



Case Report

Atypical teratoid/rhabdoid tumor presenting with subarachnoid and intraventricular hemorrhage

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Received : 13 February 19
Accepted : 26 April 19
Published : 5 July 19

DOI

10.25259/SNI-59-2019

Quick Response Code:



ABSTRACT

Background: Sellar masses comprise 14–18% of all intracranial tumors. Pituitary adenomas account for 85% of these lesions, while 15% of sellar masses stem from other etiologies. Intratumoral hemorrhage (apoplexy), while not exceptionally common, can be discovered at presentation. While the hemorrhage pattern is typically contained within the tumor, an extension of bleeding beyond the sella has been reported.

Case Description: A 55-year-old female presented with an anterior interhemispheric subarachnoid hemorrhage and extensive intraventricular hemorrhage (IVH). Initially, the IVH was thought to be due to a ruptured aneurysm. After further workup, a hemorrhagic sellar mass was diagnosed. The patient underwent transsphenoidal resection of a pituitary adenoma. The patient returned to the emergency department 6 weeks later with an atypical rapidly expanding sellar mass. After tumor debulking, the pathology revealed an atypical teratoid/rhabdoid tumor (ATRT). Here, we reviewed this and other such cases of sellar ATRT.

Conclusion: The early diagnosis of sellar ATRT with lack of integrase interactor 1 expression and elevated Ki67 proliferation indices can prompt more aggressive attempted gross total resection, chemotherapy, and radiation treatments.

Keywords: Atypical teratoid/rhabdoid tumor, Intraventricular hemorrhage, Sellar mass

INTRODUCTION

Sellar masses occur in 0.1% of the population and represent 14–18% of intracranial tumors. Hemorrhage is an uncommon finding, only occurring in 1.6–2.8% of pituitary adenomas which account for 85% of sellar tumors.^[1,5] Sellar masses with suprasellar extension presenting with intraventricular hemorrhage (IVH) are rare; even more uncommon are atypical teratoid/rhabdoid malignant sellar tumors atypical teratoid/rhabdoid tumor (ATRT) in adults. In fact, there were only 21 cases of adult sellar ATRT tumors in the literature before this case [Table 1]. The diagnosis of a sellar ATRT is a pathological diagnosis (e.g., characteristic loss of integrase interactor 1 (INI1) expression on immunohistochemistry and elevated Ki67 proliferation index).^[3] Here, we present an adult whose hemorrhagic sellar mass resulted in an extensive IVH and subarachnoid hemorrhage (SAH) that was eventually diagnosed to be an ATRT.

CASE REPORT

A 55-year-old female presented with 1 week of headache, blurred vision, and the acute onset of altered mental status. On examination, she opened her eyes to voice, followed commands in all four extremities, and had a slight right hemiparesis. She also exhibited panhypopituitarism and hyponatremia secondary to diabetes insipidus.

Computer tomography and magnetic resonance findings

The brain computer tomography (CT) revealed a suprasellar cistern and anterior interhemispheric SAH with IVH; this required placement of a right ventricular drain [Figure 1]. The brain magnetic resonance imaging (MRI) with and without contrast also demonstrated a moderately enhancing, expansive sellar mass with internal hemorrhage, extending into the suprasellar space. There was uniform enhancement throughout most of the mass, except for its most dorsal aspects which heterogeneously enhanced. There was also an invasion of the right cavernous sinus, protrusion into the sphenoid sinus, and mass effect on the optic chiasm [Figure 2].

Computer tomography and cerebral angiography

The CT angiogram suggested a 3 mm right supraclinoid internal carotid artery aneurysm. Two cerebral angiograms performed 3 days later showed incidental pathology (e.g., 2 mm left middle cerebral artery bifurcation aneurysm; left anterior choroidal artery aneurysm; and a 2 mm right paraophthalmic aneurysm), which was not considered to be contributing to her symptomatology.

Surgery

Transsphenoidal resection of the pituitary mass and evacuation of the sellar hematoma were undertaken. The frozen and permanent pathology was negative for hormonal stains. She was, therefore, originally diagnosed as having a pituitary adenoma with apoplexy and recovered within weeks without a persistent neurological deficit.

Six-week postoperative readmission

Six weeks later, however, she developed worsening of the headaches accompanied by visual loss. The MRI now showed a significant interval increase in the size of the sellar/suprasellar enhancing mass with the invasion of the cavernous sinus and encasement of the internal carotid arteries bilaterally [Figures 3 and 4]. She underwent a right frontal craniotomy for tumor debulking. The mass was highly vascular, and only partial debulking was achievable. Additional stains showed INI1 loss in neoplastic cells, and an elevated Ki67 proliferation index; both were characteristic

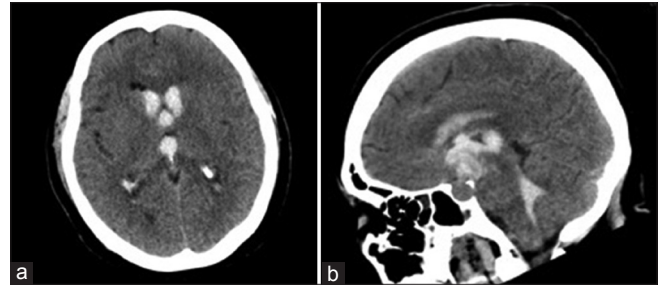


Figure 1: (a) Axial noncontrast head computer tomography showing significant bilateral frontal horn, third and fourth ventricular hemorrhage. (b) Sagittal noncontrast head computer tomography showing third and fourth ventricular hemorrhage. The lateral ventricles are not well visualized here, but some intraventricular hemorrhage is still seen.

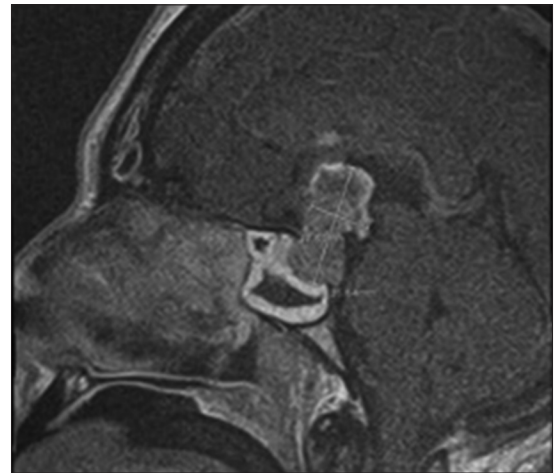


Figure 2: Magnetic resonance imaging brain, sagittal T1-WI with contrast shows a large sellar mass with suprasellar extension. Mass measures 3.5 cm × 1.7 cm.

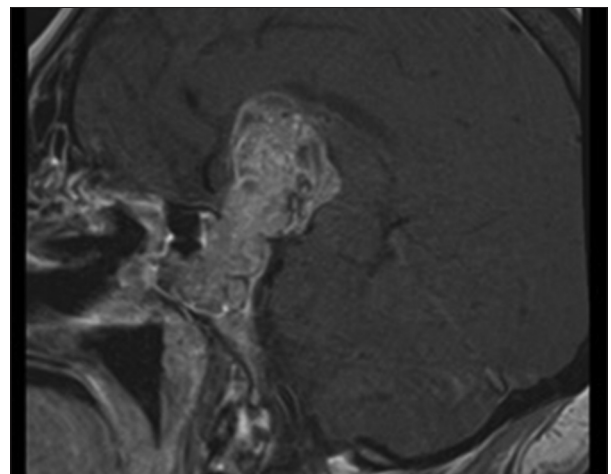


Figure 3: Magnetic resonance imaging sella, sagittal T1-WI with contrast shows a large, enhancing sellar/suprasellar mass extending into the third ventricle. Significantly enlarged compared to prior imaging, no measuring 5.7 cm × 2.9 cm.

for an ATRT (i.e., the World Health Organization Grade IV). Postoperatively, the patient remained intubated and followed commands intermittently for several days. She then, however, suddenly developed a dilated and fixed right pupil along with extensor posturing. When the repeated brain CT showed the

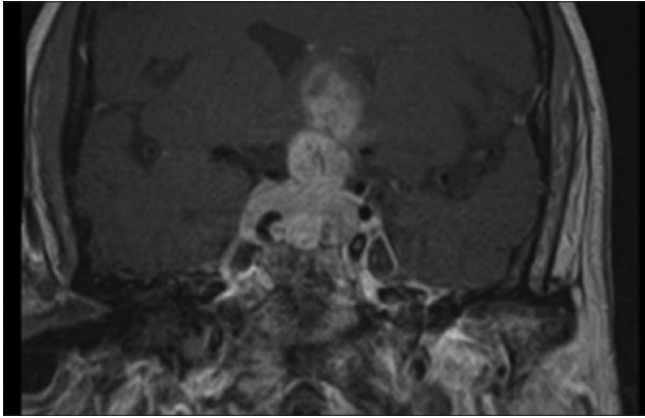


Figure 4: Magnetic resonance imaging sella, coronal T1-WI with contrast shows the mass now extending into the bilateral cavernous sinuses, right greater than left, and encasing the internal carotid arteries.

tumor had re-bleed, the family chose only palliative measures; the patient subsequently expired.

DISCUSSION

ATRT typically is a central nervous system tumor, most commonly found in the posterior fossa.^[3] It is typically diagnosed in infants <2 years of age and is associated with poor clinical outcomes. In adults, only about 50 cases of ATRT have been reported; 21 involved the sella and/or suprasellar cistern.^[9,20] Notably, ATRT in adults has a more favorable clinical course than in children and carries a mean survival time of 23 months.^[4] Common clinical presenting features of sellar ATRTs include strictly females, onset from ages 20 to 69, presentation with persistent blurry vision, headache, ophthalmoplegia, and features of hypopituitarism.^[4,8]

Behavior of sellar atypical teratoid/rhabdoid tumor

Sellar ATRT rapidly recurs after transsphenoidal resection; this also occurred in our patient. Of the previously reported adult sellar/suprasellar ATRT cases, only one presented with SAH and IVH^[4,19] [Table 1].

Table 1: Summary of adult-onset sellar/suprasellar ATRT data in past publications.

Authors (year)	Age (years)	Gender	Symptoms	Initial resection	IVH/SAH	Survival time
Kuge <i>et al.</i> ^[11]	32	Female	Visual disturbance, headache	Subtotal	No	28 months
Raisanen <i>et al.</i> ^[21]	20	Female	Visual disturbance	Total, with recurrence	No	Alive at 28 months
Raisanen <i>et al.</i> ^[21]	31	Female	Not stated	Total	No	9 months
Arita <i>et al.</i> ^[3]	56	Female	diplopia, headache	Subtotal	No	23 months
Las Heras and Pritzker ^[13]	46	Female	headache	Not stated	Not stated	Not stated
Schneiderhan <i>et al.</i> ^[22]	57	Female	Diplopia, headache	Total, with recurrence	No	Alive at 6 months
Schneiderhan <i>et al.</i> ^[22]	61	Female	Diplopia	Subtotal	No	3 months
Moretti <i>et al.</i> ^[15]	60	Female	Headache, diplopia	Total, with recurrence	No	30 months
Chou <i>et al.</i> ^[10]	43	Female	Headache, diplopia	Subtotal	No	Alive at 2 weeks
Park <i>et al.</i> ^[20]	42	Female	Visual disturbance	Subtotal	No	Alive at 27 months
Shitara and Akiyama ^[23]	44	Female	Visual disturbance	Subtotal, with recurrence	No	17 months
Biswas <i>et al.</i> ^[7]	48	Female	Visual disturbance	Total, with recurrence	No	2 months
Lev <i>et al.</i> ^[14]	36	Female	Headache, diplopia	Total, with recurrence	No	29 months
Nobusawa <i>et al.</i> ^[18]	69	Female	Diplopia	Subtotal	No	Alive at 24 months
Larrán-Escandón <i>et al.</i> ^[12]	43	Female	Headaches, diplopia	Subtotal	No	35 days
Barresi <i>et al.</i> ^[5]	59	Female	Headache, diplopia	Subtotal	No	2 months
Almalki <i>et al.</i> ^[2]	36	Female	Headache, diplopia, nausea, vomiting	Subtotal	No	Alive at 37 months
Nakata <i>et al.</i> ^[16]	21	Female	Not stated	Total	Not stated	17 months
Nakata <i>et al.</i> ^[16]	26	Female	Not stated	Total	Not stated	33 months
Nishikawa <i>et al.</i> ^[17]	42	Female	Headache, visual disturbance, vertigo	Total, with recurrence	No	11 months
Asmaro <i>et al.</i> ^[4]	62	Female	Headache, diplopia	Subtotal	Yes	<2 months
Present case (2019)	55	Female	Vision disturbance, headache	Total, with recurrence	Yes	6 weeks

SAH: Subarachnoid hemorrhage, IVH: Intraventricular hemorrhage, ATRT: Atypical teratoid/rhabdoid tumor

Radiographic presentation of sellar atypical teratoid/rhabdoid tumor on magnetic resonance

On MR, sellar ATRT invades the suprasellar space and cavernous sinus. The tumor in our patient showed moderate enhancement throughout most of the mass, including increased dorsal heterogeneous enhancement. Typically, on MR scans, these tumors demonstrate heterogeneous enhancement with peripherally located cysts.

Radiographic presentation of sellar atypical teratoid/rhabdoid tumor on computer tomography

On CT, Biswas *et al.* described ATRT as hyperdense lesions with heterogeneous enhancement, and calcifications (40%).^[7] Alternatively, pituitary adenomas are typically isodense on CT with rare calcifications.^[6]

Pathology of atypical teratoid/rhabdoid tumor

Key distinguishing features of ATRT are an elevated Ki67 proliferation index, and the loss of INI1 protein expression due to partial deletion of chromosome 22 (e.g., which contains the *SMARCB1* gene). The latter two findings were noted in our patient's pathology report.^[7,18,21]

Treatment of atypical teratoid/rhabdoid tumor

At present, there is no standard treatment for sellar ATRT. Although various therapies have been attempted including resection, radiation, and chemotherapy, they have been met with varying success and are largely based on pediatric protocols.^[6,9,20]

CONCLUSION

A 55-year-old female acutely presented with a sellar ATRT characterized by both a subarachnoid and IVH. Although the initial diagnosis was pituitary apoplexy attributed to an adenoma, the secondary bleed led to further histopathological confirmation of an ATRT (e.g., INI1 expression and elevated Ki67 proliferation index).

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Siddiqui M, Thoms D, Samples D, Caron J. Atypical teratoid/rhabdoid tumor presenting with subarachnoid and intraventricular hemorrhage. *Surg Neurol Int* 2019;10:139.