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Case Report

Unusually Long Survival of an Adult Patient with Atypical Teratoid/Rhabdoid Tumor of the Sellar Region: A Follow-Up Report

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Abstract

Atypical teratoid/rhabdoid tumor (AT/RT) is a rare and very aggressive central nervous system neoplasm that is most often seen in infants and young children. The prognosis remains poor, with a median survival time of <1 year. Here, we report a follow-up on a case of AT/RT that originated in the sellar and suprasellar region in a 42-year-old female patient with unusually long survival.

Keywords: Adult, atypical teratoid/rhabdoid tumor, central nervous system neoplasm, prognosis

Introduction

Atypical teratoid/rhabdoid tumor (AT/RT) is a rare and very aggressive central nervous system (CNS) neoplasm that is most often seen in infants and young children.[1,2] About half of AT/RTs are found in the posterior fossa, but can also occur anywhere in the brain or spinal cord. Its clinical presentation varies with tumor location. Typically, a patient with AT/RT is treated with surgery and craniospinal radiation therapy, which is often followed by systemic chemotherapy. The prognosis for AT/RT is poor, with a median survival time of <1 year.[1-3] However the average survival of 20 months had been reported in adult population in recent systematic reviews.[4,5] We have previously reported a case of AT/RT in a 42-year-old female, originated in the sellar and suprasellar region treated successfully with surgical resection along with radiotherapy and multidrug chemotherapy with short-term follow-up. Here, we present a 6-year follow-up of the same patient.[6]

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Case Report

Our patient is a 42-year-old female presented to the emergency room in October 2013 with a history of severe headache associated with double vision and vomiting for 3 months prior to presentation. She denied galactorrhea, but her menstruation had ceased 2 months earlier before her presentation. She had no weakness or convulsion. She had no significant past medical history, and family history was unremarkable. She also denied using any medication. Physical examination revealed bilateral sixth nerve palsies with the pale optic disc; the rest of the examination was unremarkable. Initial pituitary hormone profiles revealed low level of cortisol 78 nmol/L, and adrenocorticotropic hormone (ACTH) 8.4 pmol/L. Other pituitary hormone levels including; growth hormone (GH) 0.5 mIU/L; follicle-stimulating hormone (FSH) 1.2.1 IU/L; luteinizing hormone (LH) <0.11 IU/L; and FT4 was 16 pmol/l. Preoperative magnetic resonance imaging identified a large sellar mass with the suprasellar extension [Figure 1].

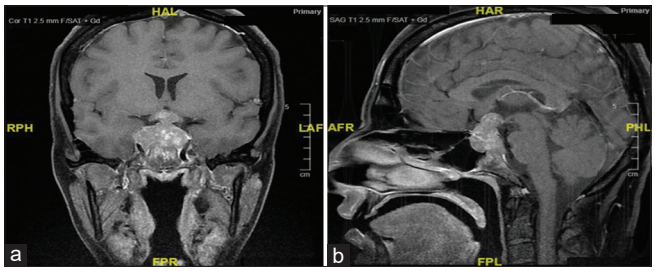

Figure 1: Baseline pituitary magnetic resonance imaging, coronal (a) and sagittal (b) images show evidence of an intrasellar mass with suprasellar extension, compressing and displacing the optic chiasm. Invasion to the cavernous sinus is noted, bilaterally with invasion of the clivus and destruction of the posterior clinoid

Whole-body computed tomography (CT) scans did not reveal any other neoplastic lesions or metastasis. Transsphenoidal approach and tumor resection were performed in November 2013 with subtotal resection of the sellar and suprasellar lesion. Postoperatively, the patient's neurological status remained unchanged. She developed diabetes insipidus that was treated with desmopressin and continued to require cortisol and thyroxin. The patient was discharged without complication. The histopathological study was consistent with the diagnosis of AT/RT [Figure 2]. The patient was treated with concurrent chemotherapy (vincristine) and radiotherapy 60 Gy in thirty fractions followed by six cycles of chemotherapy; ICE protocol (ifosfamide, carboplatin, and etoposide). The patient had a good clinical recovery without any significant toxicity or other complications [Figure 3]; however, bilateral sixth nerve palsy, optic atrophy, and panhypopituitarism persisted, which continued to require full hormonal replacement therapy, including desmopressin, thyroxin, cortisol, and female sex hormone. Seventy-four months postoperatively, the patient continue to have diplopia with no radiological evidence of recurrence since the initiation of therapy [Figure 4].

Discussion

AT/RT is a rare malignant CNS neoplasm usually diagnosed in children who are younger than 3 years of age, and it rarely occurs in adults with approximately only fifty cases reported in the literature so far.[4,5] It can occur anywhere in the CNS, with the majority of these tumors occurring in the posterior cranial fossa. A digital online search using the combination

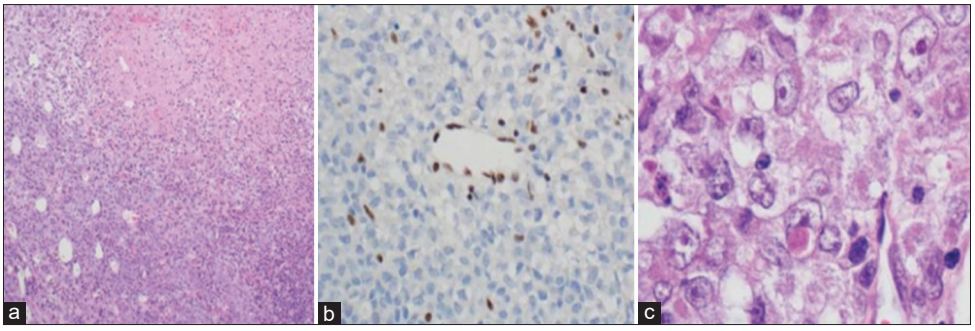

Figure 2: (a) Tumor is composed of sheets of undifferentiated cells with large area of necrosis (H and E stain, ×100 magnifications). (b) INI-1 (BAF47) immunostain shows loss of nuclear staining in the tumor nuclei and retention of nuclear staining in the lymphocytes and endothelial cells (×400 magnification). (c) Tumor cells have oval nuclei and prominent nucleoli with focal eosinophilic globular inclusions (H and E, ×1000 magnification with oil)

of "adult" AND "atypical teratoid/rhabdoid tumor" OR "atypical teratoid/rhabdoid tumor" revealed 101 records in PubMed and 291 in Scopus databases (not mutually exclusive). Its occurrence in the sellar region is particularly rare, with only 31 cases reported in the literature to date [Table 1].[5,7-10]

Sellar AT/RTs have a female predominance, although male predominance with a reported ratio of 3:2–2:1 is notable for AT/RT outside the sellar region.[11] In most adult AT/RT cases, there are no consistent treatment protocols, and any decisions on treatment are extrapolated from the pediatric literature. Treatment typically consists of surgery, chemotherapy, and radiotherapy. However, patients treated with chemotherapy may survived longer than patients who were treated with only surgery and irradiation.[12]

Conclusions

The average survival of a patient with AT/RT is usually <2 years. However, aggressive resection followed by multimodality treatment in our patient

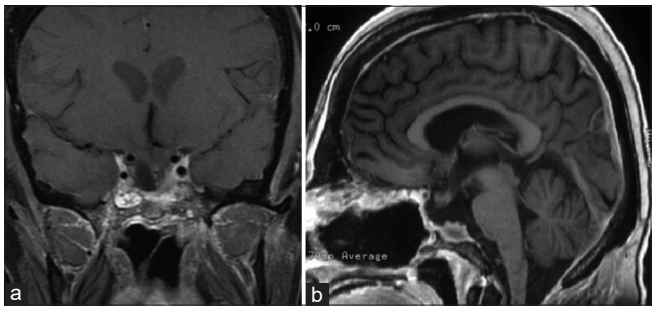

Figure 3: Postoperative pituitary MRI (6 months' post op): Coronal (a) and sagittal (b) images show post debulking of the previously seen large sellar and suprasellar tumor mass lesion, however residual sellar enhancing mass is noted with associated enhancing mass affecting the pituitary stalk and possibly invading the optic chiasm

Table 1: Summary of adult-onset sellar/suprasellar atypical teratoid rhabdoid tumors and patients' age and gender in addition to management and outcome

Case	Author*	Age	Sex	Treatment	Survival
1	Michael A *et al*., 2018	31	Female	Surgery	2 months, died
2	Michael A *et al*., 2018	36	Female	Surgery, radiation, and chemotherapy	22 months, alive
3	Michael A *et al*., 2018	46	Female	Surgery	Postoperative death
4	Michael A *et al*., 2018	47	Female	Surgery, radiation, and chemotherapy	62 months, alive
5	Michael A *et al*., 2018	65	Female	Surgery, radiation, and chemotherapy	23 months, died
6	Kuge *et al*., 2000	32	Female	Surgery, radiation, and chemotherapy	28 months, died
7	Raisanen *et al*., 2005	20	Female	Surgery, radiation, and chemotherapy	28 months, alive
8	Raisanen *et al*., 2005	31	Female	Surgery and radiation	9 months, died
9	Las Heras *et al*., 2010	46	Female	-	-
10	Arita *et al*., 2008	56	Female	Surgery and radiation	23 months, died
11	Schneiderhan *et al*., 2011	61	Female	Surgery	3 months, died
12	Schneiderhan *et al*., 2011	57	Female	Surgery, radiation, and chemotherapy	6 months, alive
13	Moretti *et al*., 2013	60	Female	Surgery, radiation, and chemotherapy	30 months, died
14	Park *et al*., 2014	42	Female	Surgery, radiation, and chemotherapy	27 months, alive
15	Shitara *et al*., 2014	44	Female	Surgery, radiation, and chemotherapy	17 months, died
16	Biswas *et al*., 2015	48	Female	Surgery and chemotherapy	2 months, died
17	Nakata S *et al*., 2017	69	Female	Surgery, radiation, and chemotherapy	38 months, alive
18	AlMalki *et al*.(Present case),2017	42	Female	Surgery, radiation, and chemotherapy	74 months, alive
19	Nakata *et al*., 2017	26	Female	Radiation and chemotherapy	33 months, died
20	Nakata *et al*., 2017	21	Female	Radiation and chemotherapy	35 months, died
21	Johann PD *et al*., 2018	66	Male	-	54 months, alive
22	Johann PD *et al*., 2018	20	Female	Chemotherapy	120 months, died
23	Johann PD *et al*., 2018	48	Female	-	4 months, alive
24	Mehdi *et al*., 2019	55	Female	Surgery	6 weeks, died
25	Asmaro *et al*., 2019	62	Female	Surgery	<2 months, died
26	Lev *et al*., 2014	36	Female	Surgery, radiation, and chemotherapy	29 months, died
27	Nobusawa *et al*., 2016	69	Female	Surgery and chemotherapy	24 months, alive
28	Larran-Escandon *et al*., 2016	43	Female	Surgery and radiation	25 days, died
29	Barresi *et al*., 2018	59	Female	Surgery and radiation	2 months, died
30	Nishikawa *et al*., 2018	42	Female	Surgery	11 months, died
31	Chou *et al*., 2013	43	Female	Surgery and radiation	2 weeks, alive

Full bibliographic information is not included due to space restrictions imposed for case reports

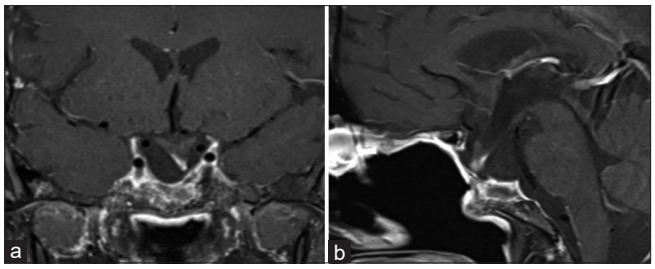

Figure 4: Postoperative pituitary magnetic resonance imaging (April 2019): Coronal (a) and sagittal (b) images show postoperative changes seen at the sellar and suprasellar region with persistent displacement of the optic chiasm inferiorly and the pituitary stalk toward the left side with no definite residual or recurrent masses at the surgical bed

yielded a much longer survival of approximately 74 months with no evidence of recurrence. The progress of knowledge in the management of adult AT/RT with a multidisciplinary approach along with the use of radiotherapy and multidrug chemotherapy might improve the approach to the management of AT/RT and successfully prolonged disease-free survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has/have given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published, and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Authors' contributions

All authors contributed to the care of the patient, drafting of the case report, and revision and approval of its final version.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Compliance with ethical principles

Ethical approval was granted for this report by King Fahad Medical City's Institutional Review Board. The patient provided consent for publication as stated above.

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