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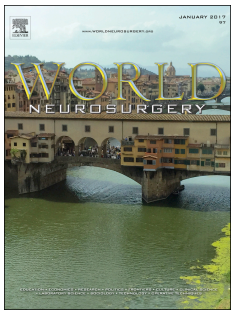

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Garret P. Greeneway, MD, Paul S. Page, MD, Viharkumar Patel, MD, Azam S. Ahmed, MD

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Garret P. Greeneway, MD[1]; Paul S. Page, MD[1]; Viharkumar Patel, MD[2]; Azam S. Ahmed, MD[1]

[1]Department of Neurological Surgery, University of Wisconsin School of Medicine and Public Health, Madison, WI, USA. Affiliation address: Department of Neurological Surgery, 600 Highland Avenue, K4/8 CSC, Box 8660, Madison, WI, USA 53792

[2]Department of Pathology and Laboratory Medicine, University of Wisconsin School of Medicine and Public Health, Madison, WI, USA. Affiliation address: Department of Pathology and Laboratory Medicine, 3170 UW Medical Foundation Centennial Building, 1685 Highland Avenue, Madison, WI, USA 53705

Corresponding author: Garret P. Greeneway

Attn: Garret P. Greeneway

Department of Neurological Surgery

600 Highland Avenue, K4/8 CSC, Box 8660

Madison, WI, USA 53792

Phone: 608-263-1410

Corresponding author's email: gpgreeneway@gmail.com

Declarations of interest: None.

Key words: Adult; Atypical teratoid/rhabdoid tumor; Brain tumor; Cerebellar tumor; Cerebellum; Rhabdoid tumor

Short title: AT/RT of the cerebellum in an adult

Abbreviations list: AT/RT – atypical teratoid/rhabdoid tumor; CNS – central nervous system; CSF – cerebrospinal fluid; CT – computed tomography; EMA – epithelial membrane antigen; GFAP – Glial fibrillary acidic protein; Gy – gray; INI-1 – integrase interactor-1; MRI – magnetic resonance imaging; SMARCB1 – SWI/SNF-related matrix-associated actin-dependent regulator of chromatin subfamily B member 1; WHO – World Health Organization

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[1]Department of Neurological Surgery, University of Wisconsin School of Medicine and Public Health, Madison, WI, USA. Affiliation address: Department of Neurological Surgery, 600 Highland Avenue, K4/8 CSC, Box 8660, Madison, WI, USA 53792

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Corresponding author: Garret P. Greeneway

Attn: Garret P. Greeneway

Department of Neurological Surgery

600 Highland Avenue, K4/8 CSC, Box 8660

Madison, WI, USA 53792

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Abstract

Background: Atypical teratoid/rhabdoid tumor (AT/RT) is a rare tumor that is most frequently encountered in the pediatric patient population. AT/RT accounts for approximately 1-2% of all pediatric central nervous system (CNS) tumors and roughly 10-20% of tumors in patients less than three years of age. While AT/RT has been encountered in the adult population, the vast majority of the cases reported occur in the supratentorial space. In the existing literature, only three adult cases have ever been reported that arise from the cerebellum.

Case Description: A 38-year-old female presented with six months of worsening nausea, emesis, vertigo, diplopia, and coordination difficulty. Magnetic resonance imaging revealed a T1 avidly contrast enhancing mass, composed of both cystic and solid areas, extending from the cerebellum into the fourth ventricle. Following a gross total resection, surgical pathology was

consistent with AT/RT, with tumor cell loss of integrase interactor-1 (INI-1) observed via immunohistochemical staining.

Conclusions: This case represents just the fourth ever reported case of AT/RT arising from the cerebellum in an adult and the oldest reported age to date of a cerebellar AT/RT occurring in a female. Due to the paucity of reported adult AT/RT cases, little is known about adults with AT/RT. Further reports will function to improve the general understanding of AT/RT in the adult population.

Introduction

Atypical teratoid/rhabdoid tumor (AT/RT) is a rare and highly malignant embryonal neoplasm of the central nervous system (CNS). AT/RT is characterized histologically by the presence of rhabdoid cells among primitive neuroectodermal, epithelial, and mesenchymal cell populations.[15,16] Epidemiologic study demonstrates that AT/RT likely accounts for only 1.6% of all pediatric CNS tumors.[13] AT/RT is very rarely encountered in the adult patient population (≥18 years of age). Of the AT/RT cases reported in adults, the large majority of them occur supratentorially.[21] Manifestation of AT/RT in the cerebellum of an adult is extremely rare, with only three cases reported in the scientific literature to date.[8,10,14] Here we describe a rare additional instance of AT/RT in an adult arising from the cerebellum. Specifically, this case identifies the fourth ever cerebellar AT/RT encountered in an adult and the oldest adult female to date, 38 years of age, presenting with AT/RT in the cerebellum.

Case Illustration

Patient Information

A 38-year-old female presented to the neurosurgery clinic with six months of worsening nausea and emesis (Table 1). She also reported vertigo, diplopia, decreased sense of taste, unintentional weight loss, and coordination difficulty. She denied headache, seizure-like activity, dysarthria, dysphagia, fevers, and chills. Outside of a maternal grandfather with an unspecified skin cancer, the patient denied having a personal or family history of cancer, specifically brain tumors. She denied a history of tobacco and illicit drug use.

Clinical Findings

- Blood pressure 115/79 millimeters of mercury, pulse 83 beats per minute and regular, respiratory rate 18 breaths per minute, body mass index 21.60 kilograms per meter squared
- Alert and oriented to person, place, time, and situation with fluent speech, good memory, and recall
- Pupils equal, round, and reactive to light bilaterally
- Extraocular motions intact but with evidence of nystagmus when looking superiorly, leftward, and rightward
- Visual fields intact to confrontation, face is symmetric, tongue protrudes midline, uvula and palate elevate midline, face sensation intact bilaterally to light touch, good sternocleidomastoid and trapezius strength
- Strength five out of five in the bilateral upper and lower limbs
- No pronator drift
- Grossly intact sensation bilaterally to light touch in the upper and lower limbs
- Romberg test is midline
- No gross evidence of dysdiadochokinesis, dysmetria, or tremor

- Patient unable to perform tandem gait

Timeline

(Table 1)

Diagnostic Assessment

Initial magnetic resonance imaging (MRI)-scan findings demonstrated a 3.1 x 3.3 x 3.6 cm contrast-enhancing mass composed of both cystic and solid areas extending from the cerebellum into the fourth ventricle (Figure 1). There was no evidence of resulting hydrocephalus. The remainder of the ventricular system, brain parenchyma, and intracranial structures appeared unremarkable.

Therapeutic Intervention

The patient was taken to the operating room and underwent a suboccipital craniotomy and C1 laminectomy with a telovelar tonsillar approach for resection of the mass. Intraoperative findings revealed a soft-consistency mass that appeared to originate from the cerebellar tonsils. The mass protruded into the fourth ventricle and abutted the caudal aspects of the brainstem. A gross total resection was achieved. The resected mass was sent as two gross specimens to the pathology laboratory. Further histologic examination of the tumor revealed a densely-cellular neoplasm of diverse morphological features, including glandular patterns and sheet-like arrangements as well as a fibrotic and myxoid background. The specimens displayed mostly a vesicular chromatin pattern with prominent nucleoli, apoptotic figures, and abundant mitoses. There were areas of cells with moderate pink cytoplasm in glandular patterns and areas of cells with moderate pink cytoplasm in sheet-like arrangements. Vascular endothelial proliferation and necrosis was not seen. The tumor stained positive with glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA), and synaptophysin. Tumor cells showed loss of integrase

interactor-1 (INI-1) staining (Figure 2). These histologic features were consistent with a diagnosis of atypical teratoid/rhabdoid tumor, World Health Organization (WHO) grade IV. The patient was started on a postoperative dexamethasone taper.

Follow-Up and Outcomes

Postoperative day one MRI-scan demonstrated expected postoperative changes following posterior fossa craniotomy without evidence of residual tumor (Figure 3). The patient progressed well postoperatively and was discharged on the sixth postoperative day to an acute rehab facility (Figure 4). She was referred to radiation oncology and medical oncology for further evaluation and management. On three weeks follow-up visit, the patient was still experiencing some vertigo and diplopia but was overall doing well. Total spine MRI three weeks postoperatively was unremarkable for metastatic disease. The patient's case was presented at a multidisciplinary tumor board where it was recommended that the patient undergo genetic counseling, computed tomography (CT)-scan of the abdomen and pelvis assessing for renal rhabdoid tumors, and spinal tap in order to rule out metastatic disease in the spine. Spinal tap at four weeks postoperatively was unremarkable for evidence of metastatic disease. CT-scan of the abdomen and pelvis at five weeks postoperatively was unremarkable for evidence of renal rhabdoid tumors. Follow-up MRI scan at five weeks postoperatively demonstrated small areas of nodularity and conspicuous enhancement in the resection cavity, concerning for disease recurrence, though postoperative changes could not be ruled out. Postoperative adjuvant radiation therapy began 6 weeks postoperatively. The patient received a total of 36 Gray (Gy) in 20 fractions to the craniospinal axis and an additional sequential boost of 19.8 Gy in 11 fractions to the posterior fossa. She tolerated the adjuvant radiotherapy regimen well and without complication. MRI-scan at three and a half months postoperatively showed decreased nodular enhancement along the margins of

the resection cavity, consistent with favorable radiotherapy treatment response. Follow-up cervical spine MRI at three and a half months postoperatively continued to remain unremarkable for evidence of metastasis. An adjuvant chemotherapy regimen began five months postoperatively, consisting of cisplatin, etoposide, and cyclophosphamide administered over six cycles. The patient developed a leukopenia and neutropenia and thus received a seven-day course of filgrastim. Following her fifth cycle, she also developed a peripheral neuropathy in her hands and feet, as well as muffled hearing, leading to cessation of cisplatin therapy. She otherwise tolerated the six cycles of adjuvant chemotherapy regimen well. Eight-month postoperative MRI scan demonstrated continued decrease in the areas of nodular enhancement along the margins of the resection cavity, with therefore no evidence of disease recurrence. The patient continues to do well with no additional reported complications at 12 months.

Discussion

AT/RTs are incredibly rare and aggressive embryonal malignancies of the CNS. They are unique in that they are characterized by the hallmark of rhabdoid cells together with the presence of epithelial, neuroepithelial, and mesenchymal cells.[14] Having only been first described in 1987, AT/RT is a relatively new diagnostic entity.[9] Since it was first described, the overwhelming majority of reported cases have been in the pediatric patient population. To the best of our knowledge, less than 60 cases of AT/RT have been described in adults.[21] The majority of reported adult AT/RT cases occur in the supratentorial compartment and rarely present infratentorially.[16] Prior to this report, there have only been three reported descriptions of AT/RT manifesting from the adult cerebellum.[8,10,14] This report serves to describe the fourth ever case of

AT/RT in the adult cerebellum and the oldest female diagnosed with AT/RT arising from the cerebellum to date.

The three previously described cases of AT/RT occurring in the cerebellum, along with the current case, are described in Table 2. The mean age at diagnosis of the four adult cerebellar AT/RT cases is 34 years of age. There have been two reported female cases and two male cases. Of the four cases, only one presented with evidence of leptomeningeal disease at time of diagnosis. None of the four cases demonstrated evidence of extracranial disease. Two of the four cases received gross total resection, one received partial resection, and the other case did not specify if gross total or partial resection was achieved. All four patients received some form of radiotherapy. Two of the four patients received whole neuraxis radiation in addition to local radiation, one patient received only local radiation, and the other case did not specify whether whole and/or local radiotherapy was used. Three of the four patients received adjuvant chemotherapy. Two of the three patients received cisplatin, etoposide, and an alkylating agent (ifosfamide or cyclophosphamide). One of the patients also received intrathecal methotrexate. The one patient that did not initially receive adjuvant chemotherapy received temozolamide following her second episode of disease recurrence. Of the four patients, only one had reportedly experienced tumor recurrence. That same patient went on to succumb from her disease. The remaining three patients were still alive at time of case report publication, with the longest survival being 24 months following initial time of diagnosis. Of the three alive patients, two patients received surgical resection, whole neuraxis radiation plus local radiation, and an adjuvant chemotherapy regimen consisting of etoposide, cisplatin, and an alkylating agent. While this is an extremely small sample size, it could possibly assist in paving the way for future studies attempting to analyze the effects of a similar treatment regimen in adults with AT/RT.

It is well reported in the scientific literature that AT/RT is often misdiagnosed in adults. AT/RT's frequent misdiagnosis is largely due to its low prevalence in the adult patient population and histologic overlap with other more prevalent neoplastic processes with rhabdoid features, such as high-grade gliomas, rhabdoid meningiomas, and some metastatic tumors. That being said, the most definitive way to properly diagnose AT/RT is via immunohistochemistry demonstrating inactivation or deletion of SMARCB1/INI-1 or through fluorescence in situ hybridization.[14] Our patient's tissue specimen demonstrated loss of INI-1 in tumor cells, therefore together with its histologic findings yielding a diagnosis of AT/RT.

Review of the literature reveals some additional considerations to be mindful of when it comes to further diagnostic evaluation of AT/RT in the adult patient population. Studies show that cerebrospinal fluid (CSF) involvement via leptomeningeal dissemination can be seen in as high as 30% of AT/RT cases.[3,4] Therefore, in order to assess for leptomeningeal spread, our patient underwent an MRI of the entire neuroaxis and a spinal tap to analyze CSF cytology, both of which were unrevealing. Studies also show that patients presenting with AT/RT may also have rhabdoid predisposition syndrome, causing them to present with multiple rhabdoid tumors elsewhere in the body, in particular the kidney.[19] Additional imaging is typically obtained to assess for kidney involvement.[4] Our patient underwent a CT-scan of the abdomen and pelvis to rule out the presence of additional rhabdoid tumor in the kidney.

When it comes to standard-of-care treatment in adults with AT/RT, there is very little data, largely in part due to the rarity of the disease process. Much of the current treatment approach in adult patients with AT/RT stems from the treatment data of pediatric patient cohorts.[17] Of the limited adult AT/RT treatment data reported in the literature, studies do suggest that gross total resection of the tumor followed by adjuvant chemoradiation may yield a better

outcome in AT/RT adult patients.[1,2,7,18,20] In contrast, one systematic review of AT/RT in adults found there to be no difference in survival between those that received gross total resection versus incomplete resection.[6] However, the same systematic review found patients that received both radiotherapy and chemotherapy to have increased survival.[6] Additionally, another report suggests that as many as 50% of AT/RT tumors may be responsive to chemotherapy.[2] While adults are better suited than pediatric patients in tolerating the proposed strong regimens of chemotherapy used in treating AT/RT, such regimens are still extremely toxic and life-threatening none the less. Therefore, adult AT/RT patients are traditionally administered medulloblastoma-like chemotherapy regimens. Brandes et al. reports a regimen consisting of etoposide, cisplatin, and cyclophosphamide that has shown stabilization of disease in 2 patients and complete remission in one patient.[5] Our patient received a gross total surgical resection. She received adjuvant radiotherapy in the form of 36 Gy to the craniospinal axis and a local sequential boost of 19.8 Gy to the posterior fossa beginning 6 weeks postoperatively. Adjuvant radiotherapy was followed by six cycles of cyclophosphamide, cisplatin, and etoposide beginning 5 months postoperatively.

While the prognosis of AT/RT in children is dismal and almost uniformly fatal within one year, the prognosis for adults seems to be better. There is evidence in the literature of some patients surviving beyond 17 years from the time of diagnosis.[11] However, the overall prognosis of adults with AT/RT still remains poor. Several studies demonstrate an average overall survival of only 20 months in adult patients.[12] Our patient continues to do well 12 months postoperatively with no evidence of disease recurrence. In conclusion, largely in part due to the limited number of reported adult AT/RT cases and their wide dispersion in the scientific literature, the overall prognosis of adults with AT/RT remains unclear. Given the poor understanding of prognosis in

adult patients with AT/RT, it is vital that more cases and further research be reported in order to better highlight the prevalence and prognosis in the adult population.

Conclusion

AT/RT is a rare and very aggressive malignant entity of the CNS. Here we present an extremely rare case of a cerebellar AT/RT in an adult female presenting with months of nausea, vomiting, and vertiginous symptoms. The diagnosis was confirmed with absent immunohistochemical expression of INI-1 in tumor cells, and our patient received a multidisciplinary approach to treatment. She continues to do well 12 months postoperatively.

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Figure Legends

Figure 1. Preoperative contrasted T1 axial MRI (panel A) and coronal MRI (panel B) demonstrating an avidly contrast enhancing mass measuring 3.1 x 3.3 x 3.6 cm, composed of both cystic and solid areas, and extending from the cerebellum into the fourth ventricle.

Figure 2. Histologic specimen with features consistent with atypical teratoid/rhabdoid tumor. Hematoxylin & eosin-stained section at 4x (panel A) and 40x (panel B) revealing a densely-cellular neoplasm with glandular patterns and sheet-like arrangements, as well as a fibrotic and myxoid background. Evidence of a vesicular chromatin pattern with prominent nucleoli, apoptotic figures, and abundant mitoses. Glial fibrillary acidic protein (GFAP) immunochemistry demonstrating diffuse positivity (panel C). Synaptophysin immunohistochemistry demonstrating positivity (panel D). Epithelial membrane antigen (EMA) immunohistochemistry with diffuse positivity (panel E). Integrase interactor-1 (INI-1) staining immunohistochemistry demonstrating INI-1 loss in the tumor cells (panel F).

Figure 3. Postoperative contrasted T1 axial MRI (panel A) and coronal MRI (panel B) demonstrate expected postoperative changes without gross evidence of residual tumor.

Figure 4. Outline of postoperative course.

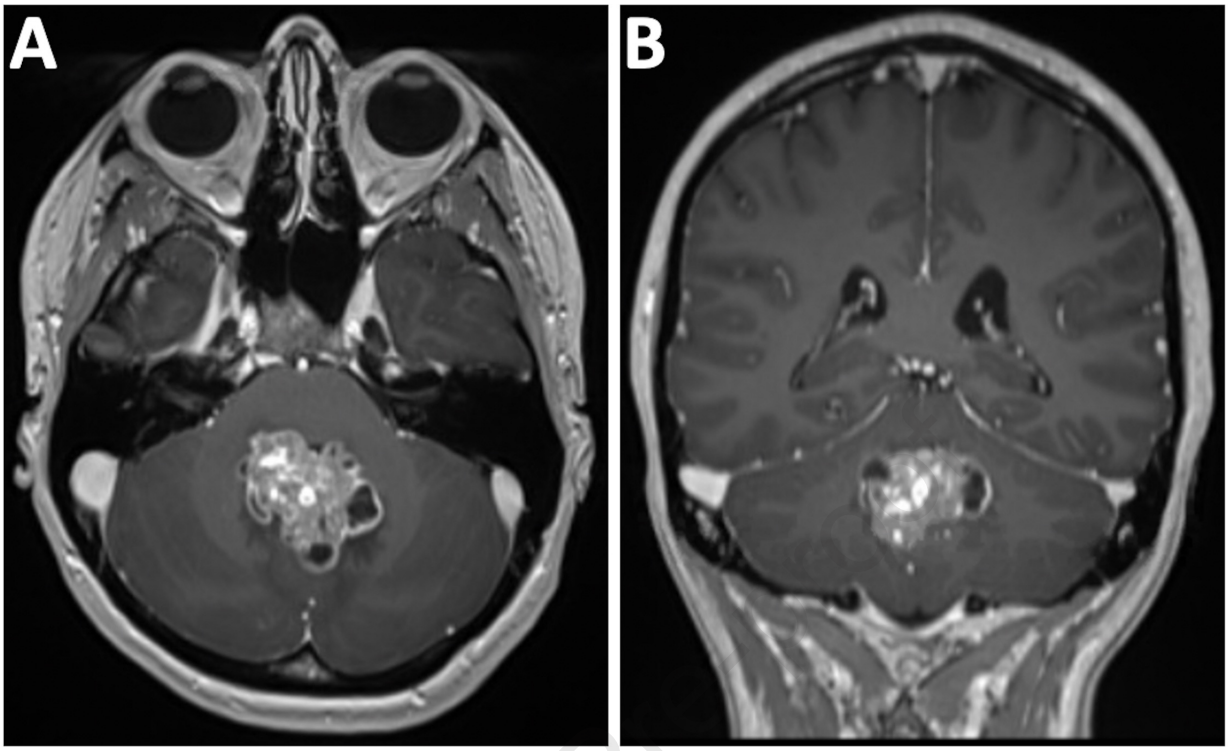
Table 1. Medical history timeline	
December 2018	Symptoms of nausea, vomiting, and vertigo begin
March 2019	Otolaryngology clinic visit with referral for vestibulotherapy
May 7th, 2019	Brain MRI obtained demonstrating mass arising from cerebellum and extending into the fourth ventricle
May 10th, 2019	Neurosurgery clinic visit
May 30th, 2019	Suboccipital craniotomy for mass removal
Postoperative Day 1	Brain MRI obtained demonstrating gross total resection
Postoperative Day 6	Discharged from hospital
June 18th, 2019	Total spine MRI unremarkable for metastatic disease
June 19th, 2019	Postoperative visit – symptomatic improvement and wound healing well
Late June 2019	Case presented at multidisciplinary tumor board
June 28th, 2019	Lumbar puncture obtained and unremarkable for metastasis
July 3rd, 2019	CT abdomen pelvis obtained and unremarkable for renal rhabdoid tumors Follow-up brain MRI obtained and concerning for disease recurrence, though postoperative changes unable to be excluded
Early July 2019	Postoperative adjuvant radiotherapy begins
Late August 2019	Postoperative adjuvant radiotherapy complete
September 17th, 2019	Follow-up brain MRI obtained and consistent with favorable radiotherapy treatment response Follow-up cervical spine MRI obtained and unremarkable for metastasis
Early October 2019	Six cycles of postoperative adjuvant chemotherapy begin
February 2nd, 2020	Follow-up brain MRI obtained and without evidence of recurrence
Early March 2020	Six cycles of postoperative adjuvant chemotherapy complete

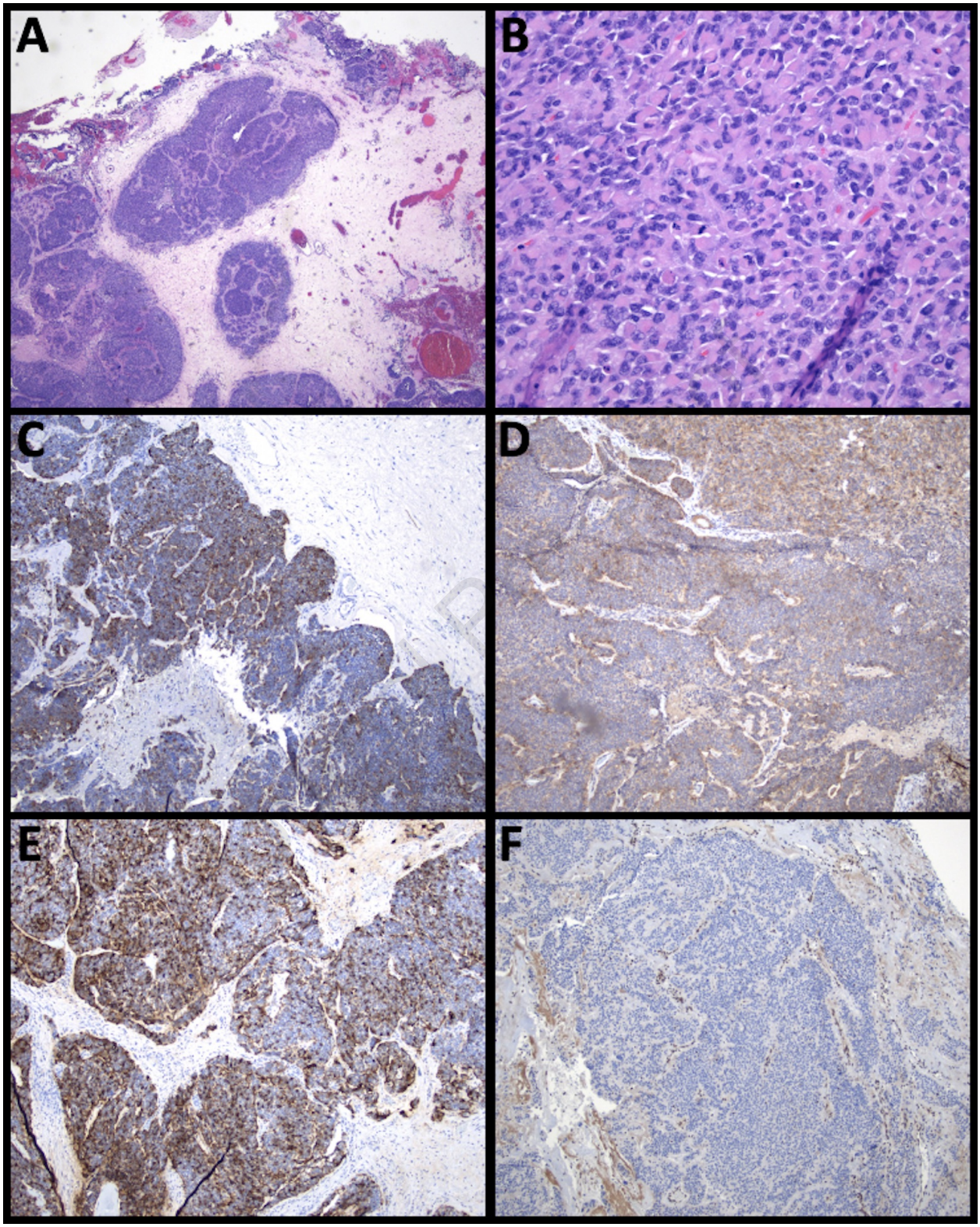
CT, computed tomography; MRI, magnetic resonance imaging

Table 2. Literature review describing four adult cases of atypical teratoid/rhabdoid tumor arising from the cerebellum

Case	Age at Diagnosis (years)	Gender	Leptomeningeal Disease	Extracranial Disease	Resection	Radiotherapy	Chemotherapy	Recurrence	Survival after Diagnosis (months)
Lutterbach et al.[10]	30	Female	No	No	Gross total	Local, 54 Gy	No	Distant (7 mo) – radiosurgery Local and distant (9 mo) – temozolamide	11 (deceased)
Kawaguchi et al.[8]	22	Male	Brain stem and entire spinal cord	-	Partial	Whole neuraxis, 30 Gy Local, 30 Gy	Ifosfamide, cisplatin, etoposide IT methotrexate	No	24 (alive)
Raisanen et al.[14]	45	Male	No	-	Yes	Yes	Yes	-	15 (alive)
Present case	38	Female	No	No	Gross total	Whole neuraxis, 36 Gy Local, 19.8 Gy	Cyclophosphamide, cisplatin, etoposide	No	11 (alive)

Gy, gray; IT, intrathecal; Mo, month

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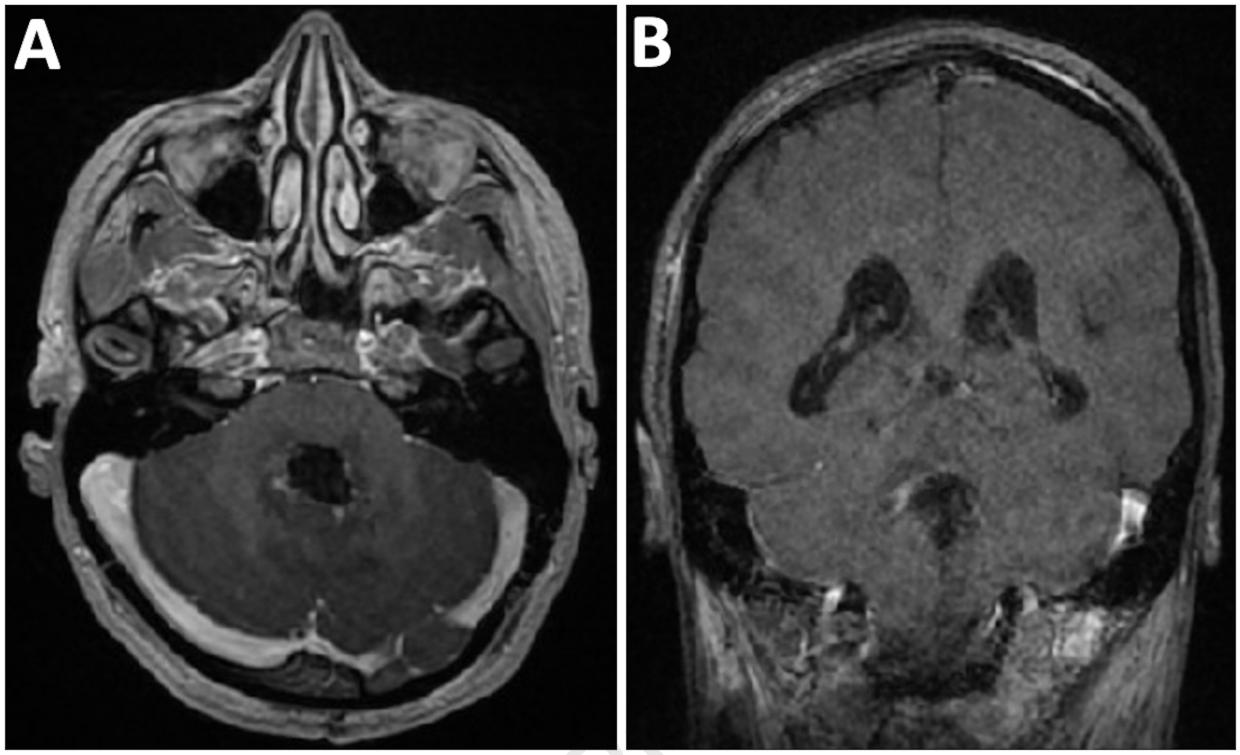

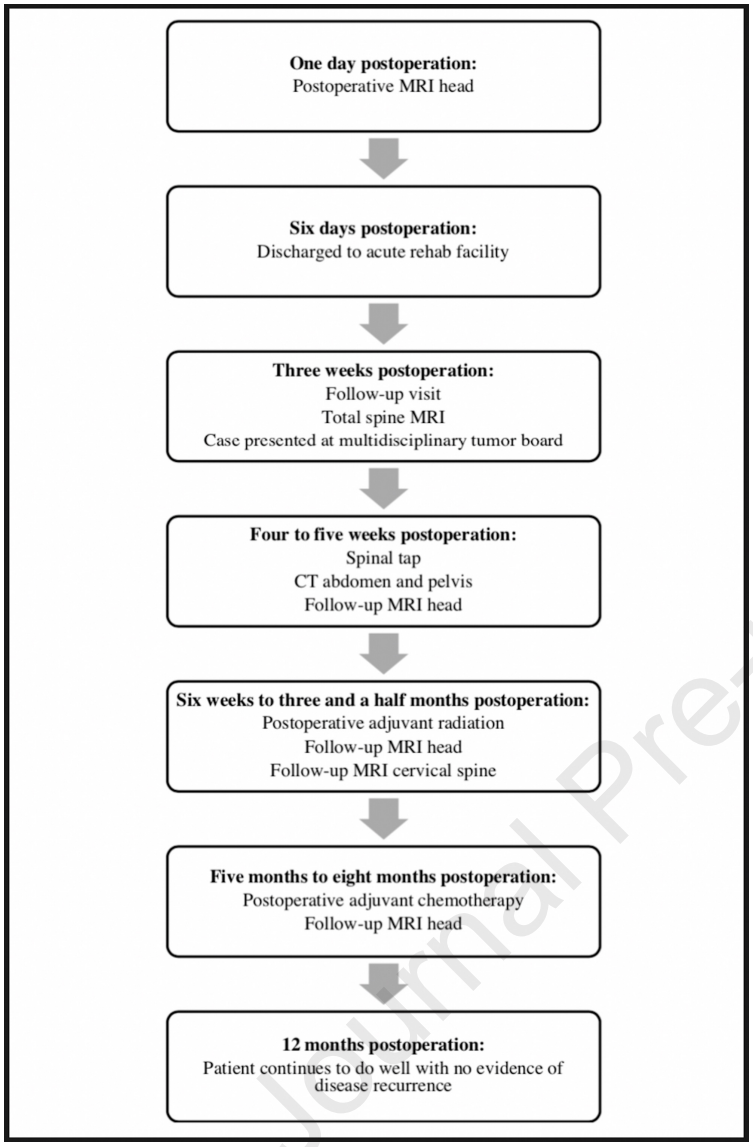

Abbreviations list: AT/RT – atypical teratoid/rhabdoid tumor; CNS – central nervous system; CSF – cerebrospinal fluid; CT – computed tomography; EMA – epithelial membrane antigen; GFAP – Glial fibrillary acidic protein; Gy – gray; INI-1 – integrase interactor-1; MRI – magnetic resonance imaging; SMARCB1 – SWI/SNF-related matrix-associated actin-dependent regulator of chromatin subfamily B member 1; WHO – World Health Organization

Declaration of interests

☒ The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

☐The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: