Thyroid

THYROID CANCER CASE REPORTS II

Poorly Differentiated Thyroid Carcinoma Metastatic to the Adrenal Gland

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Background: Poorly differentiated thyroid carcinoma (PDTC) constitutes 1-15% of all thyroid cancers.[1] Invasive adrenal metastases secondary to PTDC are exceedingly rare.

Clinical Case: A 64-year-old woman with a non-toxic multinodular goiter presented with right upper quadrant abdominal pain and distension for three months. CT imaging revealed a 13.5 cm right suprarenal retroperitoneal mass invading the liver and inferior vena cava (IVC), concerning for adrenocortical carcinoma. She underwent resection of the mass with *en block* right adrenalectomy, partial hepatectomy, and IVC resection. Pathology demonstrated metastatic thyroid cancer with necrosis of the adrenal gland and IVC. Immunohistochemical staining was positive for PAX8, TTF1, and thyroglobulin (Tg). Completion thyroidectomy revealed an encapsulated 2 cm focus of PDTC with Hurthle cell phenotype in the right thyroid lobe. The mitotic activity was 5/10 per HPF. There were focal areas of tumor necrosis, 3 foci of capsular invasion, and extensive angioinvasion. Surgical margins were free of tumor invasion. Eight resected lymph nodes were negative for malignancy (Stage T1bN0M1; AJCC 8, Stage IVb). Genetic testing was positive for somatic mutations of *NRAS*, *TERT*, *PTEN*, and *GNAS* with broad copy number loss on chromosome 22q conferring aggressive tumor behavior.[3]

MRI of the brain and spine ruled out additional metastases. A radioactive iodine (RAI) whole-body scan (WBS) showed residual uptake of 7.6% in the right thyroid bed and a focus of increased uptake at the right sternoclavicular joint. A therapeutic dose of 206 mCi of I-131 was administered. A post-therapy WBS demonstrated focal activity in the right thyroid bed, distal right clavicle, and lower lung lobes. Chest CT and MRI of the right shoulder revealed no structural evidence of metastases corresponding to radiotracer uptake. The stimulated Tg level prior to RAI was 323 ng/mL with a TSH of 66 uU/mL (0.4-4.6 uU/mL). Tg antibodies were undetectable. She was maintained on 150 mcg of levothyroxine with the goal of TSH suppression. Tg levels declined to 4.8 ng/mL at three months, and to 0.3 ng/mL eight months post-RAI.

Discussion: PDTC is an aggressive thyroid cancer subtype with distant metastasis reported in 36-85% of cases.[2] Distant metastasis is predictive of poorer outcomes, with patients three times more likely to die from the disease than those without metastatic disease.[1] Adrenal metastasis of thyroid cancer is rare, and unlike in our patient, usually asymptomatic and frequently detected on a post-therapy scan. Despite a dramatic response to therapy, given the poorly differentiated features of the primary tumor, a whole-body PET-CT is warranted to evaluate for RAI refractory disease.

References:
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Neuroendocrinology and Pituitary

CASE REPORTS IN UNUSUAL PATHOLOGIES IN THE PITUITARY II

A Case of the Suprasellar Atypical Teratoid Rhabdoid Tumor (ATRT) Presenting in an Adult Treated with Intrathecal Chemotherapy

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Background: Atypical teratoid rhabdoid tumors (ATRTs) are highly malignant tumors that usually present as a posterior fossa mass in children less than 3 years old. Only 38 cases have been reported in adults. They are also typically located in the supratentorial region. In none of the reported cases of suprasellar ATRT in the adult, intrathecal chemotherapy (via ommaya) has been used. **Clinical case:** A 70-year-old woman presented with a severe headache and magnetic resonance imaging (MRI) revealed a suprasellar mass measuring 2.9 x 2.1 x 3.0 cm. Shortly after her presentation, she developed an acute 3rd nerve palsy, and repeat MRI found dramatic interval growth. A transsphenoidal approach for biopsy/resection was attempted, but the lesion was not accessible via this corridor. She then underwent a right frontotemporal craniotomy and subsequently developed panhypopituitarism, including diabetes insipidus. Pathology revealed poorly differentiated malignant cells. Immunohistochemistry was positive for synaptophysin, Epithelial Membrane Antigen (EMA), Tumor protein p53, and negative for integrase interactor 1 (INI-1 antibody) with loss of expression in tumor nuclei with positive internal control in endothelial cells. These findings confirmed the diagnosis of ATRT. The Ki-67 index was 60% consistent with a highly proliferative tumor. One month later, she developed acute mental status change. Repeat computed tomography, and MRI showed recurrence of the tumor at the same location with new leptomeningeal enhancement involving the left facial nerve. Multimodal treatment was instituted, consisting of intraventricular/intrathecal chemotherapy with etoposide and topotecan plus fractionated external beam cranial irradiation (30 Gy in 10 fractions). She continued to deteriorate, and following consultation with her family, she was transferred to hospice care and died six months following her initial surgery. **Conclusion:** This is the first case of adult suprasellar ATRT that has been treated with intrathecal chemotherapy. There is no consensus on the best combination of chemotherapy, and often the St. Jude's protocol used in the treatment of pediatric ATRT is used. In line with the biological behavior reported for this tumor in children and adults in different locations, the tumor was very aggressive, resulting in the patient's death only after 6 months from the diagnosis despite aggressive surgical and medical treatment.

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