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| Title | AT/RT in Association with Germ Cell Tumor |
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| Aim | Atypical teratoid/rhabdoid tumor (AT/RT) is a rare, highly malignant embryonal tumor of the CNS. The tumor is found almost exclusively in children less than three years of age, with only a handful of reported adult cases. Because of the considerable morphologic overlap with other primary CNS primitive neuroectodermal tumors (PNETs), AT/RT can be viewed as an extension to the spectrum of CNS PNETs. Germ cell tumors (GCTs) are unique in their ability to differentiate along multiple lineages and are known to give rise to a potpourri of benign and malignant neoplasms. We have recently shown that most PNETs arising in the setting of testicular GCT resemble pediatric-type CNS embryonal tumors (Ulbright et al). |
| Materials & Methods | Herein, we describe a case of AT/RT occurring in an adult with a history of GCT. |
| Result | The patient was diagnosed with disseminated mediastinal GCT at the age of 18 and received chemotherapy at this time. He soon developed brain metastases and underwent whole brain radiation. Eighteen years after his initial diagnosis of primary mediastinal GCT, the patient developed a new right temporal lobe mass. He underwent gross total resection twice, once after the original presentation and another following a recurrence five months later. Pathology was consistent with AT/RT. We hypothesized that AT/RT was related to this patient's GCT and sat out to establish a molecular link between the two. We performed FISH for 12p abnormalities, which are commonly seen in GCTs but not in AT/RTs, on both the GCT and AT/RT specimens. Interestingly, isochromosome 12p was identified in both samples, essentially establishing a GCT origin for the AT/RT. This is the first report of an AT/RT arising in the context of GCT. |
| Conclusion | We suggest that AT/RT be added to the short list of highly malignant tumors that may arise in the setting of disseminated germ cell tumors. |