of the gene for IGF-2 results in increased production of the more biologically active pro-IGF-2 that does not form a complex with IGFBP3 and is available to interact with insulin receptors. In patients with normal IGF-2 (chronic kidney disease, poor nutritional status), an IGF-2: IGF-1 ratio >3 can be used to confirm NICTH. IGFBP-3 was not measured in this patient but it is likely this was low due to her poor nutritional status. The most definitive treatment for NICTH is resection of the tumor. Pharmacological management can be considered in patients who are not surgical candidates, but is not always successful as was observed in this patient. Conclusion: This is an unusual case of malignancy associated hypoglycemia in a woman with type 1 diabetes who required discontinuation of all insulin therapy as well as continuous dextrose infusions to achieve euglycemia and briefly maintain an acceptable quality of life over a period of several months.

Tumor Biology
ENDOCRINE NEOPLASIA CASE REPORTS

Presentation of Pheochromocytoma, Papillary Thyroid Cancer and Hyperparathyroidism in Three Family Members

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Introduction: Although rare, one of the most common inherited disorders is multiple endocrine neoplasia. It is an autosomal dominant disorder that predisposes individuals to certain endocrine abnormalities depending on which type. The type 2A is a combination of medullary thyroid cancer, hyperparathyroidism, and pheochromocytoma which have been explained to be due to a mutation in the RET proto-oncogene. This abstract present a case of a patient with hyperparathyroidism whom family members also have pheochromocytoma and papillary thyroid cancer. Case description: A 45-year-old Hispanic male came to the endocrinology clinic complaining of constipation and headache. He has a personal history of non-toxic multinodular goiter and underwent right sided thyroidectomy in 2015 with pathology report showing follicular adenoma. He is currently on thyroid replacement therapy. He is clinically and biochemically euthyroid with TSH of 2.29 IU/ml. Physical examination was unremarkable. His labs were pertinent for calcium 11.5mg/dl, parathyroid 245.7pg/ml, creatinine of 1.5mg/dl. Two years ago, parathyroid was 189.5pg/ml and calcium was 11mg/dl. 1.5 year ago parathyroid level was 235.5pg/ml, calcium was 11.4mg/dl, urine calcium 9.3mg/dl, 24hr urine calcium 286.4mg, calcitonin <2pg/ml, vitamin D 23ng/ml, 1,25 vitamin D 53ng/ml. In 2017, Sestambi scan showed equivocal focus of faint parathyroid activity in the region of the mid to lower left thyroid lobe versus faint residual thyroid activity and in 2019, scan showed no definite parathyroid adenoma. Surgical intervention was recently recommended due to patient’s DEXA scan showing osteoporosis of the femoral neck. The family history of this patient is pertinent for two sisters; one with pheochromocytoma and the other with papillary thyroid cancer. One of the sisters is a 60 years old diagnosed with pheochromocytoma at 51. Her free normetanephrine level was 682pg/ml and total metanephrine was 727pg/ml at time of diagnosis. Her MRI report showed right adrenal mass measuring 3.5x2.8cm. Laparoscopic right adrenalectomy was done and pathology confirmed pheochromocytoma which was RET negative. She still follows up with endocrinology and calcitonin, chromogranin A and plasma metanephrines have been normal. The second sister is now 53 years old diagnosed with papillary thyroid cancer at age 27 and had total thyroidectomy with pathology confirming papillary thyroid cancer. Discussion: Based on the clinical presentation of these family members, the most likely explanation is familial inheritance. This pattern of inheritance cannot be explained by MEN 2A or 2B due to the absence of medullary thyroid cancer. It has also been reported that this unusual presentation could be a variant of MEN 2A.[i] Due to the family history, close follow up is required to monitor for the possible development of other endocrinopathies in the future.

Tumor Biology
ENDOCRINE NEOPLASIA CASE REPORTS

Recurrent Pheochromocytoma, as Component of Multiple Endocrine Neoplasia

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Background: Pheochromocytoma is a rare neuroendocrine tumor from the adrenal medulla’s chromaffin cells that secrete catecholamines. The mainstay of treatment is surgery. Although rare, it has a recurrence rate of 6.5-16.5% even after adequate surgical removal with a notable increase in prevalence among genetic syndromes, extra-adrenal tumors, larger tumor size, and younger age of diagnosis. Case: A 23-year-old Filipino male with no known familial disease presented with episodic headache, palpitations, dizziness, and resistant hypertension. Two masses in the left adrenal gland, with the larger one measuring 6.0cm x 5.0cm x 3.0cm, were surgically removed. Histologic examination revealed pheochromocytoma. Post-operatively, there was the normalization of urinary metanephrines and the resolution of the signs and symptoms. He was lost to follow-up and returned five years later with an abdominal ultrasound demonstrating a right adrenal mass. He had no accompanying signs and symptoms; the physical examination was unremarkable. On workup, biochemical testing revealed two 24-hour urinary metanephrine levels were markedly elevated, 14.49 and 19.97 (NV:0-1mg/24 hours). Calcitonin: 644 (NV:0-18pg/ml) were also elevated. Mild hypercalcemia was noted with elevated intact PTH 101.74 (NV:0-65pg/ml). Parathyroid scintigraphy was normal. The neck ultrasound showed multiple bilateral thyroid nodules, with the largest measuring 1.2cmx0.9cm (TIRADS IV).
The abdominal CT scan showed two well-defined, homogenous right adrenal masses, with the largest measuring 6.3cm x 5.6cm x 7.4cm. He underwent right adrenalectomy, and histopathology showed pheochromocytoma with Pheochromocytoma of the Adrenal Scale Score (PASS) of 3. One month later, he underwent total thyroidectomy with neck dissection and inferior parathyroid gland resection. Histopathology confirmed medullary thyroid cancer with nodal metastasis and parathyroid adenoma. The patient recovered well without complications. Biochemical tests normalized eight weeks post-surgery. The presence of synchronous recurrent pheochromocytoma, medullary thyroid cancer (MTC), and primary hyperparathyroidism is consistent with multiple endocrine neoplasia 2A (MEN2A) syndrome. Conclusion: Lifelong follow-up is essential in patients treated for pheochromocytoma despite the complete removal of the adrenal masses due to recurrence risk. Recurrence should likewise raise the suspicion of MEN2, a genetic syndrome. In addition, the pheochromocytoma has malignant characteristics, and the MTC has metastases to the cervical lymph nodes. Early detection and prompt intervention are essential for the treatment of the disease.

**Tumor Biology**

**ENDOCRINE NEOPLASIA CASE REPORTS**

**Reversible Catecholamine Induced Cardiomyopathy**

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**Background:** Pheochromocytomas and paragangliomas (PPGL) are rare neuro-endocrine tumors associated with a myriad of poor outcomes as a result of long-term exposure to catecholamines. Although paragangliomas are less commonly associated with increased catecholamine production than adrenal pheochromocytomas, there have been a few reports of catecholamine-induced cardiomyopathy in patients diagnosed with PPGL. We report a case of a PPGL associated with hypercoagulability and cardiomyopathy.

**Clinical Case:** 42-year-old man with uncontrolled hypertension presented to the emergency department with abdominal pain. On CT imaging, he was found to have hepatic lesions, aortocaval lymph node concerning for metastatic disease, left renal infarct, and a left ventricular thrombus. Soon after his admission, he developed acute ataxia, gaze palsies and left hemiparesis. CTA of the head showed a basilar artery thrombus [FJ1] which was treated with emergent thrombectomy. In addition patient had absent distal basilar artery thrombus [FJ1] which was treated with emergent intervention. In addition patient had absent distal basilar artery thrombus which was treated with emergent intervention. In addition patient had absent distal basilar artery thrombus which was treated with emergent intervention.

Further workup with abdominal MRI showed retroperitoneal mass [FJ3] and multiple hepatic lesions concerning for metastatic extra-adrenal neuroendocrine tumor. Plasma normetanephrine was 4.5 nmol/L (ULN 0.89), plasma metanephrine 0.3 nmol/L (ULN 0.49) Chromogranin A was 387 ng/ml (ULN 160). Ga-68 DOTATE scan was consistent with an extra adrenal paraganglioma with less prominent radiotracer activity in hepatic lesion concerning for dedifferentiated metastatic disease. In addition, echocardiogram showed reduced LV ejection fraction of 24% with global hypokinesis, and confirmed the LV thrombus. Cardiac MRI showed infiltrative nonspecific cardiomyopathy and mild dilation of left ventricle, as well as patchy delayed enhancement in the basal and inferoseptal walls suggestive of myocarditis. Treatment included rivaroxaban [FJ4], lisinopril, doxazosin, furosemide, and carvedilol. Several months after discharge, his EF improved to 48%. Hepatic lesions concerning for dedifferentiated tumor vs unrelated malignancy was biopsied [FJ5] and consistent with neuroendocrine tumor. Future plan for his PPGL include revaluation for resection of retroperitoneal mass or DOTA Lutathera therapy. **Conclusions:** This case highlights a young man who was incidentally found to have metastatic paraganglioma with catecholamine-induced cardiomyopathy. The patient was asymptomatic until he developed significant heart failure. Cardiomyopathy in this setting is thought to be secondary to uncontrolled hypertension, as well as sympathetic overdrive from overstimulation of norepinephrine. We present the case to highlight the management challenges in a patient with PPGL with significant cardiovascular compromise and limited therapeutic options.

**Tumor Biology**

**ENDOCRINE NEOPLASIA CASE REPORTS**

**Serendipity and the Second Malignancy: Clear Cell Renal Carcinoma Workup Revealing a Paraaortic Paraganglioma With Post-Operative Vasoplegia**

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Pheochromocytoma and paraganglioma (PPGL) are neuro-endocrine tumors requiring careful pre- and post-operative management to dampen fluctuations in catecholamines. These lesions are not typically biopsied due to risk of catecholamine-induced hemodynamic instability. This case illustrates the importance of considering PPGL in patients without typical symptoms, and highlights challenges with post-operative blood pressure management following use of phenoxybenzamine. A 52 year old man presented with a 40 lb weight loss. PET/CT revealed a right renal mass, enlarged retroperitoneal lymph nodes, and increased uptake in paraaortic lesions. P araortic lymph node biopsy was consistent with paraganglioma. This finding was unexpected as the patient did not report palpitations, paroxysmal hypertension, or diaphoresis. He had no family history of neuroendocrine tumors. Fortunately, he had no adverse effects during biopsy despite subsequent testing showing elevated plasma metanephrines (271 pg/mL, normal ≤ 57 pg/mL) and normetanephrines (770 pg/mL, normal ≤ 148 pg/mL). I-123 MIBG scan revealed an enlarged left paraaortic mass with increased activity consistent with paraganglioma. He started preoperative alpha blockade with phenoxybenzamine 14 days prior to surgery. Propranolol was added 10 days later to provide combined sympathetic blockade.