

## CASE REPORT

## Pediatrics

# Atypical teratoid rhabdoid tumor in the cerebellum of a 7-year-old boy presenting with headache after a fall

Leena Iqbal MD | Philip S. Nawrocki MD | Christine Radivoj MSN, APRN

Department of Emergency Medicine, Trinity Medical Center West, Steubenville, Ohio, USA

**Correspondence**

Leena Iqbal, MD, Department of Emergency Medicine, Trinity Medical Center West, 4000 Johnson Road, Steubenville, OH 43952, USA.  
Email: [iqballeena@gmail.com](mailto:iqballeena@gmail.com)

**Funding and support:** By *JACEP Open* policy, all authors are required to disclose any and all commercial, financial, and other relationships in any way related to the subject of this article as per ICMJE conflict of interest guidelines (see [www.icmje.org](http://www.icmje.org)). The authors have stated that no such relationships exist.

**Abstract**

A 7-year-old boy presented to a community emergency department (ED) after sustaining a minor fall. Although he was found to have a normal neurologic examination, additional history revealed the patient had been having mild intermittent headaches and dizziness in the months preceding the fall. The emergency clinicians ordered neuroimaging, which demonstrated a right cerebellar mass, ultimately diagnosed as atypical rhabdoid/teratoid tumor. Atypical rhabdoid/teratoid tumor is a rare, aggressive brain tumor with a poor prognosis. The objectives of this case report are to emphasize the importance of detailed history with pediatric head trauma, in particular on reassessment, and to discuss briefly the epidemiology and management of atypical teratoid rhabdoid tumor.

**KEYWORDS**

atypical teratoid rhabdoid tumor, central nervous system tumor, pediatric headache

## 1 | INTRODUCTION

Head trauma is one of the most common reasons children are evaluated in the emergency department (ED). Traumatic brain injury from head trauma is diagnosed in nearly 1 million ED visits annually in the United States.<sup>1</sup> Fall and motor vehicle accidents are the most common mechanisms.<sup>2</sup> The vast majority of patients with traumatic brain injury have mild injuries that resolve uneventfully over days to weeks.

Clinically important traumatic brain injury is rare, including intracranial bleeding requiring intervention. The risks of ionizing radiation, concerns about imaging overuse, and the low frequency of significant injury prompted efforts to develop clinical decision aides to identify patients at low risk for clinically significant traumatic brain injury. The Pediatric Emergency Care Applied Research Network (PECARN) criteria are perhaps the most prominent and widely used decision aids for pediatric head trauma.<sup>3</sup> Although the PECARN criteria are well validated and highly accurate, they were designed to identify patients at

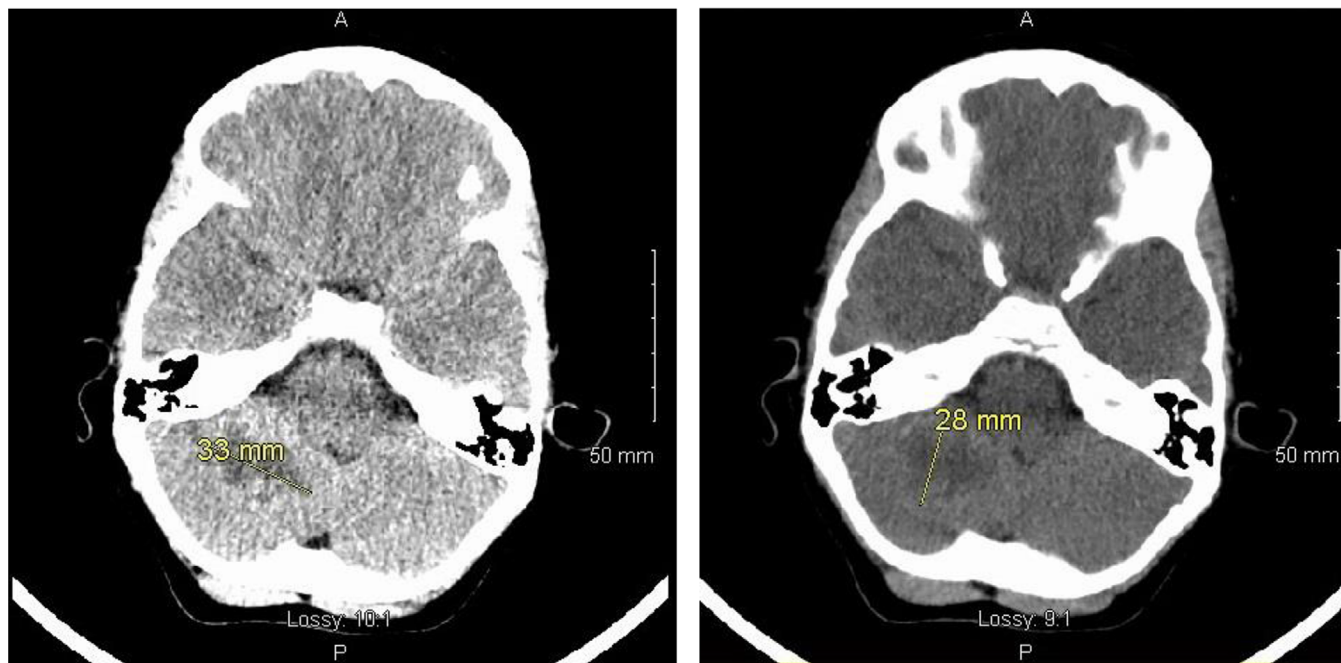
low risk of traumatic brain injury, not all serious central nervous system disease processes. As head and other trauma are common in children, a history of trauma may be a red herring. Thus, clinicians are at risk of anchoring on head trauma and may miss other less common central nervous system disease processes. Use of the PECARN criteria may falsely reassure clinicians in these cases, preventing or limiting reassessment and additional history taking.

We describe the case of a 7-year-old boy who presented to our community ED after an unwitnessed, ground-level fall without loss of consciousness. The patient had a normal neurological assessment and was in the lowest risk group for clinically important traumatic brain injury by the PECARN criteria. The emergency physician obtained additional history during reassessment of the patient, which was concerning for brain tumor. The objectives of this report are to emphasize the importance of detailed and especially repeated history taking and to discuss briefly the epidemiology and management of atypical rhabdoid/teratoid tumor.

Supervising Editor: Benjamin T. Kerrey, MD, MS.

This is an open access article under the terms of the [Creative Commons Attribution-NonCommercial-NoDerivs](https://creativecommons.org/licenses/by-nc-nd/4.0/) License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.

© 2021 The Authors. *JACEP Open* published by Wiley Periodicals LLC on behalf of the American College of Emergency Physicians.



**FIGURE 1** Computed tomography imaging of the brain demonstrating a low-density right cerebellar mass measuring 3.3 cm × 2.8 cm with local mass effect.

## 2 | CASE

A 7-year-old boy was brought by his mother to a community ED after sustaining a ground-level fall. He reported that he was running in gym class and struck the back of his head after falling. The fall was witnessed by the school staff, and the mother reported she was told there was no loss of consciousness. Immediately after the fall, the patient complained of dizziness to his teacher, who informed his mother. The patient and his mother denied nausea, vomiting, and alteration in his level of consciousness since the fall. His mother reported that he was born at 36 weeks of gestation with an uncomplicated neonatal course. He had been generally healthy, with no history of serious head injury or significant disease.

The patient was triaged by the nursing staff as Emergency Severity Index level 4. He was placed in the fast track of the ED and seen by a mid-level clinician. On initial examination, the child was found to be awake and alert, with a Glasgow Coma Score of 15. There was no external evidence of injury to his head or neck; he denied tenderness to the cervical spine and had full range of motion of his neck without pain. He had no focal deficits on complete neurologic examination, including a normal gait and normal speech.

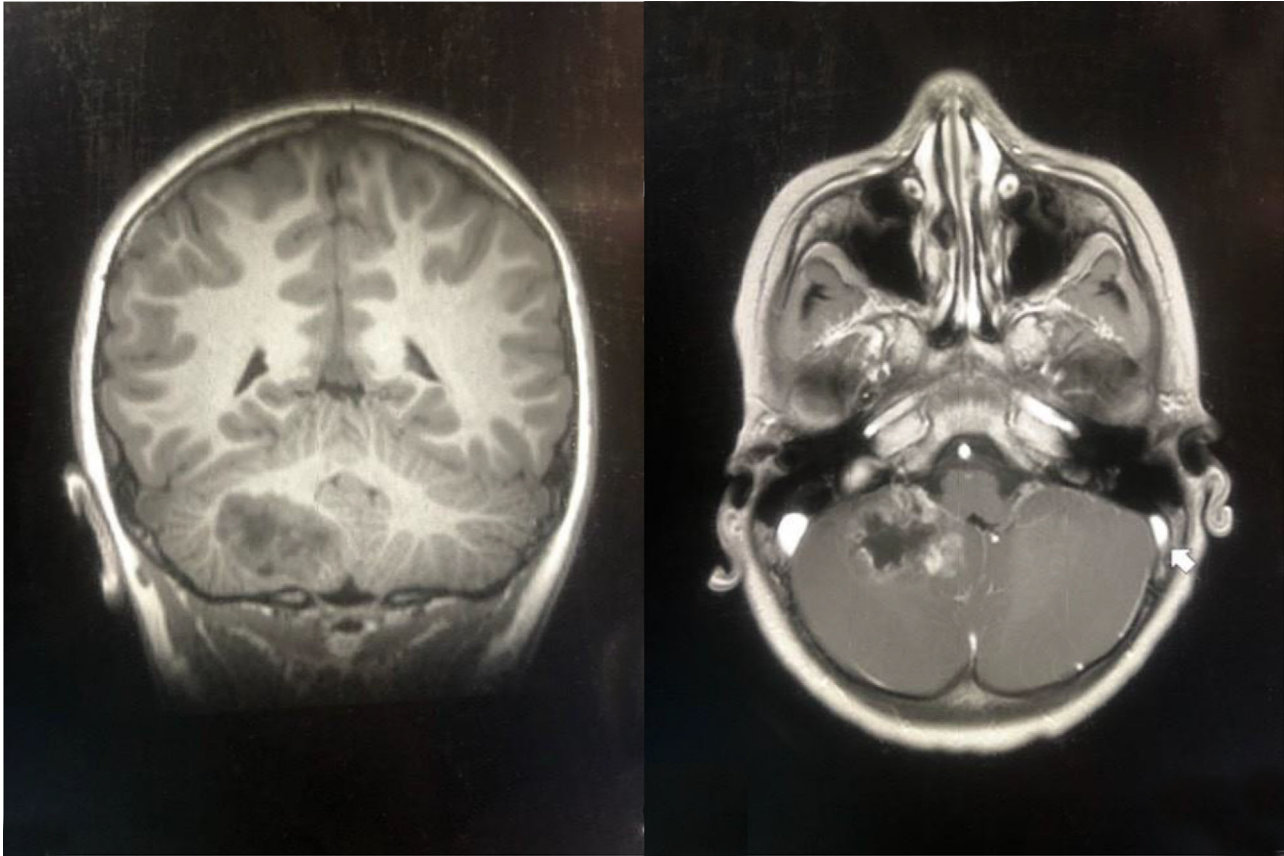
The mid-level clinician applied the PECARN head injury criteria and determined that the patient was in the lowest risk group for clinically important traumatic brain injury. Based on this assessment, a computed tomography (CT) scan of the head was not indicated. The mid-level clinician did not order imaging but wanted to discuss this decision further with the attending physician. The mid-level clinician and attending physician reassessed the patient together. The attending physician's examination and assessment, including PECARN criteria, were unchanged.

The attending physician and the mother discussed the decision not to order imaging. During this discussion, the mother expressed concerns that the patient had been experiencing intermittent headaches and dizziness before the fall. The patient confirmed he had been having headaches and occasional dizziness for about 4 months. He described the headaches as mild, aching, diffuse, and intermittent. The headaches reportedly had not increased in intensity or in frequency, including in the early morning or on awakening. The mother and the patient denied other falls before the index visit. They also denied nausea and vomiting and changes in vision, physical activity, or gait during the period he had been having headaches. The mother again denied a family history of headaches. The mother reported that the patient was evaluated by his primary care physician for headaches 2 months before the index visit and that she had been keeping a headache diary.

Because of the additional history obtained during the reassessment, the mid-level clinician and attending physician ordered a head CT. A head CT revealed a low-density right cerebellar mass, measuring 3.3 cm × 2.8 cm and with local mass effect (Figure 1). The ED's institution did not have pediatric neurosurgical services, and the child was transferred to a pediatric tertiary care facility.

## 3 | DISCUSSION

The diagnosis of pediatric patients with rare but serious diseases is challenging. With central nervous system tumors, the clinical variation is very wide, based on tumor type and location and patient age and stage of development. In a systematic review of 74 studies, 17 individual symptoms or signs were present in at least 5% of children among all types of central nervous system tumors; 56 total symptoms or signs



**FIGURE 2** Magnetic resonance imaging of the brain demonstrating a right cerebellar centrally necrotic mass measuring 3.7 cm

were recorded in at least 1 patient.<sup>4</sup> The relative rarity of central nervous system tumors in children combined with a high degree of clinical variation and subtle or non-specific symptoms, such as headache, increase the time to diagnosis. In a 6-year study of children with brain tumors in Austria, the median time from symptom onset to diagnosis was 60 days.<sup>5</sup>

Headache and abnormal gait are common symptoms in the pediatric patient in the ED, including both those with common conditions such as head trauma and in patients with relatively rare conditions such as a central nervous system tumor. In the aforementioned systematic review that included >4000 pediatric patients, headache (33%) and abnormalities of gait and coordination (27%) were among the most common presenting signs and symptoms.<sup>4</sup> In a separate study of pediatric patients with central nervous system tumors presenting to an ED, two-thirds complained of headache at presentation (66.7%), and almost half of the patients reported some disturbance in gait (42.5%).<sup>6</sup> In a secondary analysis of children with central nervous system tumors from the English National Audit of Cancer Diagnosis in Primary Care, a history of headache and episodes of fainting or falls were each present in 20% of patients.<sup>7</sup>

After a fall and possible head injury, complaints of headache and dizziness will typically prompt clinicians to focus on traumatic brain injury. Clinically important traumatic brain injury is the chief concern, especially various forms of intracranial bleeding that may require urgent intervention. The incidence of clinically important

traumatic brain injury is very low, however, especially in children with ground-level mechanisms and normal neurologic examinations. In the original PECARN head injury study, among the >40,000 patients enrolled, although head CT was performed for >30% of the patients, only 0.9% had clinically important traumatic brain injury, and only 0.1% of the entire sample required neurosurgical intervention.<sup>3</sup>

Emergency medicine and pediatric authorities have long been advocating for the judicious use of neuroimaging in children after head trauma. One study estimated that of the 600,000 abdominal and head CT scans performed annually in the United States, 500 children might ultimately die from cancer attributed to CT radiation exposure.<sup>8</sup> The original PECARN head injury study has been externally validated<sup>9,10</sup> and provides an opportunity to decrease the use of head CT, although evidence suggests that is not yet happening outside of academic pediatric centers.<sup>8</sup>

Applying the PECARN criteria to the patient described in this case, the clinicians determined that the patient was in the lowest risk group and a CT scan of the head was not indicated. Although the clinical decision aid was correct in predicting a low probability of clinically important traumatic brain injury, decision aids such as the PECARN criteria must be interpreted in the context for which they were designed. In this case, the initial clinical focus was understandably on traumatic brain injury. In most patients with this history, even mild traumatic brain injury would be relatively unlikely, with most patients appearing well and symptom free by the time of ED evaluation. The reassessment and

additional history in our patient, who had a normal neurologic assessment, were essential to the clinician's recognition that this patient was different. Complaints of symptoms preceding a history of minor trauma may be easily missed. Anchoring on a diagnosis heavily framed by the initial details of a patient's presentation is likely a key contributor to missing serious but rare diagnoses. The clinicians in this case, however, were not so anchored on traumatic brain injury that they ignored or dismissed the mother's and patient's concerns. Intentionally reassessing a patient combined with the mental habits of openness to additional history and challenging initial conceptions may be among the most effective tools available to clinicians to avoid missing diagnoses. Although relatively rare, clinician recognition of symptoms preceding a fall or other minor trauma may be essential to earlier diagnosis of central nervous system tumors, and delayed diagnosis is associated with greater morbidity.<sup>11</sup>

### 3.1 | Case resolution

At the pediatric tertiary care center, magnetic resonance imaging of the brain and spine confirmed a tumor in the right cerebellar hemisphere with central necrosis (Figure 2). The patient had the tumor resected and a ventriculoperitoneal shunt placed because of concern for obstructive hydrocephalus. The tumor was histologically classified as an atypical teratoid rhabdoid tumor.

### 3.2 | Atypical teratoid rhabdoid tumor

Central nervous system tumors are the second most common childhood malignancy, comprising up to 20% of all childhood cancers.<sup>12</sup> The existing literature on atypical teratoid rhabdoid tumor mostly consists of case series of infants or adults and is primarily derived from tumor registries. Atypical teratoid rhabdoid tumor is most common in male children <3 years of age<sup>13</sup> and most often involves the cerebellum, ventricles, and frontal lobes. One study from the Central Brain Tumor Registry of the United States found atypical teratoid rhabdoid tumor comprised 1.6% of all central nervous system tumors in persons 19 years of age and younger.<sup>14</sup>

Atypical teratoid rhabdoid tumor is highly malignant and aggressive, and the prognosis is therefore poor. A 10-year study found an overall median survival of 11 months after diagnosis. The literature suggests improved survival with older age and less extensive disease. Among children treated with radiation and high-dose alkylating agents, one study showed a 2-year overall survival of 98% in children 3 years of age and older compared with 17% in children younger than 3 years of age.<sup>13</sup> Improved outcomes have also been associated with multimodal therapy, radiotherapy in patients older than 3 years of age, and gross total resection compared with less extensive surgery.<sup>14,15</sup> Our patient had no metastasis and underwent gross total resection. He was subsequently enrolled in the Saint Jude's Phase 2 Study of Alisertib Therapy for Rhabdoid Tumors (SJATRT)<sup>16</sup> and received craniospinal radiation along with Alisertib treatment. At the 24-month follow-up, there

was no reported recurrence of disease or symptoms. We believe early diagnosis at a stage without metastasis may have played a role in his outcome.

## 4 | CONCLUSION

Central nervous system tumors in pediatric patients are challenging to diagnose given the clinical variation, rarity, and overlap in symptomatology with more common and relatively benign conditions such as minor traumatic brain injury. Intentionally reassessing a patient after an initial history and examination, including the mental habit of challenging the initial clinical framing, can allow emergency clinicians the opportunity to identify patients at risk for serious but rare diagnoses, such as central nervous system tumors. Atypical teratoid rhabdoid tumor is a highly malignant tumor with a poor prognosis, although patients with earlier diagnosis before metastasis may have better outcomes.

## REFERENCES

- Corwin DJ, Grady MF, Joffe MD, Zonfrillo MR, Pediatric mild traumatic brain injury in the acute setting. *Pediatr Emerg Care*. 2017;33(9):643-649.
- Dewan MC, Mummareddy N, Wellons JC, Bonfield CM, Epidemiology of global pediatric traumatic brain injury: qualitative review. *World Neurosurg*. 2016;91:497-509.e1.
- Kuppermann N, Holmes JF, Dayan PS, et al. Identification of children at very low risk of clinically-important brain injuries after head trauma: a prospective cohort study. *Lancet*. 2009;374(9696):1160-1170.
- Wilne S, Collier J, Kennedy C, Koller K, Grundy R, Walker D, Presentation of childhood CNS tumours: a systematic review and meta-analysis. *Lancet Oncol*. 2007;8(8):685-695.
- Azizi AA, Heßler K, Leiss U, et al. From symptom to diagnosis-the pre-diagnostic symptomatic interval of pediatric central nervous system tumors in Austria. *Pediatr Neurol*. 2017;76:27-36.
- Snyder H, Robinson K, Shah D, Brennan R, Handrigan M, Signs and symptoms of patients with brain tumors presenting to the emergency department. *J Emerg Med*. 1993;11(3):253-258.
- Ozawa M, Brennan PM, Zienius K, et al. Symptoms in primary care with time to diagnosis of brain tumours. *Fam Pract*. 2018;35(5):551-558.
- Burstein B, Upton JEM, Terra HF, Neuman MI, Use of CT for Head Trauma: 2007-2015. *Pediatrics*. 2018;142(4):e20180814.
- Schonfeld D, Bressan S, Da Dalt L, Henien MN, Winnett JA, Nigrovic LE, Pediatric emergency care applied research network head injury clinical prediction rules are reliable in practice. *Arch Dis Child*. 2014;99(5):427-431.
- Easter JS, Bakes K, Dhaliwal J, Miller M, Caruso E, Haukoos JS, Comparison of PECARN, CATCH, and CHALICE rules for children with minor head injury: a prospective cohort study. *Ann Emerg Med*. 2014;64(2):145-152, 152.e1-5.
- Wilne SH, Dineen RA, Dommett RM, Chu TPC, Walker DA, Identifying brain tumours in children and young adults. *BMJ*. 2013;347:f5844.
- Udaka YT, Packer RJ, Pediatric brain tumors. *Neurol Clin*. 2018;36(3):533-556.
- Tekautz TM, Fuller CE, Blaney S, et al. Atypical teratoid/rhabdoid tumors (ATRT): improved survival in children 3 years of age and older with radiation therapy and high-dose alkylator-based chemotherapy. *J Clin Oncol*. 2005;23(7):1491-1499.
- Richardson EA, Ho B, Huang A, Atypical teratoid rhabdoid tumour: from tumours to therapies. *J Korean Neurosurg Soc*. 2018;61(3):302-311.

15. Richards A, Ved R, Murphy C, et al. Outcomes with respect to extent of surgical resection for pediatric atypical teratoid rhabdoid tumors. *Childs Nerv Syst.* 2020;36(4):713-719.
16. Phase 2 Study of Alisertib Therapy for Rhabdoid Tumors—Full Text View—ClinicalTrials.gov. <https://clinicaltrials.gov/ct2/show/NCT02114229>. Accessed May 7, 2020.

**How to cite this article:** Iqbal L, Nawrocki PS, Radivoj C. Atypical teratoid rhabdoid tumor in the cerebellum of a 7-year-old boy presenting with headache after a fall. *JACEP Open.* 2021;2:e12353. <https://doi.org/10.1002/emp2.12353>