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"Atypical Teratoid/Rhabdoid Tumor of the Sellar Region in an Adult Male: A Case report" (2022-11-19)

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Atypical Teratoid/Rhabdoid Tumor of the Sellar Region in an Adult Male: A Case report

Abstract

Background:
Atypical teratoid/rhabdoid tumor (AT/RT) is a rare, fast growing, aggressive tumor, that is almost exclusively seen in pediatric population. It has a poor prognosis despite aggressive treatment. Nonetheless, there are over 50 cases of AT/RT in adults up to this date. Sellar occurrence is rarely reported, with only 23 cases reported in the literature. All of those cases were female, which raised the question of whether sellar AT/RT is a sex-related disease.

Case presentation:

We report a case of sellar atypical teratoid/rhabdoid tumor (AT/RT) in a 35-year-old male that posed a unique clinical and diagnostic challenge. To the best of our knowledge, this is the third case of male patient with sellar AT/RT in the world.

Conclusion:

Sellar AT/R is extremely rare in an adult male. We report the third case in the literature. Thus disputing the belief that it is a female-exclusive disease. Nonetheless, Sellar AT/RT poses a unique entity that mandates further research and understanding. The rarity and the complexity of managing such cases mandate sharing them whenever possible.

Introduction:

Atypical teratoid/rhabdoid tumor (AT/RT) was first described in 1985. (1) It is a rare, fast growing, aggressive tumor, that is mostly exclusively seen in pediatric population with poor prognosis despite aggressive treatment. (2) It constitutes 1-2% of all pediatric CNS tumors. (3) Nonetheless, there are over 50 cases of AT/RT in adults up to this date. Horn et al were the first to report AT/RT in adult patient in 1992. (4) AT/RT can occur anywhere in CNS. As all CNS tumors, clinical presentation varies with tumor location. Sellar occurrence is rarely reported, with only 23 cases reported in the literature. (5) interestingly, almost all adult patients with sellar AT/RT were female, which raised the question of it being a sex-related disease.

Hereby, We report the third case of sellar atypical teratoid/rhabdoid tumor (AT/RT) in a 35-year-old male that posed a unique clinical and diagnostic challenge.

Case presentation:

A 32 year old male, who is a known case of schizophrenia and obsessive compulsive disorder, brought to our emergency department by his father as he was complaining of headache, high grade fever, and vomiting for 4 days. Also, he had left eye redness and greenish discharge. the patient became unresponsive and dyspneic shortly after presentation, so he was intubated and stabilized. Later, the patient developed generalized tonic clonic seizure. On examination, his Glasgow Coma Scale (GCS) was 7/15, his pupils were equal and reactive. No signs of meningism, clonus, or further abnormal movement were

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Discussion:

AT/RT is prototypically an aggressive tumor of infancy, constituting 10% of CNS tumors in this age group. (6,7) AT/RT is defined by inactivation of SMARCB1, or SMARCA4 in rare cases. (7) Occurrence of AT/RT in adult is extremely rare, with less than 60 cases reported in the literature up to date. (5) Distinguished from pediatric AT/RT, Adult-onset AT/RT is typically supratentorial, with slight predilection for mid-line structures.(8) Moreover, many cases reported long-term survival which may indicate more favorable outcome than pediatric cases. (9) Sellar occurrence is exclusively reported in adult cases. Reported findings suggest that

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Key words:
Atypical teratoid/ rhabdoid tumor, AT/RT, sellar lesion, Inactivation of SMARCB1, SMARCB1

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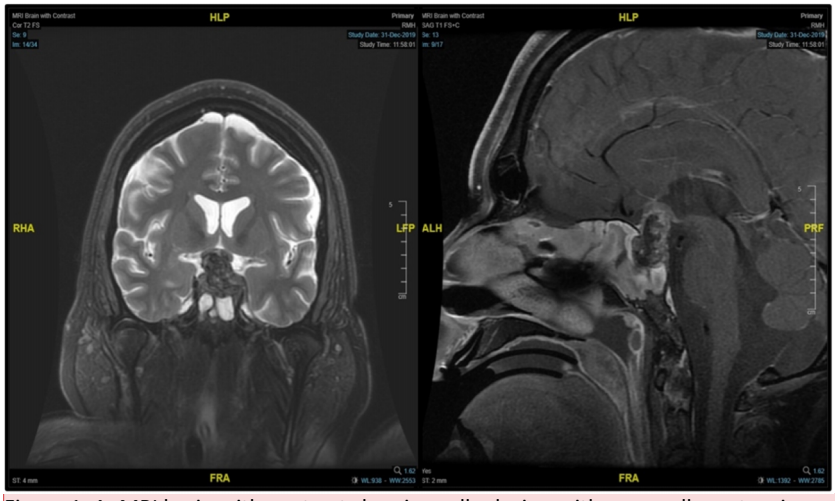

Figure 1: A: MRI brain with contrast showing sellar lesion with suprasellar extension and extension inferiorly to the sphenoid sinus. B: Post gadolinium study showed the lesion to be enhancing in its periphery only with mild heterogeneous enhancement in the center

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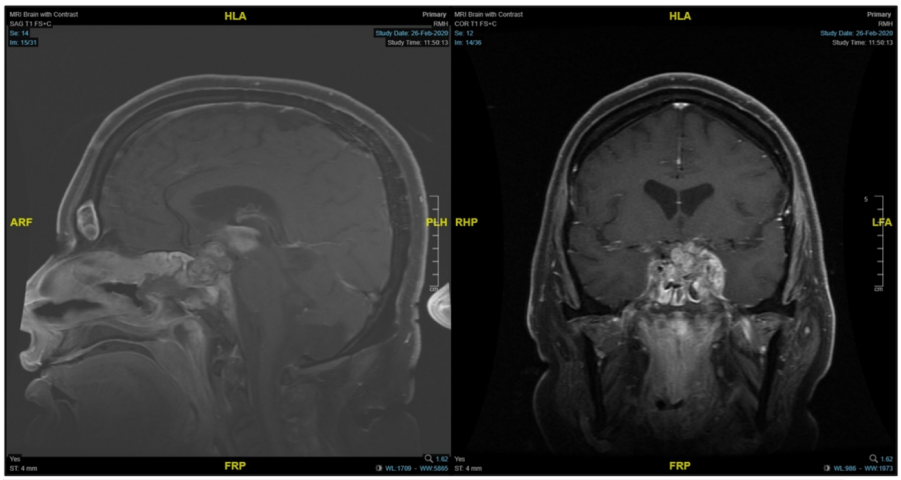

Figure 2: MRI brain with contrast showing Slight interval progression of lesion size with heterogeneous texture/enhancement and ill-defined margins. And development of subacute intraventricular hemorrhage predominantly involving third ventricle.

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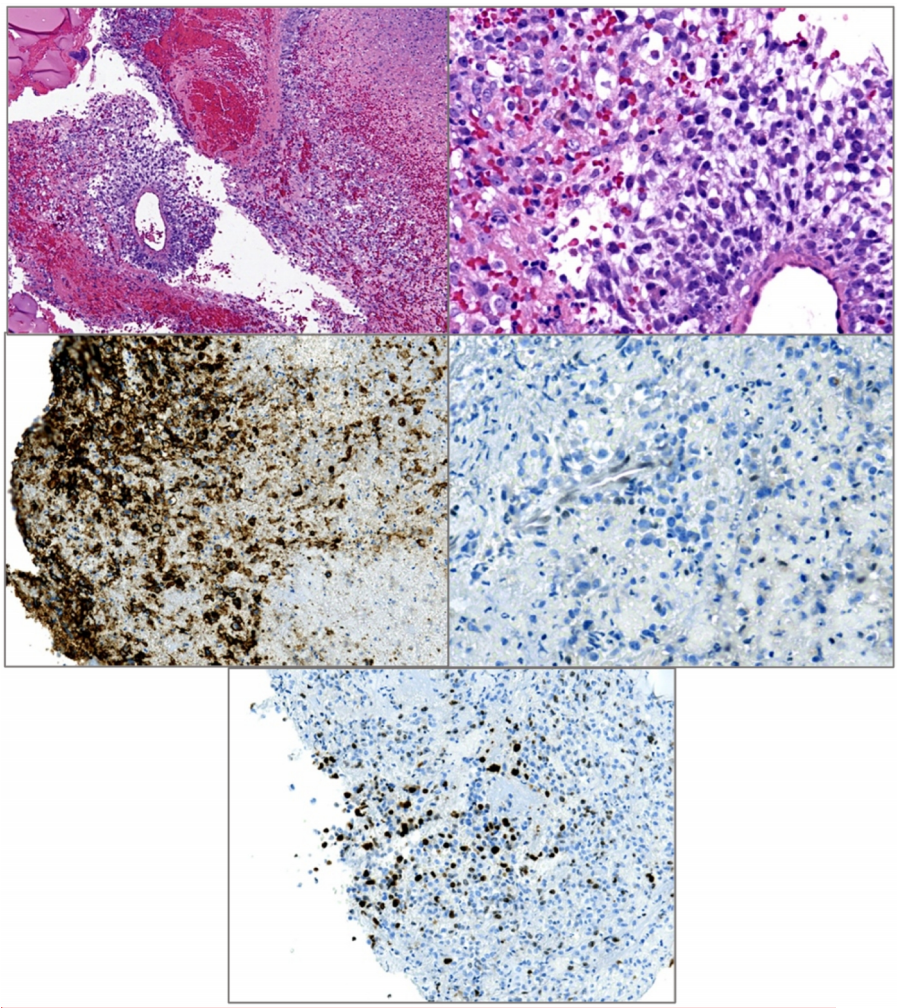

Figure 3: Histopathological examination: A: H&E slide shows hemorrhagic and necrotic neoplasm with focal papillary architecture. B: The tumor is composed of cells with round nuclei, prominent nucleoli and scant cystoplasm. C: Immunohistochemical stain (EMA), positive in the tumor cells. D: Immunohistochemical stain (INI-1), loss of expression in the tumor cells and retention in the internal positive controls encluding endothelila cells. E: (KI67) proliferation index, focally elevated.

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Abstract

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Key words:
Atypical teratoid/ rhabdoid tumor, AT/RT, sellar lesion, Inactivation of SMARCB1, SMARCB1

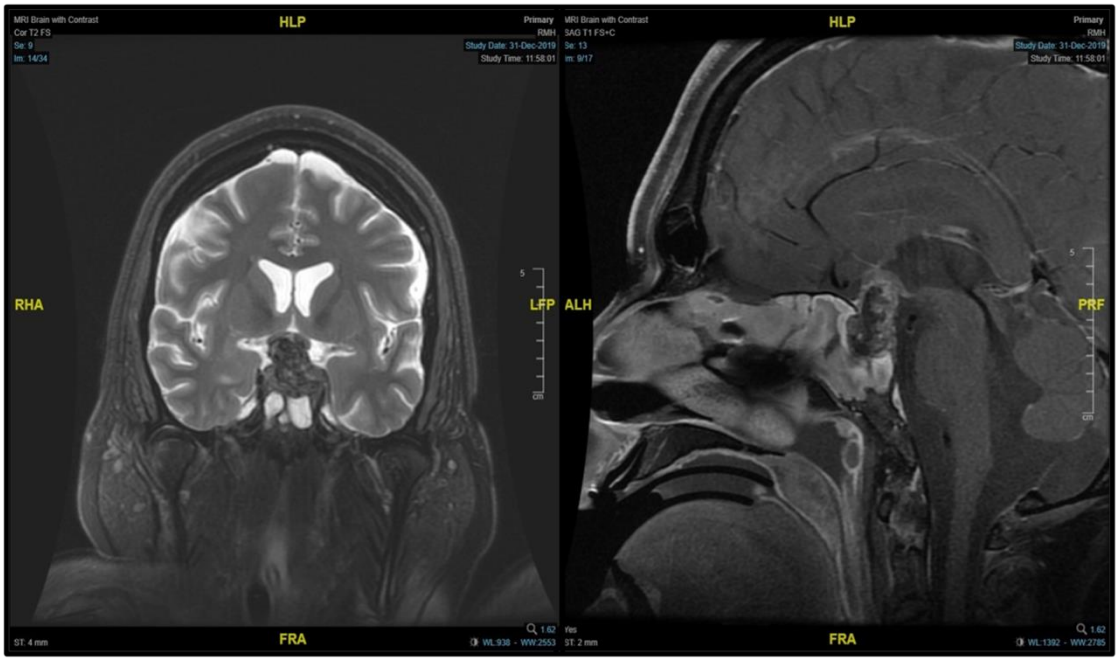

Figure 1: A: MRI brain with contrast showing sellar lesion with suprasellar extension and extension inferiorly to the sphenoid sinus. B: Post gadolinium study showed the lesion to be enhancing in its periphery only with mild heterogeneous enhancement in the center

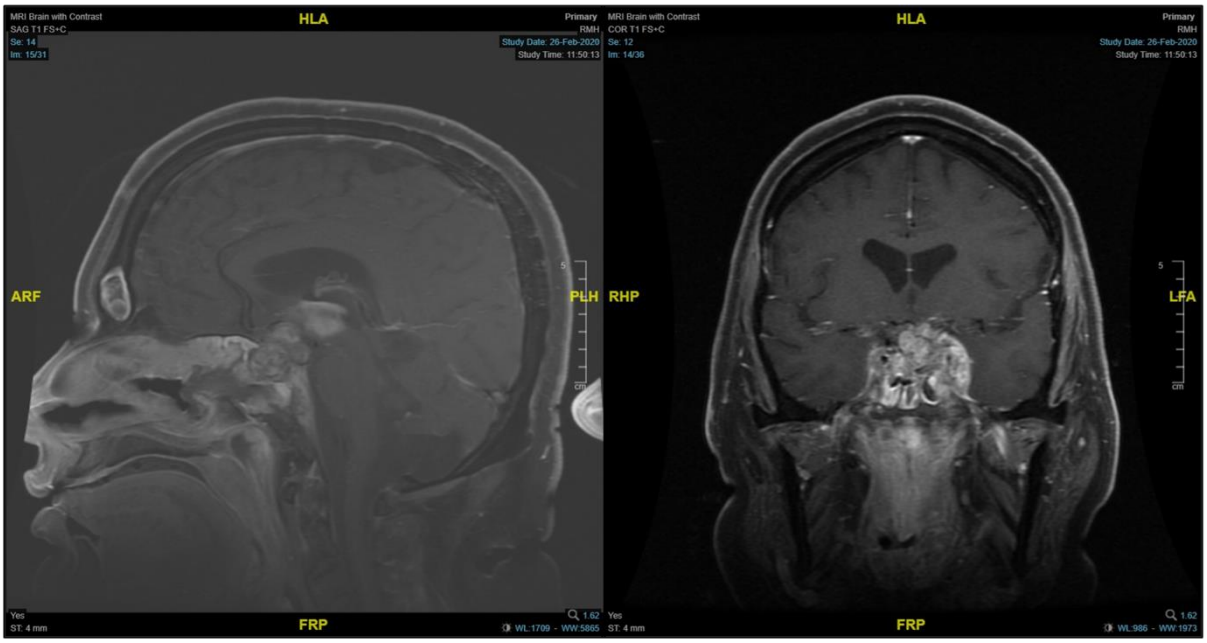

Figure 2: MRI brain with contrast showing Slight interval progression of lesion size with heterogeneous texture/enhancement and ill-defined margins. And development of subacute intraventricular hemorrhage predominantly involving third ventricle.

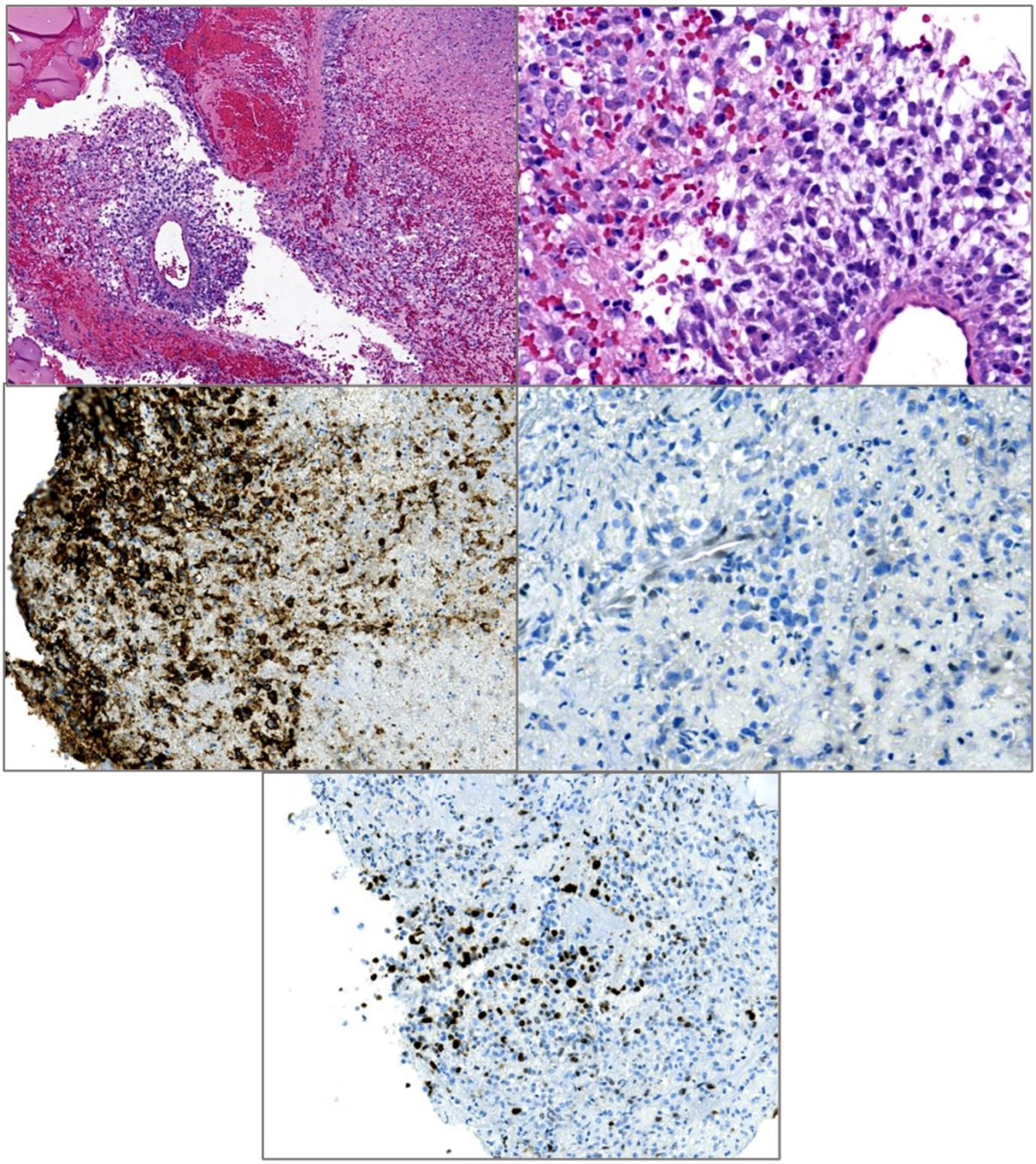

Figure 3: Histopathological examination: A: H&E slide shows hemorrhagic and necrotic neoplasm with focal papillary architecture. B: The tumor is composed of cells with round nuclei, prominent nucleoli and scant cystoplasm. C: Immunohistochemical stain (EMA), positive in the tumor cells. D: Immunohistochemical stain (INI-1), loss of expression in the tumor cells and retention in the internal positive controls encluding endothelila cells. E: (KI67) proliferation index, focally elevated.