An Atypical Case of Atypical Teratoid Rhabdoid Tumor (ATRT)

Sharmeen Mahmood
Henry Ford Health System

Hadi Mohammed
Henry Ford Health System

Follow this and additional works at: https://scholarlycommons.henryford.com/merf2019caserpt

Recommended Citation
Mahmood, Sharmeen and Mohammed, Hadi, 'An Atypical Case of Atypical Teratoid Rhabdoid Tumor (ATRT)' (2019). Case Reports. 43.
https://scholarlycommons.henryford.com/merf2019caserpt/43
An Atypical case of Atypical Teratoid Rhabdoid Tumor (ATRT)

Dr. Sharmeen Mahmood (PGY-2) & Dr. Hadi Mohammed (PGY-2), Department of Internal Medicine, Henry Ford Hospital; Dr. Kamran Thanikachalam & Dr. Jawad Z. Sheqwara (Attending), Department of Hematology & Oncology, Henry Ford Hospital.

Henry Ford Health System, Detroit, Michigan

Introduction

- Atypical Teratoid Rhabdoid Tumors are rare pediatric tumors that usually occur at age <3 years.1
- These tumors are scarcely seen in adults, with the first adult case appearing in 1992.2
- Since then, about 64 cases have been reported in the literature.3
- As such, much has been learned about adult ATRT cases and extrapolation from pediatric cases.
- The loss of INI1 (SMARCB1) or BRG1 (SMARCA4) genes are implicated in pathogenesis of ATRT.3
- Specifically, the INI1/SMARCB1 gene is classified as a tumor-suppressor gene that encodes a core subunit protein of the ATP-dependent SWI/SNF chromatin remodeling complex.4

Case Presentation

- A 62-year-old Caucasian right-handed female with history of hypertension and sinusitis presented with 2-month history of bilateral, frontal headaches reaching 5/10 in severity with associated nausea, emesis, polydipsia and polyuria.
- Polyuria occurred every hour while polydipsia included drinking (15-20) 16 oz. bottles per day.
- She presented to a local hospital and was found to have hyponatremia (Sodium 154 mEq/mL).
- Non-contrast brain Computerized Tomography (CT) revealed a 1.2 cm x 1.1 cm x 1.7 cm sellar mass with suprasellar extension.
- Urine studies diagnosed central diabetes insipidus, responsive to D-amino D-arginine vasopressin (DDAVP). However, she developed seizures and Abducen’s nerve palsy.
- Magnetic resonance imaging (MRI) of the brain demonstrated intraventricular and subarachnoid hemorrhage along with optic nerve edema.
- Prior to surgery, she was found on the floor of her room in pool of urine with incoherent speech and a sluggish papillary reflex on right side.
- STAT non-contrast head CT revealed 2.7 x 1.8 x 2.5 cm extension of hemorrhage into interpeduncular cisterns and ventricles with associated 3rd ventricle & lateral ventricle. There was no midline shift or impending herniation.
- She received bi-coronal craniotomy with excision of sellar mass and right frontal external ventricular drain placement.
- Hydrocephalus and hemorrhage improved on subsequent MRI a few days later.
- The pathology report came back positive for a malignant epithelioid neoplasm, specifically sellar ATRT, WHO grade V. Tumor was shown to be SMARCB1/INI1 deficient and no metastatic lesions were found.
- Recommendations were made for craniospinal radiation and chemotherapy afterward.

Histology

- Figure 1: Rare cells with rhabdoid features (arrow), eccentric nuclei, and eosinophilic cytoplasmic hyaline inclusions (hematoxylin–eosin, >600).8

MRI

- Figure 2: MRI showing hemorrhage of sellar mass (Red arrows) from (A) Sagittal view and (B) Coronal view.

Conclusion

- She was discharged to facility for radiation therapy. During rehab however, she was noted to be less responsive than usual.
- CTH showed worsening hydrocephalus with IVH and she was transferred to HFH.
- She underwent Left Frontal VP shunt with EVD placement.
- Initial GCS on admission was E2V1TM3.
- Her shunt was externalized to facilitate drainage.
- There was a deterioration in neurological status and CT head showed new ICH related to tumor.
- Goals of care discussion lead to terminal extubation.

Discussion

- ATRTs remain rare and aggressive brain tumors seen in both the pediatric and adult population.
- The management of ATRT remains a difficult challenge with multimodal approaches to treatment remaining the mainstay.
- Resection followed by radiation and chemotherapy has been shown to significantly increase 5-year overall survival rate,5 yet median time to progression remains in the range of 6-10 months.6
- Much more standardization is required in the treatment of disease, since patients continue to get variable approaches to treatment.
- Additional, radiation doses and optimal chemotherapy regimens have yet to be determined.7
- A promising step towards these answers have been in-vitro studies of Insulin-growth factor receptor (IGF-1R) inhibition in sensitizing the tumor to chemotherapy and radiation.5

References