy, intraoperative ultrasound was used to ensure that the intradural mass was localized rostrally and caudally within the margins of the bony exposure. The dura was then incised under microscopic visualization and the cauda equina and nerve roots overlying the mass immediately herniated dorsally through the dural and arachnoid defect. These nerve roots were micro-dissected with Penfield 3 and Rhoton 7 micro dissectors to expose a purple, friable mass with a central capsule (Figure 2A-C). Nerve roots were intertwined with the mass both rostrally and caudally. Following central debulking with bipolar cautery and suction, the capsule was mobilized rostrally and separated from the overlying nerve roots. Debulking continued in a caudal to rostral fashion with SSEP and EMG monitoring to avoid conus and nerve root injury. The most rostral aspect of the tumor was noted to be tightly adherent and ventral to the conus with no clear dissection plane identified. At this stage, the frozen specimen sent for pathology resulted and was concerning for a small round blue cell tumor with a differential of sarcoma, lymphoma, or germ cell tumor being considered likely. Due to the possibility of medical management for these differential diagnoses, resection was halted with ~80% of the tumor resected. Total tumor resection was avoided given concern for neurological injury given the remaining 20% of the tumor was abutting the conus medullaris with no clear dissection plane.

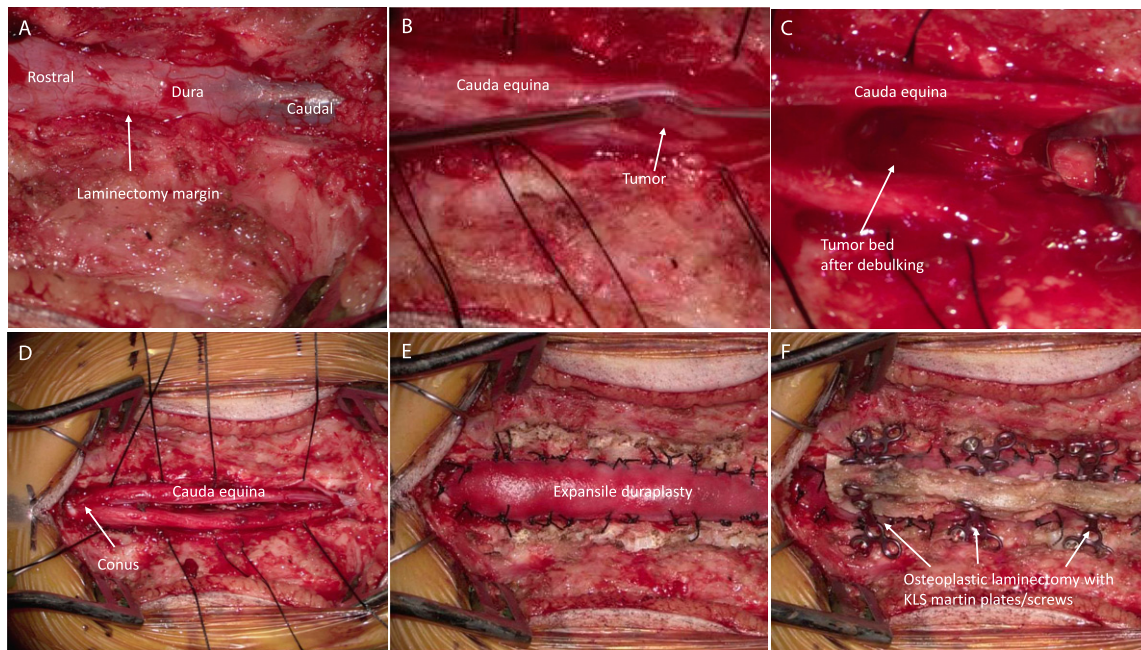


Figure 2. Intraoperative pictures. (A) Dorsal approach posterior L1-4 laminectomy with exposure of underlying dura (caudal is to the right). (B) Following dural opening, Penfield micro-dissectors are used to dissect the cauda equina and nerve roots ventral to the mass exposing a purple, friable mass. (C) Following exposure, the mass was centrally debulked with bipolar cautery and suction. The cauda equina nerve roots can be seen dorsally displaced. (D) After subtotal resection of spinal mass with residual mass ventral to the conus. (E) Expansile duraplasty was performed with Durerepair for dural closure. (F) The L1-L4 spinous processes and laminae were then reattached with KLS martin plates and screws to complete the laminoplasty.

Following satisfactory hemostasis, primary dural closure was attempted but unsuccessful due to continued dorsal herniation of the cauda equina and nerve roots (Figure 2D-F). Expansile duraplasty was then performed with Durepair (TEI Biosciences, Medtronic, Boston, MA) with 4-0 Nurolon sutures and Tisseel (Baxter International Inc., Westlake Village, CA) for dural sealant. Gel foam was placed on the dura, and the L1-L4 spinous processes and laminae were then reattached with KLS martin plates and screws (KLS Martin LLC, Jacksonville, FL). Vancomycin antibiotic powder was applied, and the patient's fascia and skin were closed using 3-0 nylon in a simple running fashion. The patient tolerated the procedure well with no postoperative complications.

Final histopathology of the tumor demonstrated poorly differentiated and discohesive cells with high nuclear to cytoplasmic ratios, vesicular chromatin, and prominent nucleoli (Figure 3A-D). Immunohistochemistry revealed immunoreactivity for epithelial membrane antigen (EMA), pankeratin, neurofilament, and smooth muscle-specific actin (SMSA) with loss of INI-1/SMARCB1, confirming the diagnosis of ATRT. The patient was initiated on 2 cycles of multiagent chemotherapy using cisplatin, cyclophosphamide, vincristine, etoposide, and methotrexate, followed by 3-course of high-dose chemotherapy with proton-based radiation treatment and autologous peripheral blood stem cell transplant per ACNS0333 guideline. Post-operatively, the patient remained neurologically stable with continued improvement of her lower extremity weakness throughout her hospital stay. The patient was

discharged on post-operative day 27 following completion of her first chemotherapy regimen. She was noted to be ambulatory with moderate assistance. 6 weeks after surgery, post-operative MRI spine confirmed residual tumor along the conus with no new enhancing spinal lesions (Figure 4A-D). The patient continued with 3 cycles of chemotherapeutic regimen per ACNS0333 guidelines over a 6-month period. At her most recent follow-up over 2 years after surgery, the patient's cancer remains in remission with age-appropriate developmental milestones (Figure 5).

Discussion

Atypical teratoid rhabdoid tumors (ATRTs) are rare embryonal tumors, representing only 1-2% of all pediatric CNS tumors.^{13,14} ATRTs originating primarily within the spine are estimated to account for less than 2% of all CNS ATRTs.¹⁵ Despite their rarity, ATRTs disproportionately affect younger children; 40-50% percent of CNS malignancies in children aged less than 1 are ATRTs, and it is imperative to consider ATRTs in the differential diagnosis of pediatric craniospinal neoplasms.¹⁴ Our patient initially presented to an outside hospital with lower extremity weakness and was referred to the University of Iowa Hospitals and Clinics for further evaluation after a lumbar lesion was discovered incidentally on MRI. Subsequent spinal imaging prompted tumor workup and evaluation, culminating in intradural mass resection in conjunction with ACNS0333 chemotherapy and adjuvant proton-beam radiation treatment.

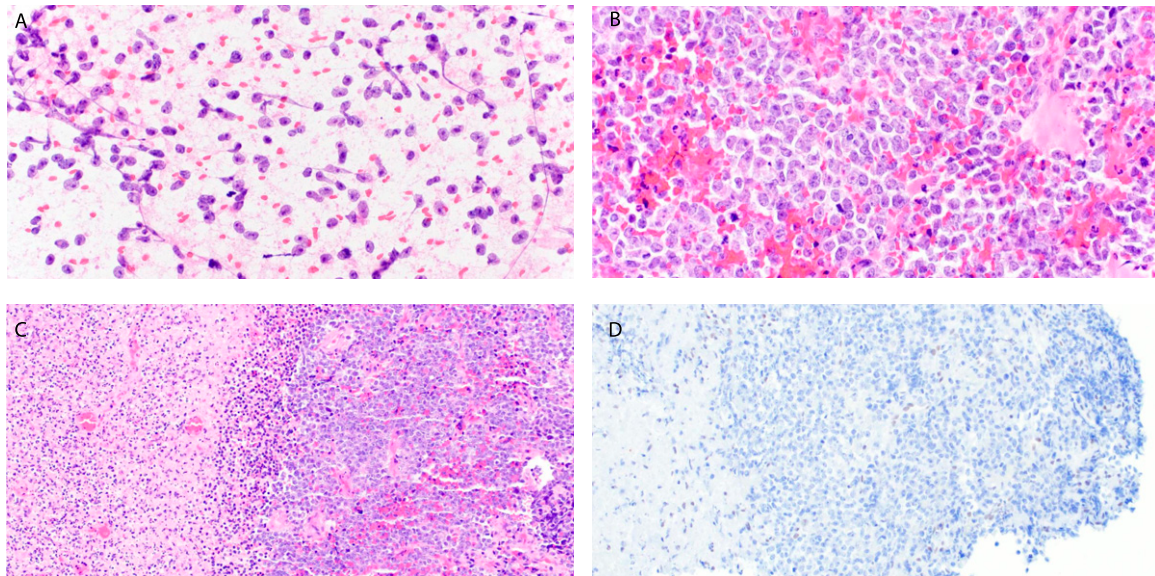


Figure 3. (A) H&E, 400x magnification: Intraoperative touch prep demonstrating dis-cohesive tumor cells with irregular nuclear contours, vesicular chromatin, and scant cytoplasm. (B) H&E, 400x magnification: Sheets of tumor cells with pleomorphic nuclei, prominent nucleoli, and scant cytoplasm. (C) H&E, 200x magnification: Small-round blue cell neoplasm with broad regions of necrosis. (D) INI1 immunohistochemical stain, 200x: Loss of nuclear expression of INI1 within tumor cells with preservation of expression within internal control (endothelial cells).

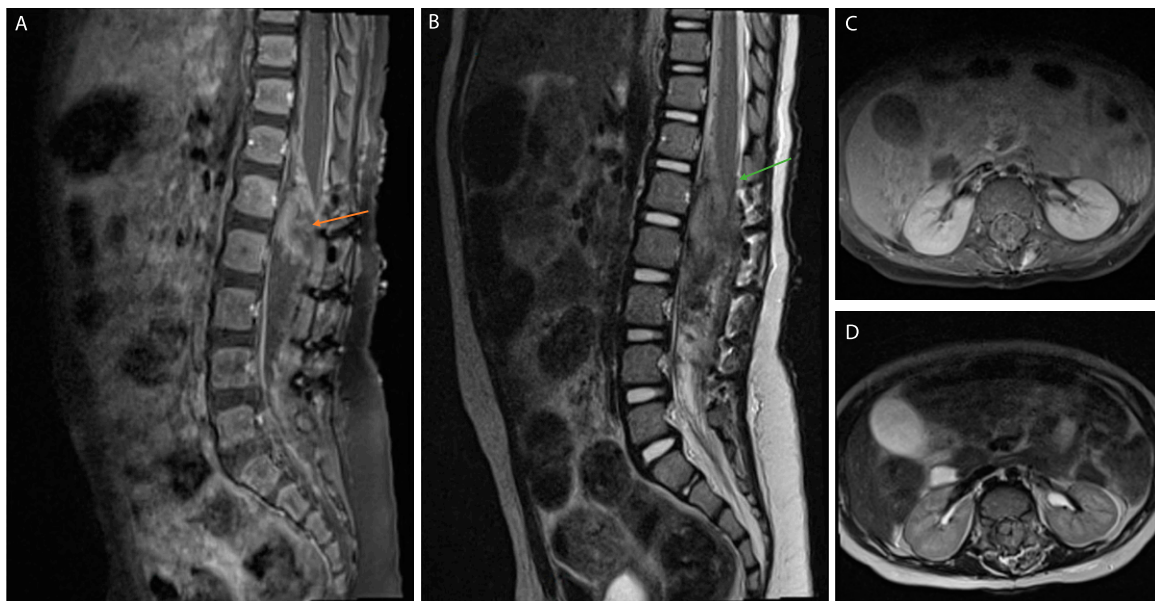


Figure 4. Post-operative MRI of the spine: Sagittal T1 post-gadolinium contrast (A), sagittal T2 (B), axial T1 post-contrast (C), and axial T2 (D) demonstrating persistent intradural extramedullary mass postoperatively (orange arrow) after incomplete resection with expected internal hemorrhagic changes and decreased mass effect on the conus medullaris (green arrow).

Histopathology/molecular features

Rhabdoid cell tumors were originally described in the context of renal tumors; however, in 1996 a seminal study established the presence of rhabdoid cell tumors within the central nervous system as being distinct from other lesions, such as medulloblastomas or primitive neuroectodermal tumors.^{16,17} In addition to rhabdoid cells, the researchers identified epithelial,

mesenchymal, and large and small cells consistent with a teratoid morphology, from which the tumor derives its name.⁸ Since then, spindle cells and sickle-shaped cells have also been identified on histologic sectioning.⁹

Despite this cellular diversity, all ATRTs are associated with mutations in the *SMARCB1* or *SMARCA4* genes, causing a loss-of-function protein deactivation.¹⁰ These genes encode proteins involved in the SWI/SNF chromatin remodeling

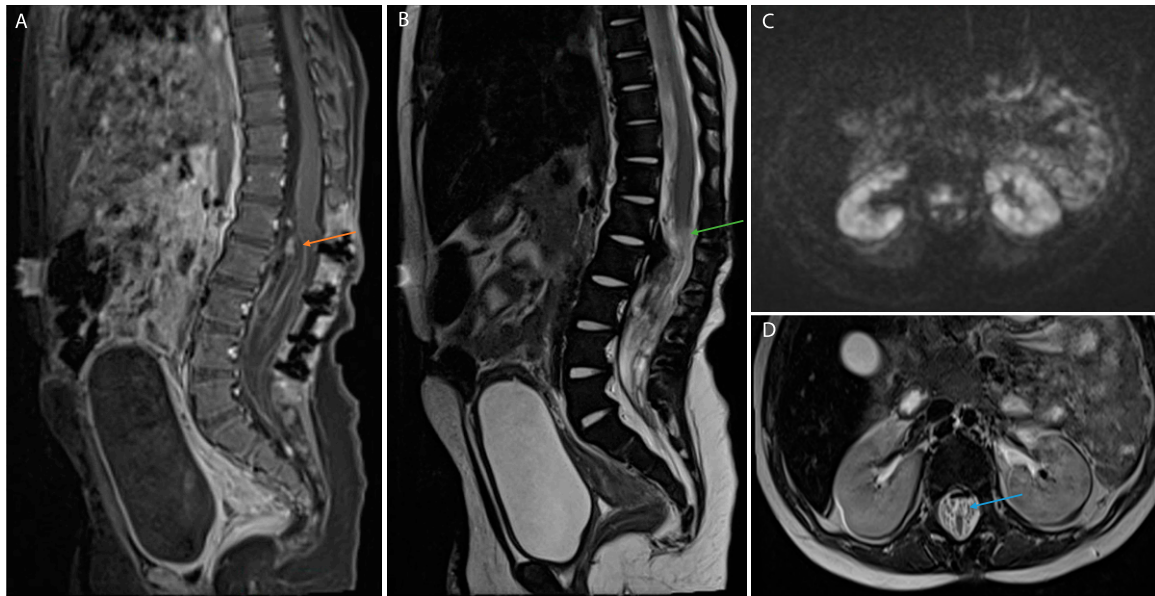


Figure 5. Repeat MRI spine 5 months post-adjuvant chemoradiation: Sagittal T1 post-gadolinium contrast (A), sagittal T2 (B), axial DWI (C), and axial T2 (D) demonstrating reduced tumor size (orange arrow) without new enhancing lesions and decompressed conus medullaris (green arrow) and cauda equina (blue arrow).

complex, named SMARCB1 and SMARCA4.¹⁰ The SWI/SNF complex is responsible for several cellular processes including DNA mismatch repair and chromatin structure regulation.¹⁸ Mutations in these key proteins result in decreased strand fidelity due to faulty mismatch repair, as well as uncontrolled cell growth resulting from dysregulated chromatin remodeling and increased DNA replication with looser chromatin structure.¹⁹ Finally, mutations in the SMARCB1 and SMARCA4 are associated with variable levels of DNA hypomethylation, suggesting epigenetic alterations of cell growth regulation.¹⁰

Although bi-allelic SMARCB1 loss is the critical genetic event in the vast majority of ATRTs, despite this genetic homogeneity, ATRTs are an epigenetically diverse group of tumors with distinct enhancer landscapes.²⁰⁻²² At least three molecular subtypes of ATRTs exist, each with unique targetable pathways and potential therapeutic vulnerabilities: ATRT-TYR, ATRT-SHH, and ATRT-MYC.²⁻²² ATRT-TYR and ATRT-MYC subtypes display a critical dependence on receptor tyrosine kinase pathways. The tyrosine kinase inhibitors (TKIs) dasatinib and nilotinib display selective toxicity to ATRT-MYC and TYR cell lines, and dasatinib significantly improved survival in an intracranial orthotopic xenograft model.²³ ATRT-SHH tumors appear to be more critically dependent on multiple targetable epigenetic regulators for survival like EZH2 and inhibition with an EZH2 inhibitor (UNC1999) has been shown to be selectively toxic to cell lines from this subgroup.²³ A phase-I trial using the EZH2 inhibitor (tazemetostast) for relapsed or refractory SMARCB1-deficient tumors is currently ongoing (NCT02601937). All these ongoing clinical trials and laboratory research focused on

translating these exciting new insights into efficacious therapeutics, will hopefully improve the outcome of this devastating disease. In terms of histopathology, intraoperative frozen pathology obtained from our patient was notable for a small round blue cell tumor with differentials of sarcoma, lymphoma and germ cell tumor considered more likely given the patient's age and tumor location. However, due to the histological similarity between other embryonal tumors and ATRTs, further molecular and histological work-up was pursued. Our patient's tumor molecular profile disclosed a *SMARCB1* pathogenic variant at a 50% allelic fraction and loss of INI-1 expression, consistent with one of the two ATRT mutations. Immunohistochemistry revealed EMA-1, pankeratin, neurofilament, and SMSA positive staining, further supporting the diagnosis of spinal ATRT by identifying rhabdoid, epithelial, and PNET cells.^{24,25} Most importantly, IHC was absent for SMARCB1 protein, allowing a definite diagnosis of ATRT to be made.²⁶

Clinical and radiological features

The clinical presentation of craniospinal ATRTs mainly depends on the location of the lesion and is often due to compression of adjacent neurovascular structures.^{5,26} Previously described symptoms included upper/lower extremity weakness or paralysis, paresthesia, myelopathy, gait instability, bowel/bladder control issues, and/or neck/back pain.^{5,27} Our patient's presentation is consistent with what has been reported in the literature (Table 1). At 15 months of age, she began to exhibit increased fussiness, decreased lower extremity movement, and preferred to stand in hip flexion over a span of 2 weeks. The

Table 1. Summary of published cases of pediatric spinal ATRT in the English literature.

AUTHORS	YEAR PUBLISHED	SEX	AGE	PRESENTING SYMPTOMS	TUMOR LOCATION	TREATMENT	CHEMO	RT	OVERALL SURVIVAL
Tantana et al ³⁰	1988	—	—	Symptomatic spinal cord compression	ED bony destruction	—	—	—	—
Horie et al ³¹	1992	F	4 m	Dorsal soft tissue mass	Lt thoracic ED space and paravertebral region	Laminectomy for subtotal resection	Y	N	4 m, DOD
Rosemberg et al ³²	1994	F	2 yr	—	IDEM	—	—	—	—
Rorke et al ²⁵	1996	—	Infant	—	Cervical IDEM	—	—	—	—
Tamiya et al ³³	2000	F	7 m	Progressive paraplegia & bilateral lower extremity hypesthesia	T7–L3 IDIM	T12–L3 laminectomies for subtotal resection	Y	Y	3 m, DOD
Bambakidis et al ³⁴	2002	M	22 m	Paraplegia	T11–L3 IDIM	subtotal resection	Y & ASCR	Y	10 m, DOD
Bambakidis et al ³⁴	2002	M	17 yr	—	Diffuse IDIM	Biopsy	N	Y	1 m, DOD
Mahmood et al ³⁵	2003	M	11 yr	Bilateral neck swelling, dysphagia, anorexia, right shoulder pain	Post oropharynx & cervical chain w/ invasion of clivus, left petrous apex, occipital bone and C1-2 vertebral bodies	Lymph node biopsy	—	—	—
Cheng et al ³⁶	2005	F	2 yr	Low-back pain & bilateral lower extremity weakness	T12–S1 ID spine (IDEM)	Multimodal intensive therapy (unspecified)	—	—	2 m, DOD
Chen et al ³⁷	2005	M	15 yr	Mid-back pain	IDEM w/invasion of rootlets	subtotal resection	Y & ASCR	Y	34 m, NED
Tanizaki et al ³⁸	2006	F	10 m	Progressive paraplegia	T10–conus IDIM	Decompressive laminectomy for biopsy	Y	Y	3 m, DOD
Bannykh et al ³⁹	2006	M	4 yr	Progressive bilateral lower extremity flaccid paralysis, pain in low back & right hip, areflexic bilateral lower extremities, sensory loss below L1, constipation	T9–L1 IDEM & ED extension	T9–L1 laminectomies for subtotal resection of ED component	Y, IT	Y	18 m, NED
Moeller et al ⁴⁰	2007	M	9 yr	Right lower extremity numbness, weakness, urinary incontinence	T11–L2 IDEM	Gross total resection	N	Y	3 m, NED
Yang et al ²⁷	2007	M	7 yr	Back pain & unstable gait	L2-4 IDEM	L2-4 laminectomies	Y	Y	7 m, DOD
Kodama et al ⁴¹	2007	M	9 m	Tetraparesis, severe loss of muscle tone, increased knee jerk	C4–T6 IDEM w/ED extension to right C6/7 neural foramen	subtotal resection	Y	Y	20 m, DOD

(Continued)

Table 1. Continued.

AUTHORS	YEAR PUBLISHED	SEX	AGE	PRESENTING SYMPTOMS	TUMOR LOCATION	TREATMENT	CHEMO	RT	OVERALL SURVIVAL
Tinsa et al ⁴²	2008	F	4 yr	Unstable gait, fever, weakness, anorexia	C1–T1 IDIM	C1 laminectomy, C2-7 laminoplasties & biopsy	N	N	0.5 m, DOD
Yano et al ⁴³	2008	F	1.75 yr	Acute, progressive tetraparesis	Cervical spine IDEM	Gross total resection	Y & ASCR	Y	27 m, NED
Agrawal et al ⁴⁴	2009	F	18 m	Increasingly painful swelling in low back with firm lump	Lumbar spine para-spinal mass w/ erosions of left L2 transverse process	Near-total resection	Y	N	NS
Athale et al ⁶	2009	M	4.25 yr	Mild fever, progressive generalized weakness, back pain & bilateral lower extremity pain, difficulty walking, right upper extremity weakness	C5–T2 IDEM w/ predominant extradural extension into neck	Cervicothoracic laminectomies for subtotal resection	Y	Y	11 m, DOD
Niwa et al ⁴⁵	2009	M	6 yr	Left neck pain & upper extremity pain	C4-6 IDEM w/ED extension into left neural foramen	Emergent C3-6 laminectomy & resection of IDEM component (subtotal resection)	—	—	—
Fridley et al ⁴⁶	2009	—	13 m	Symptomatic spinal cord compression	Cervical spine	Resected twice	Y	—	4 m, DOD
Mohapatra et al ⁴⁷	2010	M	4.5 yr	—	C1-2 IDEM	Resection (extent unspecified)	N	N	Unknown
Heuer et al ⁴⁸	2010	M	7 yr	Pain w/head turning, photophobia	Clival–C2 ED	Transoral gross total resection & occiput–C5 fusion	Y, IT	Y	42 m, DOD
Stabouli et al ⁴⁹	2010	M	2 m	Progressive tetraparesis, respiratory failure, bilateral lower extremity hyperreflexia & positive Babinski reflex	C1-5 IDEM w/ED extension into brachial plexus	Open biopsy	Y	N	6 m, DOD
Huddleston et al ⁵⁰	2010	M	6 m	—	Cervical spine	—	—	—	—
Dobbs et al ⁵¹	2011	F	17 yr	Left S1 pain & paresthesia	Left presacral space & S2 neural foramen	Biopsy of left S2 lamina → marginal subtotal resection of presacral mass	Y	N	NS
Makis et al ⁵²	2011	M	8 m	Bilateral lower extremity weakness, hyporeflexia, loss of motor milestones	L4–SC IDEM w/ED extension	Biopsy	Y	N	.75 m, NED
Imagama et al ⁵³	2012	F	2 yr	Recent fever, bilateral lower extremity paralysis and pain	T12–S1 IDEM	Emergency T10–S1 laminectomy for gross total resection	Y, IT	N	9 m, DOD

(Continued)

Table 1. Continued.

AUTHORS	YEAR PUBLISHED	SEX	AGE	PRESENTING SYMPTOMS	TUMOR LOCATION	TREATMENT	CHEMO	RT	OVERALL SURVIVAL
Kelley et al ⁵⁴	2012	M	4 yr	Constipation & several days of bilateral lower extremity paraplegia	T10–L1 IDEM mass	T9–L3 laminectomies for subtotal resection	Y, IT	Y	5 yr, NED
Yang et al ⁵⁵	2014	F	8 m	Progressive limb weakness	C2–T2 IDIM	subtotal resection	N	N	1 m, DOD
Xin et al ⁵⁶	2014	F	10 yr	Neck pain	C2-5 ED w/C3 osteolysis	subtotal resection	N	N	8 m, DOD
Dhir et al ⁵⁷	2015	M	2.5 yr	Inability to ambulate, encopresis, urinary dribbling since 2 weeks of age, decreased rectal tone, bilateral foot drop	L3 & below IDEM	L3–S2 laminoplasty	Y	N	8 m, NED
Tang et al ⁵⁸	2015	F	13 yr	Neck pain & left neck mass	C4 vertebra	Percutaneous needle biopsy	Y	Y	40 m, NED
Mankotia et al ⁵⁹	2016	M	5 yr	Rapidly ascending quadriparesis, respiratory distress, sensory loss below T4, myelopathic features	T5-10 IDEM	T4-10 laminoplasty for subtotal resection	Y	N	1 m, NED
Chao et al ⁶⁰	2017	M	16 m	Urinary retention, change in bowel habits, unable to walk w/decreased sitting balance, bilateral lower extremity loss of muscle tone	T11–L3 IDEM	T10–L3 laminectomies w/ neuro-endoscopic resection	Y, IT	Y	7 m, NED
Babgi et al ⁶¹	2018	M	6 yr	—	Thoracolumbar junction (IM,EM)	Resection ×2	Y	Y	16 m, DOD
Singla et al ⁶²	2016	M	3 m	Right lower extremity flaccid paralyses	T12–S1 ED	Gross total resection	N	N	—
Nishimoto et al ⁶³	2018	M	3 yr	Tetraparesis & dysphagia	C1-2 ED & osteolysis lower clivus	Emergent C1 laminectomy & delayed lat approach for subtotal resection & occiput-C2 fusion	N	Y	29 m, DOD
Song et al ⁶⁴	2018	M	16 m	Bilateral lower extremity weakness, areflexic	L1-3 IDEM w/ED extension	L1-4 laminoplasty for resection	N	Y	—
Garling et al ⁶⁵	2018	F	15 yr	Bilateral lower extremity weakness, decreased sensation, weight loss, back pain, & progressive bowel and bladder incontinence	L3–S2 IDEM w/ED extension into sacral plexus	L3-5 laminectomies for subtotal resection	—	—	—

(Continued)

Table 1. Continued.

AUTHORS	YEAR PUBLISHED	SEX	AGE	PRESENTING SYMPTOMS	TUMOR LOCATION	TREATMENT	CHEMO	RT	OVERALL SURVIVAL
Hong et al ⁶⁶	2018	M	3 yr	Right upper extremity weakness → quadriparesis, bowel and bladder incontinence	C3-5 IDEM w/ED extension	Gross total resection	Y	Y	—
Buccoliero et al ⁷	2019	F	4 yr	Difficulty walking, pain in pelvis & legs	T11–L2 IDIM	subtotal resection	Y	N	4 m, DOD
Buccoliero et al ⁷	2019	M	18 m	Pain & weakness left lower extremity weakness	L1-5 IDEM w/ED extension	subtotal resection	Y	N	3 m, DOD
Tsitsopoulos et al ⁶⁷	2020	M	4 m	Hypotonia, poor head control, gradual reduction of consciousness	Intraspinal extradural	Chemotherapy and radiation therapy	Y	Y	4 m, DOD
Kramer et al ⁶⁸	2020	F	5 yr	Progressive bilateral lower extremity pain	conus medullaris	surgical decompression	—	—	—
Shiflett et al ⁶⁹	2020	F	17 m	Stopped walking, fussiness	T11 down to cauda equina	Midthoracic to L5 laminoplasties gross total resection w/resection of lumbar nerve roots, meninges, & conus medullaris	N	N	1 m, DOD
Meena et al ⁷⁰	2020	M	3 yr	Facial grimacing, loss of all function below C4 with flaccid quadriplegia, preceded by difficulty walking hemato-myelia and spinal subarachnoid hemorrhage	C3-C7 IDEM	subtotal resection	N	N	1 m, DOD
Biswas et al ⁷¹	2021	M	6 yr	—	D10-D12 with a 9 mm enhancing lesion at L5-S1	Subtotal resection, chemoradiation	Y	—	30 m NED
Biswas et al ¹⁵	2023	F	18 m	—	C3-D1	Subtotal resection and multiagent adjuvant chemotherapy and focal RT	Y	Y	30 m NED

Legend: — = not reported; m = months, yr = years; ED = extradural, ID = intradural, EM = extramedullary, IM = intramedullary, IT = intracranial; Y = yes, N = no; ASCR = autologous stem cell rescue, SRS = stereotactic radiosurgery, DOD = died of disease, NED = no evidence of recurrence.

patient's tumor was found incidentally while exploring more common etiologies of pediatric hip and back pain such as osteomyelitis, transient synovitis, or septic joint. The rapid progression of symptoms was concerning for a more aggressive neoplastic process and has previously been reported in patients with spinal ATRT. Subsequently, further workup was performed with MRI of the neural axis, given the likelihood of early metastasis on presentation of pediatric craniospinal embryonal tumors.²⁸ ATRTs, specifically, have a high rate of leptomeningeal seeding and metastasis, with 46% of cases reported to have metastatic disease on presentation.²⁹ It's paramount to

obtain thorough neural axis imaging to evaluate for the presence of distant seeding from the primary tumor site on initial workup. Fortunately, in our patient, neural axis MRI showed no signs of distant metastasis or seeding.

Treatment and long-term outcomes

The prognosis of ATRTs is highly variable depending on tumor location, clinical presentation, and presence of metastases. A 2022 systematic review by Egiz et al of 39 retrospective studies found the median overall survival rate of pediatric ATRT

diagnoses to be 29 months.⁷² ATRTs of spinal origin, as was found in our patient, demonstrate a significantly lower median survival rate compared to those that are cranial in origin. For primary tumors, surgical resection is a mainstay of treatment and prognosis may correlate with the extent of the tumor that can be safely resected.⁷² Egiz et al found that complete tumor resection increased the median survival rate to 4.2 years as compared to .5-.9 years in patients with partial or subtotal resection. Adjuvant therapy is also recommended, with chemotherapy, combination chemotherapy and photon or proton-based radiation treatments, all having been utilized for multimodal management.⁷³ A combination of chemotherapy and radiation is most common with a median survival rate of 8.842 years.⁷² Prognosis worsened to a median survival rate of 4.942 years in patients who receive proton-based radiation therapy, and only .833 years in patients who receive chemotherapy as a lone adjuvant to surgical resection.⁷² This data suggests that maximal tumor resection alongside chemotherapy and radiation may provide optimal patient outcomes. However, due to the lack of a standard treatment protocol, various multimodal regimens are still being explored through clinical trials as seen in our patient's case.

Our patient underwent surgical resection followed by high-dose chemotherapy with peripheral blood stem cell rescue as outlined by the ACNS0333 clinical trial protocol, followed by 6 weeks of proton-based radiation therapy.⁷⁴ Given her young age, proton-based radiation treatment was chosen to reduce the risk of late radiation treatment related effects. Proton-based radiation therapy has gained wide acceptance in the pediatric oncology community as a modality to minimize the toxicities associated with photon-based radiation treatments.^{75,76} Given the physical properties of the proton beam, it allows for conformation of the radiation dose more strictly to the volume of interest, avoiding critical organs with greater normal tissue sparing than occurs with photons and is a theoretical advantage because of the reduction of integral dose to normal tissues (ex. conus medullaris in our patient).^{75,76} This ability of protons to spare normal tissues beyond a specified depth results in lower or no dose to surrounding organs with subsequent reduced toxicities.^{75,76}

Currently, our patient remains in remission 27 months from her initial diagnosis. We demonstrate that sub-total surgical resection followed by aggressive adjuvant therapy can allow patients to achieve remission and continue development appropriately, with minimal neurologic impairment.

Ethical statement

Ethics approval and consent to participate

The authors of this study confirm that written informed consent was obtained from the patient's parental guardians for participation in this study. The University of Iowa Hospital

institutional review board approved all aspects of this study under IRB # 201902751.

Consent for publication

The authors of this study confirm that a written informed consent for publication of the patient's medical images and data was obtained from the patient's parents.

Author contributions

Conception and design: Nahom Teferi. Acquisition of data: Hashim Syed, Alec Hanson. Analysis and interpretation of data: Hashim Syed, Alec Hanson. Drafting of the article: Hashim Syed, Alec Hanson. Critically revising the article: Nahom Teferi, Meron Challa, Kathryn Eschbacher, Patrick Hitchon. Approved the final version of the manuscript on behalf of all authors: Nahom Teferi.

ORCID iD

Hashim Syed  <https://orcid.org/0009-0005-5284-9895>

REFERENCES

- Ho B, Johann PD, Grabovska Y, et al. Molecular subgrouping of atypical teratoid/rhabdoid tumors—a re-investigation and current consensus. *Neuro Oncol.* 2020;22(5):613-624. doi:10.1093/neuonc/noz235
- Biswas A, Kashyap L, Kakkar A, Sarkar C, Julka PK. Atypical teratoid/rhabdoid tumors: Challenges and search for solutions. *Cancer Manag Res.* 2016;8:115-125. doi:10.2147/CMAR.S83472
- Frühwald MC, Biegel JA, Bourdeau F, Roberts CW, Chi SN. Atypical teratoid/rhabdoid tumors—current concepts, advances in biology, and potential future therapies. *Neuro Oncol.* 2016;18(6):764-778. doi:10.1093/neuonc/nov264
- Karim A, Shaikhyzada K, Suleimenova A, Ibraimov B, Nurgaliev D, Poddighe D. Case report: Atypical teratoid/rhabdoid tumor of the lateral ventricle in a male adolescent (case-based review and diagnostic challenges in developing countries). *Front Oncol.* 2022;12:985862. doi:10.3389/fonc.2022.985862
- Li D, Heiferman DM, Syed HR, et al. Pediatric primary spinal atypical teratoid rhabdoid tumor: A case series and review of the literature. *J Neurosurg Pediatr* 2019; 24(3):1-17. doi:10.3171/2019.4.PEDS19113
- Athale UH, Duckworth J, Odame I, Barr R. Childhood atypical teratoid rhabdoid tumor of the central nervous system: A meta-analysis of observational studies. *J Pediatr Hematol Oncol.* 2009;31(9):651-663. doi:10.1097/MPH.0b013e3181b258a9
- Buccoliero AM, Caporalini C, Scagnet M, et al. A diagnostic pitfall: Atypical teratoid rhabdoid tumor versus dedifferentiated/poorly differentiated chordoma: Analysis of a mono-institutional series. *Appl Immunohistochem Mol Morphol.* 2019; 27(2):147-154. doi:10.1097/PAI.0000000000000554
- Ho DM, Hsu CY, Wong TT, Ting LT, Chiang H. Atypical teratoid/rhabdoid tumor of the central nervous system: A comparative study with primitive neuroectodermal tumor/medulloblastoma. *Acta Neuropathol.* 2000;99(5):482-488. doi:10.1007/s004010051149
- Zin F, Cotter JA, Haberler C, et al. Histopathological patterns in atypical teratoid/rhabdoid tumors are related to molecular subgroup. *Brain Pathol.* 2021;31(5):e12967. doi:10.1111/bpa.12967
- Holdhof D, Johann PD, Spohn M, et al. Atypical teratoid/rhabdoid tumors (ATRTs) with SMARCA4 mutation are molecularly distinct from SMARCB1-deficient cases. *Acta Neuropathol.* 2021;141(2):291-301. doi:10.1007/s00401-020-02250-7
- Nesvick CL, Lafay-Cousin L, Raghunathan A, Bouffet E, Huang AA, Daniels DJ. Atypical teratoid rhabdoid tumor: Molecular insights and translation to novel therapeutics. *J Neuro Oncol.* 2020;150(1):47-56. doi:10.1007/s11060-020-03639-w
- Ud Din N, Barakzai A, Memon A, Hasan S, Ahmad Z. Atypical teratoid/rhabdoid tumor of brain: A clinicopathologic study of eleven patients and review of literature. *Asian Pac J Cancer Prev.* 2017;18(4):949-954. doi:10.22034/APJCP.2017.18.4.949
- Calandrelli R, Massimi L, Pilato F, et al. Atypical teratoid rhabdoid tumor: Proposal of a diagnostic pathway based on clinical features and neuroimaging findings. *Diagnostics.* 2023;13(3):475. doi:10.3390/diagnostics13030475

14. Rechberger JS, Nesvick CL, Daniels DJ. Atypical teratoid rhabdoid tumor (ATRT): Disease mechanisms and potential drug targets. *Expert Opin Ther Targets*. 2022; 26(3):187-192. doi:10.1080/14728222.2022.2040017
15. Biswas A, Ghosh V, Roy S, et al. Spinal atypical teratoid rhabdoid tumor—narrative review and report of a rare case managed with multimodality approach. *Childs Nerv Syst*. 2023;39(8):2019-2026. doi:10.1007/s00381-023-05977-2
16. Kleihues P, Louis DN, Scheithauer BW, et al. The WHO classification of tumors of the nervous system. *J Neuropathol Exp Neurol*. 2002;61(3):215-229. doi:10.1093/jnen/61.3.215
17. Louis DN, Ohgaki H, Wiestler OD, et al. The 2007 WHO classification of tumours of the central nervous system. *Acta Neuropathol*. 2007;114(2):97-109. doi:10.1007/s00401-007-0243-4
18. Cheng SW, Davies KP, Yung E, Beltran RJ, Yu J, Kalpana GV. c-MYC interacts with INI1/hSNF5 and requires the SWI/SNF complex for transactivation function. *Nat Genet*. 1999;22(1):102-105. doi:10.1038/8811
19. Li Y, Yang X, Zhu W, et al. SWI/SNF complex gene variations are associated with a higher tumor mutational burden and a better response to immune checkpoint inhibitor treatment: A pan-cancer analysis of next-generation sequencing data corresponding to 4591 cases. *Cancer Cell Int*. 2022;22(1):347. doi:10.1186/s12935-022-02757-x
20. Torchia J, Picard D, Lafay-Cousin L, et al. Molecular subgroups of atypical teratoid rhabdoid tumours in children: An integrated genomic and clinicopathological analysis. *Lancet Oncol*. 2015;16(5):569-582. doi:10.1016/S1470-2045(15)70114-2
21. Johann PD, Erkek S, Zaparka M, et al. Atypical teratoid/rhabdoid tumors are comprised of three epigenetic subgroups with distinct enhancer landscapes. *Cancer Cell*. 2016;29(3):379-393. doi:10.1016/j.ccell.2016.02.001
22. Han ZY, Richer W, Fréneaux P, et al. The occurrence of intracranial rhabdoid tumours in mice depends on temporal control of Smarcb1 inactivation. *Nat Commun*. 2016;7:10421. doi:10.1038/ncomms10421
23. Torchia J, Golbourn B, Feng S, et al. Integrated (epi)-Genomic analyses identify subgroup-specific therapeutic targets in CNS rhabdoid tumors. *Cancer Cell*. 2016; 30(6):891-908. doi:10.1016/j.ccell.2016.11.003
24. Kleinert R. Immunohistochemical characterization of primitive neuroectodermal tumors and their possible relationship to the stepwise ontogenetic development of the central nervous system. 1. Ontogenetic studies. *Acta Neuropathol*. 1991;82(6): 502-507. doi:10.1007/BF00293385
25. Rorke LB, Packer RJ, Biegel JA. Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood: Definition of an entity. *J Neurosurg*. 1996;85(1): 56-65. doi:10.3171/jns.1996.85.1.0056
26. Wu HY, Xu WB, Lu LW, et al. Imaging features of spinal atypical teratoid rhabdoid tumors in children. *Medicine (Baltim)*. 2018;97(52):e13808. doi:10.1097/MD.000000000013808
27. Yang CS, Jan YJ, Wang J, Shen CC, Chen CC, Chen M. Spinal atypical teratoid/rhabdoid tumor in a 7-year-old boy. *Neuropathology*. 2007;27(2):139-144. doi:10.1111/j.1440-1789.2007.00752.x
28. Dufour C, Beaugrand A, Le Deley MC, et al. Clinicopathologic prognostic factors in childhood atypical teratoid and rhabdoid tumor of the central nervous system: A multicenter study. *Cancer*. 2012;118(15):3812-3821. doi:10.1002/cncr.26684
29. Parmar H, Hawkins C, Bouffier E, Rutka J, Shroff M. Imaging findings in primary intracranial atypical teratoid/rhabdoid tumors. *Pediatr Radiol*. 2006;36(2):126-132. doi:10.1007/s00247-005-0037-6
30. Tantana S, Sotelo-Avila C, Pilla TJ, Sundaram M. Malignant rhabdoid tumor of the thoracic spine. *Orthopedics*. 1988;11(6):905-908. doi:10.3928/0147-7447-19880601-10
31. Horie H, Etoh T, Maie M. Cytogenetic characteristics of a malignant rhabdoid tumor arising from the paravertebral region. A case report. *Acta Pathol Jpn*. 1992; 42(6):460-465. doi:10.1111/j.1440-1827.1992.tb03253.x
32. Rosemberg S, Menezes Y, Sousa MR, Plese P, Ciquini O. Primary malignant rhabdoid tumor of the spinal dura. *Clin Neuropathol*. 1994;13(4):221-224.
33. Tamiya T, Nakashima H, Ono Y, et al. Spinal atypical teratoid/rhabdoid tumor in an infant. *Pediatr Neurosurg*. 2000;32(3):145-149. doi:10.1159/000028920
34. Bambakidis NC, Robinson S, Cohen M, Cohen AR. Atypical teratoid/rhabdoid tumors of the central nervous system: Clinical, radiographic and pathologic features. *Pediatr Neurosurg*. 2002;37(2):64-70. doi:10.1159/000065107
35. Mahmood MN, Salama ME, Shah VV. Pathologic quiz case: An 11-year-old boy with cervical lymphadenopathy. Metastatic malignant extrarenal rhabdoid tumor. *Arch Pathol Lab Med*. 2003;127(8):e361-e362. doi:10.5858/2003-127-e361-PQCAYB
36. Cheng YC, Lirng JF, Chang FC, et al. Neuroradiological findings in atypical teratoid/rhabdoid tumor of the central nervous system. *Acta Radiol*. 2005;46(1): 89-96. doi:10.1080/02841850510020987
37. Chen ML, McComb JG, Krieger MD. Atypical teratoid/rhabdoid tumors of the central nervous system: management and outcomes. *Neurosurg Focus*. 2005;18(6A): E8.
38. Tanizaki Y, Oka H, Utsuki S, Shimizu S, Suzuki S, Fujii K. Atypical teratoid/rhabdoid tumor arising from the spinal cord—case report and review of the literature. *Clin Neuropathol*. 2006;25(2):81-85.
39. Bannykh S, Duncan C, Ogle E, Baehring JM. Atypical teratoid/rhabdoid tumor of the spinal canal. *J Neuro Oncol*. 2006;76(2):129-130. doi:10.1007/s11060-006-4032-z
40. Moeller KK, Coventry S, Jernigan S, Moriarty TM. Atypical teratoid/rhabdoid tumor of the spine. *AJNR Am J Neuroradiol*. 2007;28(3):593-595.
41. Kodama H, Maeda M, Imai H, Matsubara T, Taki W, Takeda K. MRI of primary spinal atypical teratoid/rhabdoid tumor: A case report and literature review. *J Neuro Oncol*. 2007;84(2):213-216. doi:10.1007/s11060-007-9359-6
42. Tinsa F, Jallouli M, Douira W, et al. Atypical teratoid/rhabdoid tumor of the spine in a 4-year-old girl. *J Child Neurol*. 2008;23(12):1439-1442. doi:10.1177/0883073808319319
43. Yano S, Hida K, Kobayashi H, Iwasaki Y. Successful multimodal therapies for a primary atypical teratoid/rhabdoid tumor in the cervical spine. *Pediatr Neurosurg*. 2008;44(5):406-413. doi:10.1159/000149910
44. Agrawal A, Bhake A, Cincu R. Giant lumbar paraspinous atypical teratoid/rhabdoid tumor in a child. *J Cancer Res Ther*. 2009;5(4):318-320. doi:10.4103/0973-1482.59906
45. Niwa T, Aida N, Tanaka M, et al. Diffusion-weighted imaging of an atypical teratoid/rhabdoid tumor of the cervical spine. *Magn Reson Med Sci*. 2009;8(3): 135-138. doi:10.2463/mrms.8.135
46. Fridley JS, Chamoun RB, Whitehead WE, et al. Malignant rhabdoid tumor of the spine in an infant: Case report and review of the literature. *Pediatr Neurosurg*. 2009; 45(3):237-243. doi:10.1159/000224622
47. Mohapatra I, Santosh V, Chickabasaviah YT, et al. Histological and immunohistochemical characterization of AT/RT: A report of 15 cases from India. *Neuropathology*. 2010;30(3):251-259. doi:10.1111/j.1440-1789.2009.01075.x
48. Heuer GG, Kiefer H, Judkins AR, et al. Surgical treatment of a clival-C2 atypical teratoid/rhabdoid tumor. *J Neurosurg Pediatr*. 2010;5(1):75-79. doi:10.3171/2009.8.PEDS08421
49. Stabouli S, Sdougka M, Tsitsopoulos P, et al. Primary atypical teratoid/rhabdoid tumor of the spine in an infant. *Hippokratia*. 2010;14(4):286-288.
50. Huddleston BJ, Sjostrom CM, Collins BT. Atypical teratoid/rhabdoid tumor involving cerebrospinal fluid: a case report. *Acta Cytol*. 2010;54(5 Suppl):958-962.
51. Dobbs MD, Correa H, Schwartz HS, Kan JH. Extrarenal rhabdoid tumor mimicking a sacral peripheral nerve sheath tumor. *Skeletal Radiol*. 2011;40(10): 1363-1368. doi:10.1007/s00256-011-1160-0
52. Makis W, Ciarallo A, Hickey M. Malignant extrarenal rhabdoid tumor of the spine: Staging and evaluation of response to therapy with F-18 FDG PET/CT. *Clin Nucl Med*. 2011;36(7):599-602. doi:10.1097/RLU.0b013e31821773f0
53. Imagama S, Wakao N, Ando K, et al. Treatment for primary spinal atypical teratoid/rhabdoid tumor. *J Orthop Sci*. 2012;17(6):822-827. doi:10.1007/s00776-011-0122-7
54. Kelley BJ, Johnson MH, Vortmeyer AO, Smith BG, Abbed KM. Two-level thoracic pedicle subtraction osteotomy for progressive post-laminectomy kyphotic deformity following resection of an unusual thoracolumbar intradural extramedullary tumor. *J Neurosurg Pediatr*. 2012;10(4):334-339. doi:10.3171/2012.7.PEDS11526
55. Yang M, Chen X, Wang N, et al. Primary atypical teratoid/rhabdoid tumor of central nervous system in children: a clinicopathological analysis and review of literature in China. *Int J Clin Exp Pathol*. 2014;7(5):2411-2420.
56. Xin X, Zhu B, Shen J, Tian C, Fan X, Liu BR. A primary spinal extradural atypical teratoid/rhabdoid tumor of the cervical spine with bony involvement. *J Child Neurol*. 2014;29(5):670-673. doi:10.1177/0883073813509017
57. Dhir A, Tekautz T, Recinos V, et al. Lumbar spinal atypical teratoid rhabdoid tumor. *J Clin Neurosci*. 2015;22(12):1988-1989. doi:10.1016/j.jocn.2015.06.007
58. Tang Y, Li S, Qu J, Zhou Y, Xiao J. Malignant rhabdoid tumor with cervical vertebra involvement in a teenage child: case report and review of the literature. *Pediatr Neurosurg*. 2015;50(3):173-178. doi:10.1159/000430448
59. Mankotia DS, Tandon V, Sharma BS, Rajeshwari M, Sharma MC. A case of primary spinal atypical teratoid/rhabdoid tumor in a 5-year-old child. *J Pediatr Neurosci*. 2016;11(2):121-124. doi:10.4103/1817-1745.187634
60. Chao MF, Su YF, Jaw TS, Chiou SS, Lin CH. Atypical teratoid/rhabdoid tumor of lumbar spine in a toddler child. *Spinal Cord Ser Cases*. 2017;3:16026. doi:10.1038/scsanc.2016.26
61. Babgi M, Samkari A, Al-Mehdar A, Abdullah S. Atypical teratoid/rhabdoid tumor of the spinal cord in a child: Case report and comprehensive review of the literature. *Pediatr Neurosurg*. 2018;53(4):254-262. doi:10.1159/000488459
62. Singla N, Kapoor A, Chatterjee D, Radotra BD. Ultra early recurrence in giant congenital malignant rhabdoid tumor of spine. *Childs Nerv Syst*. 2016;32(12): 2471-2474. doi:10.1007/s00381-016-3178-z
63. Nishimoto T, Nomura S, Fukano R, Kimura T, Ikeda E, Suzuki M. A primary extradural malignant rhabdoid tumor at the craniocervical junction in a 3-year-old boy. *Childs Nerv Syst*. 2018;34(2):367-371. doi:10.1007/s00381-017-3631-7
64. Hong S, Ogiwara H. Dumbbell-shaped atypical teratoid rhabdoid tumor in the cervical spine mimicking schwannoma. *Childs Nerv Syst* 2018;34(1):27-28.

65. Garling RJ, Singh R, Harris C, Haridas A. Intradural lumbosacral malignant extrarenal rhabdoid tumor: A case report. *Childs Nerv Syst.* 2018;34(1):165-167. doi:10.1007/s00381-017-3571-2
66. Hong S, Ogiwara H. Dumbbell-shaped atypical teratoid rhabdoid tumor in the cervical spine mimicking schwannoma. *Childs Nerv Syst.* 2018;34(1):27-28. doi:10.1007/s00381-017-3603-y
67. Tsitsopoulos PP, Marinos K, Chochliourou E, et al. Infantile atypical teratoid rhabdoid tumor of the spine presenting with acute hydrocephalus. *Pediatr Neurosurg.* 2020;55(5):313-318. doi:10.1159/000511423
68. Kramer DE, Kerolus MG, Nunna RS, et al. Atypical teratoid/rhabdoid tumor of the conus medullaris. *Pediatr Neurosurg.* 2020;55(4):215-221. doi:10.1159/000509165
69. Shiflett JM, Herrington BL, Joyner DA, Saad AG. Atypical teratoid rhabdoid tumor of the cauda equina in a child: Report of a very unusual case. *Appl Immunohistochem Mol Morphol.* 2020;28(7):e58-e62. doi:10.1097/PAI.0000000000000620
70. Meena RK, Doddamani RS, Chipde H, Mahajan S, Chandra SP, Sawarkar DP. Primary spinal atypical teratoid/rhabdoid tumour presenting with hematomyelia and subarachnoid haemorrhage—a case report. *Childs Nerv Syst.* 2020;36(3):655-659. doi:10.1007/s00381-019-04412-9
71. Biswas A, Velu U, Sharma S, et al. Successful multimodality management of atypical teratoid/rhabdoid tumour of the lower dorsal spine with spinal drop metastasis: Illustrated review. *Pediatr Neurosurg.* 2021;56(2):184-196. doi:10.1159/000513936
72. Egiz A, Kannan S, Asl SF. The impact of surgical resection and adjuvant therapy on survival in pediatric patients with atypical teratoid/rhabdoid tumor: Systematic review and pooled survival Analysis. *World Neurosurg.* 2022;164:216-227. doi:10.1016/j.wneu.2022.04.073
73. Chi SN, Zimmerman MA, Yao X, et al. Intensive multimodality treatment for children with newly diagnosed CNS atypical teratoid rhabdoid tumor. *J Clin Oncol.* 2009;27(3):385-389. doi:10.1200/JCO.2008.18.7724
74. Reddy AT, Strother DR, Judkins AR, et al. Efficacy of high-dose chemotherapy and three-dimensional conformal radiation for atypical teratoid/rhabdoid tumor: A report from the children's oncology group trial ACNS0333. *J Clin Oncol.* 2020;38(11):1175-1185. doi:10.1200/JCO.19.01776
75. Yuh GE, Loredó LN, Yonemoto LT, et al. Reducing toxicity from craniospinal irradiation: Using proton beams to treat medulloblastoma in young children. *Cancer J.* 2004;10(6):386-390. doi:10.1097/00130404-200411000-00009
76. Kuhlthau KA, Pulsifer MB, Yeap BY, et al. Prospective study of health-related quality of life for children with brain tumors treated with proton radiotherapy. *J Clin Oncol.* 2012;30(17):2079-2086. doi:10.1200/jco.2011.37.0577