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Proposal of a New Therapeutic Approach for Adult Atypical Teratoid/Rhabdoid Tumor Based on Literature Review and Case Report

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Abstract

Rhabdoid tumors are aggressive soft tissue sarcomas predominantly affecting children under three years of age. While rare, cases of Atypical Teratoid/Rhabdoid Tumors (AT/RT) have been reported in adults, presenting unique challenges for treatment. This article presents a new therapeutic approach for adult AT/RT based on a case report and literature review. The case involves an adult male diagnosed with AT/RT who achieved a ten-year survival through a comprehensive treatment regimen. The approach includes surgical resection, intensive chemotherapy, and radiation therapy. The study highlights the importance of adopting an intensive chemotherapy protocol, typically used in pediatric cases, as a promising strategy for adult AT/RT management. This innovative proposal underscores the need for further research and therapeutic advancements to improve outcomes in this rare disease.

Keywords

Adult, Atypical Teratoid/Rhabdoid Tumor, Brain, Childhood Tumor, Survival

1. Background

Rhabdoid tumors are very aggressive soft tissue sarcomas with an extremely poor prognosis, predominantly affecting children (Siegfried et al., 2016), in kidneys, liver and other soft tissues of the body (Arrazola et al., 2000). When these lesions are localized on the Central Nervous System (CNS), they are called Atypical Teratoid/Rhabdoid Tumor (AT/RT). This tumor affects preferentially patients under 3 years of age, representing about 10% of all CNS tumors in children. Rare cases

have been described in adults since the 1990s (Horn et al., 1992; Cossu et al., 1993; Fischer et al., 1996; Ashraf et al., 1997; Byram, 1999), mainly affecting the cerebral hemispheres and the sellar region (Chan et al., 2018).

The diagnosis of ATRT is based on the presence of rhabdoid cells (Siegfried et al., 2016), but also, according to the 2016 World Health Organization Classification, by the deletion of the INI1 protein (Louis et al., 2016; Voisin et al., 2019) (SMARCB1 gene, on the long arm of chromosome 22) or the BRG1 protein (Louis et al., 2016; Voisin et al., 2019) (SMARCA4 gene, on the short arm of chromosome 19) (Peng et al., 2021). To our understanding of ATRT, the presence of an INI1 mutation is increasingly recognized as an essential criterion for its diagnosis (Peng et al., 2021).

Numerous therapeutic methods have been tried as treatment for ATRT in adults, with disappointing results in the majority of cases (Eap et al., 2010). Our paper reports a proposal for a new oncological management approach based on a literature review and case of an adult male diagnosed with ATRT.

We conducted a literature review using Google Scholar and PubMed/Medline references to identify all cases of primary cerebral rhabdoid tumors in adults published in French and English. Our research, conducted in July 2022, utilized the following keywords: "ATRT" OR "AT/RT" OR "Rhabdoid tumor" OR "Rhabdoid tumour" OR "TTRA" OR "Tumeur rhabdoïde". We applied filters to select only articles categorized as case report, review, systematic review and meta-analysis. We reviewed all titles and abstracts of the identified articles and thoroughly examined the full copies of related articles. Given the significant variations in patients, prognosis, and treatment approaches associated with different locations, our study exclusively focused on hemispheric localizations while excluding other locations such as sellar, pineal, posterior fossa, and medullary lesions. Additionally, cases of secondary ATRT, or primary mixed tumors were excluded (**Figure 1**).

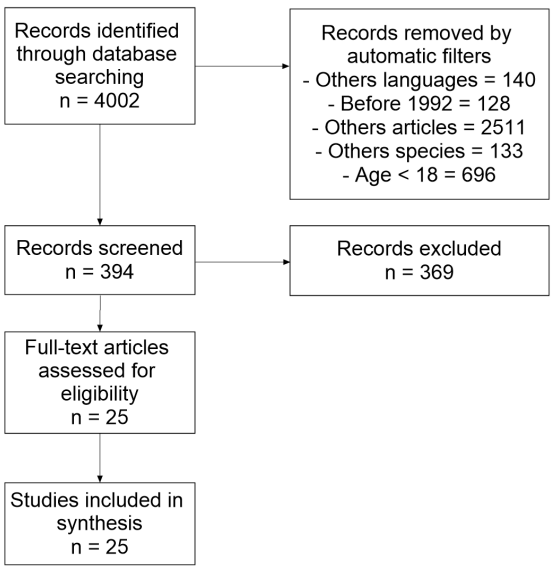

Figure 1. Flowchart of screened, selected and excluded articles.

The clinical, radiological and histological information pertaining to the case we report was collected retrospectively for this study (Table 1). All radiological and histological data, as well as reports from multidisciplinary meetings, were available.

Table 1. Comprehensive literature review of all reported cases of hemispheric ATRT in adult patients.

Author	Publication date	Age	Sex	Symptoms	MRI	INI1	Ki67%	Surgery	Treatment	Survival
Horn et al.	1992	21 yo.	M	Hyperacusis + Hemifacial paresthesias	Left temporal	ND	8.40%	Partial	Radiotherapy	6 years
Cossu et al.	1993	18 yo.	M	Headache	Left frontal	ND	ND	Total	Chemotherapy	18 months
Fisher et al.	1996	32 yo.	M	IH + Asthenia	Left frontal	ND	ND	Biopsy	None	5 months
Ashraf et al.	1997	34 yo.	M	Paresthesias + Dysphasia	Left parietal	ND	ND	Biopsy	Radiotherapy	6 months
Byram	1998	35 yo.	M	Hemifacial paresthesias	Left temporal	ND	ND	Partial	Radiotherapy	5 years
Arrazola et al.	2000	20 yo.	M	Seizure + Paresthesias	Left parietal	ND	ND	Total	Radiotherapy	24 months*
Bruch et al.	2001	34 yo.	F	ND	Parietal	Monosomy 22	ND	ND	ND	6 months
Pimentel et al.	2003	31 yo.	F	Seizure	Right frontal	ND	ND	Partial	Chemotherapy + Radiotherapy	6 months
Erickson et al.	2005	20 yo.	F	IH	Right occipital	Non-deletion	40.00%	Total	Radiotherapy	1 month*
Rezanko et al.	2006	27 yo.	M	Headache	Right frontal	ND	ND	Total	Radiotherapy	4 months
Makuria et al.	2008	23 yo.	M	Headache + Seizure	Left temporal	Deletion	ND	Total	Chemotherapy + Radiotherapy	30 months*
		25 yo.	F	Headache + Bilateral gaze palsy	Right frontal	Deletion	ND	Total x 6	Chemotherapy + Radiotherapy	17 years*
		42 yo.	M	Dysphasia	Right fronto-parietal	Deletion	ND	Partial	Chemotherapy + Radiotherapy	18 months*
		37 yo.	M	Seizures	Right fronto-parietal	Non-deletion	ND	Yes	Radiotherapy	ND
Samaras et al.	2009	18 yo.	M	Headache + Seizure	Right fronto-temporal	Deletion	50.00%	Total	Radiotherapy	4 months
Umredkar et al.	2010	32 yo.	M	Headache + Seizure	Left frontal	Deletion	ND	Total	Chemotherapy + Radiotherapy	6 months*
Han et al.	2011	24 yo.	M	ND	Right temporo-occipital	ND	> 40%	Total	Chemotherapy + Radiotherapy	10 months
		25 yo.	M	ND	Left parieto-occipital	ND	> 40%	Total	Chemotherapy + Radiotherapy	25 months
		32 yo.	M	ND	Right frontal	ND	> 40%	Total	Chemotherapy + Radiotherapy	32 months*
		35 yo.	F	ND	Right frontal	ND	> 40%	Partial	Chemotherapy + Radiotherapy	20 months
		50 yo.	F	ND	Right temporal	ND	> 40%	Total	Chemotherapy + Radiotherapy	13 months
Takahashi et al.	2010	27 yo.	F	Upper limb numbness	Left parietal	Deletion	1% - 50%	Total	Radiotherapy	9 years*
Gorayski et al.	2013	58 yo.	F	Headache	Right parieto-temporal	Deletion	ND	Total	Radiotherapy	20 months
Souki et al.	2014	44 yo.	F	Headache	Right occipital	Deletion	ND	Total	Chemotherapy + Radiotherapy	9 months*
Jin et al.	2015	38 yo.	M	IH	Left occipital	Non-deletion	ND Mib1 = 19.84%	Yes	Radiotherapy	3 months
Wu et al.	2015	18 yo.	M	Emesis	Right temporal	ND	2.00%	Total	None	6 months from diagnosis, 14 days from surgery

Continued

Horiguchi et al.	2016	24 yo.	M	Loss of consciousness + Hemiparesis	Left occipital	Deletion	1.00%	Partial	Chemotherapy + Radiotherapy	5 years*
Zhu et al.	2018	22 yo.	F	Headache + Dizziness	Right temporal	ND	70.00%	Total	Chemotherapy + Radiotherapy	12 months*
	2018	20 yo.	F	IH	Right temporal	ND	60.00%	Total	ND	6 months*
Bodi et al.	2018	22 yo.	F	Upper limb numbness	Left frontal	Deletion	12% - 15%	Total	Chemotherapy + Radiotherapy	ND
Roy et al.	2020	24 yo.	F	HTIC + Ptosis	Right temporal	Deletion	ND Mib1 = 15%	Partial	Chemotherapy + Radiotherapy	11 months*
Pittman et al.	2020	55 yo.	M	IH	Parietal	Deletion	ND	Total	ND	ND
Moujahed et al.	2022	24 yo.	M	Headache + Hemiparesia	Left parietal	Deletion	10.00%	Total	Chemotherapy + Radiotherapy	4 years*
Our case		39 yo.	M	Hemiparesia + Asthenia	Right parietal	Deletion	50% (recurrence = 2%)	Total	Chemotherapy + Radiotherapy	10 years*

*Patients who were alive at the time of writing; IH: Intracranial hypertension.

2. Case Presentation

A right-handed patient with only a history of left wrist and septum surgery and an old episode of prostatitis, presented in 2013, a confusion, bradykinesia and left hemiparesis, at the age of 39. A brain MRI was performed, revealing a right parietal tumor of 40 mm (Figures 2(A)-(D)). After a discussion in a multidisciplinary meeting with neurosurgeons, oncologists and radiotherapists, a macroscopically complete surgery was performed, with complete removal of the lesion. Following surgery, the patient underwent an extension evaluation with MRI of the neuraxis and FDG-PET, which were negative. The search for pathological cells in the cerebrospinal fluid (CSF) was also negative.

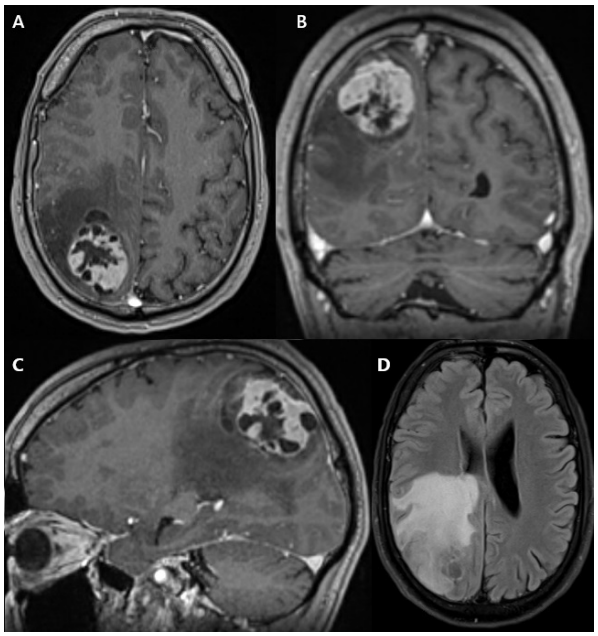

Figure 2. Right parietal tumor with heterogeneous gadolinium enhancement (A)-(C) and perilesional edema visible in FLAIR (D).

The anatomopathological analysis of the tumor revealed a dense proliferation of voluminous undifferentiated cells, with an important glial reaction: numerous enlarged astrocytes, fibrosis, and localized myxoid remodeling, as evidenced by Periodic Acid Schiff (PAS) and Alcian Blue staining. The tumor cells displayed faint cytoplasmic boundaries with eosinophilic cytoplasm, enlarged vesicular nuclei with prominent eosinophilic nucleoli and an estimated mitotic index of 6 mitoses per 10 fields at high magnification (2 mm^2). Additionally, numerous capillaries without endothelial proliferation were observed. Perls staining confirmed the presence of hemosiderin pigment at the periphery of the tumor. Immunohistochemistry revealed intense and diffuse labeling of the tumor cells for Vimentin, while Epithelial Membrane Antigen (EMA) showed heterogeneous cytoplasmic and membrane labeling. Positive staining for Alpha Smooth Muscle Actin (αSMA) and PS100 was observed, whereas Olig2, GFAP, HBM45, Melan A, CD117, neurofilament, Synaptophysin, Chromogranin A, TTF1, CK7, CK20, Estrogen Receptor, Progesterone Receptor, Desmin, CD34 and BAF47 showed negative staining. CD45 and CD3 staining indicated the presence of some T cells. The proliferation index, estimated using Ki67, was 50%, Mib1 was at 40% and p53 at 30%. Furthermore, genetic analysis revealed the presence of the BRAF 600E mutation. Most notably, a mutation in the INI1 gene was identified, further supporting the diagnosis of ATRT. To confirm the histopathological diagnosis of this particularly rare disease, a regional and national review was carried out using the French RENOCLIP network.

We initiated a 40-week course of chemotherapy and radiotherapy (Table 2), with Methotrexate, Vincristin, Doxorubicin, Cyclophosphamide, Cisplatin, Ifosfamide, Etoposide, Carboplatin and Thiotepa. This treatment resulted in transient myelosuppression, alopecia and asthenia.

Table 2. Weekly protocol used in the management of ATRT cases in adults.

Weeks 1 - 2	3 sessions of Methotrexate + Vincristine
Week 3	Rest
Week 4	Doxorubicine + Vincristine
Weeks 5 - 6	Rest
Week 7	Vincristine + cyclophosphamide
Week 8	Rest
Week 9	Cisplatine + Ifosfamide + Etoposide, then cytapheresis
Weeks 10 - 11	Rest
Week 12	Doxorubicine + Vincristine
Weeks 13 - 14	Rest
Weeks 15 - 20	Radiotherapy 30 sessions of 2Gy
Weeks 21 - 23	Rest
Week 24	Cisplatine + Ifosfamide + Etoposide
Weeks 25 - 27	Rest
Weeks 36 - 39	Conditioning with Carboplatin + Thiotepa
Week 40	Intensification through autograft of hematopoietic stem cells

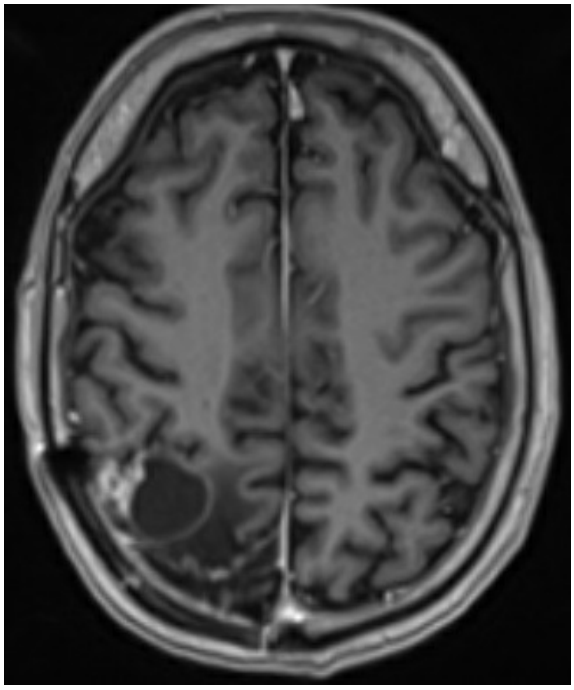

Figure 3. MRI showing cystic right parietal tumor recurrence with peripheral contrast enhancement.

The two-year follow-up MRI showed a small nodular contrast of 9 mm in the lower part of the porencephalic cavity. This lesion was non-progressive on repeated MRIs. There were no associated clinical symptoms. Six years after the surgical procedure, after symptom-free period, focal seizures reappeared on a bimonthly frequency. MRI (**Figure 3**) shows an enlargement of the cystic lesion with a thin shell with contrast enhancement. A new surgical exploration consisted of macroscopically complete excision with *in situ* CARMUSTINE wafer chemotherapy.

The anatomopathology of this second surgery confirmed the recurrence of ATRT with a significant myxoid background. OLIG 2 is negative, IDH1 is negative, and Ki67% is 2%. We decided to perform a new stereotaxic irradiation of the operating bed at a dose of 5 × 7 Gy.

At 10 years from his initial diagnosis, this patient is still alive without any symptoms. The last control MRI showed post-therapy modifications without any evolutionary argument.

3. Discussion

Primary hemispheric atypical/rhabdoid tumor in adults are exceedingly rare, with only 33 cases reported in the literature. These cases have been documented across various cerebral regions: 9 cases in a frontal lobe, 8 cases in a temporal lobe, 6 cases in a parietal lobe, 4 cases in an occipital lobe, and 6 bilobar cases. Notably, only one case showed a superior survival, at the time of writing of the articles: it is a 25-year-old patient at the time of diagnosis, still alive at the time of writing of the article, with a prolonged survival of 17 years, despite some recurrences (Han

et al., 2011).

In contrast, the majority of cases of ATRT in adults are located in the sellar region (Voisin et al., 2019). This subtype of rhabdoid tumors differs markedly from the hemispheric form: it predominantly affects mainly middle-aged women (average age 44 years) and, to date, no pediatric cases have been reported in children (Voisin et al., 2019). In children, ATRT typically affects males under 3 years of age, with a median age of diagnosis around 24 months (Peng et al., 2021). These demographic and clinical differences underscore the distinct biological behavior of ATRT in adults versus pediatric patients.

Due to the scarcity of analytical series and the lack of standardized treatment protocol for adult ATRT, our study adopted the pediatric treatment approach as a reference (Erickson et al., 2005). In pediatric cases, complete surgical resection appears to be the cornerstone of prolonged survival, as evidenced by several studies (Hilden et al., 2004; Sasani et al., 2007; Chi et al., 2009). A prospective multicenter study involving 20 infants (Chi et al., 2009) further confirmed that combining complete resection with systemic and intrathecal chemotherapy followed by irradiation significantly improves progression-free survival, although survival beyond 10 years remains exceptional.

Molecular diagnostics have also evolved, with the detection of INI1 or BRG1 deletion playing a crucial role in confirming ATRT. In addition, we also report 3 articles reporting atypical rhabdoid teratoid tumors without an INI1, or BRG1 deletion (Han et al., 2011; Samaras et al., 2009; Zhu et al., 2018), which are therefore non-conforming to the new WHO 2016 definition. In accordance with these criteria, we identified 13 cases reporting an INI1 deletion (Bodi et al., 2018; Gorayski et al., 2013; Horiguchi et al., 2016; Makuria et al., 2008; Moujahed et al., 2022; Pittman et al., 2020; Roy et al., 2020; Samaras et al., 2009; Souki & Al-Hussaini, 2014; Takahashi et al., 2011; Umredkar et al., 2010). These findings highlight the increasing importance of molecular diagnostics and suggest that complete surgical management combined with adjuvant chemotherapy and radiotherapy may be associated with imporved outcomes.

Among these cases, specific details regarding chemotherapy are available for 4 of them. Souki and Al-Hussaini (2014) and Roy et al. (2020) used an ICE protocol (Ifosfamide, Carboplatin, and Etoposide) with respective survivals of 9 and 11 months, and the patients were still alive at the time of redaction. In 2016, Horiguchi (Horiguchi et al., 2016) documented a 5-year survival using a protocol based on Temozolomide and Bevacizumab while Moujahed achieved a 4-year survival with a combination of Doxorubicin, Cyclophosphamide, Vincristine, Ifosfamide, Cisplatin, Etoposide, and Methotrexate. These disparate outcomes underscore the challenges in establishing a uniform, effective chemotherapy protocol for adult ATRT.

Radiotherapy also plays a critical role in the management of ATRT. Although most data on radiotherapy are derived from pediatric studies, several reports have demonstrated that early initiation, within two months of diagnosis, can reduce

overall mortality (Yang et al., 2020; Baliga et al., 2021). Furthermore, the extent of radiation fields appears to vary according to patient age, reinforcing the need for individualized treatment strategies.

In light of these observations, our study proposes an adaptation of pediatric treatment protocols for the adult population. Our intensive, multimodal strategy comprising complete surgical resection, a 40-week course of multi-agent chemotherapy adapted from pediatric protocols, and targeted radiotherapy which has resulted in significantly improved long-term outcomes. This approach not only addresses the limitations of previous treatment methods but also suggests a promising direction for managing this rare and aggressive tumor.

Future research, ideally involving larger, multicenter studies, is essential to validate these preliminary findings and refine treatment strategies further. Such studies should aim to standardize the therapeutic approach while incorporating detailed molecular and pharmacological analyses to optimize outcomes for adult ATRT patients.

4. Conclusion

Despite the increasing number of reported cases, ATRT remains a very rare tumor in adults, with less than 100 cases reported in all locations combined and with an extremely poor prognosis. Our study reports a particularly exceptional case, with a prolonged survival of more than 10 years, without any sign of recurrence at the last follow-up. From a therapeutic point of view, it is important to note the use of an intensive chemotherapy program, as applied in children, with the use of 9 different molecules. This represents the second case of ATRT with the longest survival, after the case reported by Makuria et al., with a survival of 17 years.

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Availability of Data and Materials

The datasets used and analysed during the current study are not publicly available due to privacy concerns but are available from the corresponding author upon reasonable request.

Authors' Contributions

NG and MR wrote the original draft of the manuscript. JCK reviewed the manuscript and made post-review corrections. CFL, PC and the other authors contributed to the manuscript by providing critical revisions and feedback during the review process. All authors read and approved the final version of the manuscript.

Conflicts of Interest

The authors declare that they have no competing interests.

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List of Abbreviations

αSMA:	Alpha Smooth Muscle Actin
ATRT:	Atypical Teratoid/Rhabdoid Tumor
CNS:	Central Nervous System
CSF:	Cerebro Spinal Fluid
EMA:	Epithelial Membrane Antigen
FDG-PET:	Fluoro Deoxy Glucose Positron Emission Tomography
ICE:	Ifosfamide, Carboplatin, Etoposide
MRI:	Magnetic Resonance Imaging
PAS:	Periodic Acid-Schiff
WHO:	World Health Organization