developed primary hyperparathyroidism, non-functional pancreatic neuroendocrine tumors, Zollinger-Ellison syndrome, bilateral adrenal hyperplasia, and bronchial carcinoid. At the age of 49, magnetic resonance imaging (MRI) and computed tomography (CT) scan of the chest incidentally demonstrated a 2.5 x 6 x 10-cm anterior mediastinal mass, with marked compression of the left brachiocephalic vein and encasement of the superior vena cava. Biopsy was consistent for malignant cells of thymic epithelial origin. A median sternotomy with en bloc resection with SVC resection and reconstruction, mediastinal lymph node dissection and placation of the right hemidiaphragm were performed. Pathology revealed WHO type B3 thymoma extending into the pulmonary parenchyma with positive tissue margins but negative lymph node involvement. Following surgery, she underwent adjuvant radiation therapy with a total dose of 59 Gy. Annual screening showed disease remission. However, nine years after initial presentation, surveillance CT scans revealed a pleural base mass with mass effect on the superior portion of the IVC, as well as hypoattenuated masses within the liver. Positron emission tomography with fluorodeoxyglucose (^{18}FDG PET-CT) confirmed multiple metastatic lesions involving thorax and abdomen. Biopsy of the retrocaval soft tissue subxiphoid mass revealed epithelioid cells in a background of lymphocytes consistent with recurrent thymoma. She was subsequently staged as IV B thymoma and was recommended to start systemic chemotherapy.

Conclusion: We described a case of an aggressive thymoma in a patient with MEN1 syndrome demonstrating that their association exists and the clinical presentation can be aggressive. Thus, it is important for practitioners to screen for thymic tumors routinely in patients with MEN1 for early detection as they can be a major cause of mortality. Although further studies are needed, improving the detection of these tumors could significantly contribute to reducing MEN1-related deaths.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS

Use Of Temozolomide In Parathyroid Carcinoma With Negative Mgmt Promoter Methylation

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Introduction: Parathyroid carcinoma (PC) is a rare malignancy with a high rate of recurrence and metastasis. **Case:** A 63-year-old man with a 13-year history of recurrent PC requiring 5 operations, including parathyroidectomies, thyroidectomy, and neck dissections presented with polyuria, polydipsia, and worsening rib pain. He had been recently treated with 6 monthly octreotide injections and maximal dose cinacalcet for gradually rising Ca/PTH levels. Tests revealed serum Ca 13.1mg/dL (8-10.5mg/dL), PTH 1750pg/mL (11-90 pg/mL), and serum Cr 3.34mg/dL (0.5-1.3mg/dL). Imaging identified tumor in the right 6th rib (3.6cm lytic lesion), and soft tissue lesions in the left thyroid bed (3 masses, the largest 1.6cm) and the suprasternal notch (1.1cm). He underwent rib resection (metastasectomy) and PTH declined from 2334pg/mL to 671pg/mL. Although metastasectomy improved the PTH level, Ca levels began to rise from the residual tumor. A multidisciplinary team deemed the risk of complications from repeat neck surgery to be prohibitively high. Temozolomide (TEM) (150-200mg/m2/d x 5d, q28d) was instituted 3 months after the rib resection. 13 months later, PTH has stably ranged from 600-800 pg/mL with a normal serum Ca of 9.8mg/dL. Recent imaging shows stable disease in the neck, without distant disease. **Discussion:** The mainstay of therapy for initial and recurrent/metastatic PC is surgery. Inoperable disease has a poor prognosis because of lack of effective systemic therapies. Radiation and chemotherapy have not shown much efficacy. Results of treatment with octreotide have not been encouraging. Anti-PTH immunotherapy and Lutathera are promising but require further investigation. Usually, no targetable mutation is found. Anti-angiogenic TKI's (sorafenib, lenvatinib) have been used with varying success. An exciting therapy used in this patient is TEM, an alkylating agent used for CNS tumors, neuroendocrine tumors (NET) and aggressive pituitary tumors. A previous report described successful use of TEM in a case of metastatic PC, whose tumor harbored high O6-methylguanine DNA methyltransferase (MGMT) promoter methylation status, a known predictor of positive response in CNS tumors. Promoter methylation is an epigenetic alteration that leads to low MGMT enzyme activity & enhances the cytotoxicity of TEM. Some studies in NET demonstrated tumor response irrespective of MGMT status. This leads to the question of whether the same is true in PC. Our patient has radiographic/biochemical stable disease on TEM, and a surprising retrospective discovery was that the MGMT promoter was unmethylated. This is a unique case of PC which seems to be responding to TEM despite absent promoter methylation. Further studies are warranted, as the incidence of PC is rising over the past decades. In the interim, clinicians could consider using TEM for in-operable PC irrespective of MGMT methylation status.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS

Well-Differentiated Malignant Neuroendocrine Cell Tumor Originating From the Jejunum With Excessive Insulin Secretion

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Background: Insulinomas are rare, well-differentiated insulin-secreting neuroendocrine tumors of the pancreas. Insulin-secreting, extra-pancreatic tumors have rarely been reported in the literature. We present a case of a well-differentiated neuroendocrine tumor with clinical, laboratory, and immunohistochemical tumor findings supportive of ectopic insulin production in a non-pancreatic neuroendocrine tumor of the small bowel.

Clinical Case: A 70 year-old female with past medical history of end stage renal disease, Type II diabetes mellitus, and hypertension presented to our facility with multiple episodes of dizziness and syncope due to hypoglycemia.