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
Kathrin Sandra Töfj, MD, Malek Mushref, MD.
1MEDICAL UNIVERSITY OF SOUTH CAROLINA, Charleston, SC, USA, 2Medical University of South Carolina, Charleston, SC, USA.

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 pulse of the right foot[FJ2], and found to have thrombus of the popliteal artery, which was treated with thrombectomy. 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 differentiated metastatic disease. In addition, echocardiogram showed reduced LV ejection fraction of 24% with global hypokinesis, and confirmed the LV thrombus. Cardiac MRI showed infraficial nonischemic 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
Robert Lee Thomas, III, MD PhD1, Preethika Sabashini Ekanayake, MD2, Jeremy Pettus, MD3, Mizuho Mimoto, MD, PhD4.
1UCSD, LA JOLLA, CA, USA, 2UCSD, Coronado, CA, USA, 3UCSD, La Jolla, CA, USA, 4University of California, San Diego, Encinitas, CA, USA.

Pheochromocytoma and paraganglioma (PPGL) are neuroendocrine 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 retroperitoneal and paraaortic lesions. Paraaortic 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.
The patient underwent partial right nephrectomy and resection of the paraaortic mass. Pathology showed renal cell carcinoma (RCC) and paraganglioma, respectively. On post-operative day 1, maintenance fluids were discontinued and he developed palpitations and tachycardia to 140 beats per minute, with blood pressure in the 130s/80s. RCC associated pulmonary embolism, beta-blocker withdrawal, and vasopengia due to phenoxybenzamine use and paraganglioma resection were considered. The patient was treated with 4 additional liters of normal saline over the next two days to address post-operative vasopengia, and his tachycardia resolved. Genetic testing for neuroendocrine tumor syndromes including Von Hippel-Lindau (VHL) is ongoing.

Clinical Lessons: 1. PPGL should be considered in patients with newly identified intraabdominal masses, even in patients without typical symptoms of catecholamine excess to avoid high risk biopsy. 2. Paraganglioma resection is associated with vasopengia due to post-operative reduction in circulating catecholamines. 3. The covalent, irreversible alpha antagonist phenoxybenzamine accumulates in adipose tissue, and clinical effects can last up to 7 days after discontinuation. Hypotension can be avoided with aggressive fluid resuscitation. Beta antagonists should be used with caution as they may precipitate hypotension. 4. For patients presenting with paraganglioma and renal cell cancer, genetic syndromes including VHL or RAPTAS etiologies should be considered.

Tumor Biology
ENDOCRINE NEOPLASIA CASE REPORTS
Simultaneous Occurrence of Germline Pathogenic Allele Variants of TMEM127 and TP53 in a Brazilian Family With Li-Fraumeni Syndrome
Jose Viana Lima, MD1, Nilza Maria Scalliisi, MD, PhD2, Gustavo Pich Ricard, Mr Gustavo Pich3, Rosa Paula Mello Biscolla, MD, PhD4, Maria Izabel Chiamolera, MD, PhD5, caroline olivati, Dr, MD6, Wagner Baratela, Dr, MD, PhD6, Elisa Napolitano Ferreira, PhD6, Claudio E. Kater, MD, PhD7.
1Santa casa de So Paulo, So Paulo, Brazil, 2Santa Casa de SP, Sao Paulo, Brazil, 3Santa Casa de SP, Sao Paulo, Brazil, 4Universidade Federal de Sao Paulo, Fleurly Medicina e Sade, Sao Paulo, Brazil, 5UNIFESP, Sao Paulo, Brazil, 6Fleury, So Paulo, Brazil, 7Federal University of Sao Paulo, Sao Paulo SP, Brazil.

Background: We will describe a Brazilian family whose index case had pheochromocytoma and in the evaluation of the genetic panel by Next Generation Sequence (NGS), the germline pathogenic variants in the TMEM127 and TP53 genes were identified. Clinical Case: A 32-year-old female patient with a clinical picture of paroxysms and difficult to control arterial hypertension, with a personal history of stroke and acute myocardial infarction. She had a 6.5 cm tumor in the right adrenal and urine metanephrine levels of 5.5 mg / g creatinine (VR <1 mg / g creatinine) compatible with pheochromocytoma. She underwent laparoscopic right adrenalectomy. There was a reversal of arterial hypertension and paroxysms. 10 years after adrenalectomy, she was diagnosed with bilateral breast cancer, she underwent radical total mastectomy and 2 years ago there was a recurrence of breast cancer and currently undergoing chemotherapy. Germinative genetic panel carried out by NGS had identified pathogenic variants c.1010G> A, p. (Arg337His) in heterozygosity in the TP53 gene and c.117_120del p. (Ile41Argfs * 39) in heterozygosis in the TMEM127 gene. Her 28-year-old daughter diagnosed bilateral breast cancer and meningoema in the central nervous system and she had the same pathogenic variants germlines. Thus far, there is no clinical, laboratory or radiological picture of pheochromocytoma. Her 11-year-old granddaughter has only the pathogenic allele variant c.117_120del p. (Ile41Argfs * 39) in heterozygosity in the TMEM127 gene and thus far she has no clinical, laboratory and radiological picture of pheochromocytoma.

Conclusion: This is the first case report of the simultaneous occurrence of pathogenic germline variants in the TMEM127 and TP53 genes. Reference: 11) Toledo RA et al Consensus Statement on next-generation-sequencing-based diagnostic testing of hereditary pheochromocytomas and paragangliomas. Nature Reviews Endocrinology 13, 233-247 (2017).

Tumor Biology
ENDOCRINE NEOPLASIA CASE REPORTS
Surgical Management of Large Gastrinomas in a Young Patient With MEN1
Nydia Burgos, MD, Janet Marie Colon Castellano, MD, Yadiel Rivera Nieves, MD, Nicolle Canales Ramos, MD, Alberto Javier Grana Santini, MD, Nicole Hernández Cordero, MD, Melba Feliciano-Emmanuelli, MD, Loida Alejandra Gonzalez-Rodriguez, MD, Milliette Alvarado-Santiago, MD, Margarita Ramirez-Vick, MD.
Division of Endocrinology, Diabetes and Metabolism, Department of Medicine, University of Puerto Rico School of Medicine, San Juan, PR, USA.

Multiple Endocrine Neoplasia 1 (MEN1) is a well-described hereditary disorder that requires a multidisciplinary approach. Gastrinomas are the most common enteropancreatic tumors found in MEN1. They often appear as small (< 0.5 cm) multifocal lesions and are rarely found as large masses in the pancreas. The crossroads of deciding between medical versus surgical management when treating these tumors requires an evidenced-based- and patient-centered approach. We describe a rare case of a young patient with MEN1 and large pancreatic gastrinomas. A 23-year-old female patient with MEN1 (prolactinoma, primary hyperparathyroidism) was evaluated for the development of hypocalcemia after surgical excision of 3 parathyroid glands. A prior history of a perforated peptic ulcer prompted further evaluation that revealed gastrin levels of 481 pg/ml (13 - 115 pg/ml) off any acid suppression therapy, and an abdominopelvic MRI that revealed two T1 hypointense lesions measuring 2.4 cm and 1.4 cm at pancreatic head and tail, respectively. Both lesions resulted to be grade 2/3 neuroendocrine tumors consistent with gastrinoma. Abdominopelvic CT scan for staging showed 6 isodense lesions distributed within the pancreas measuring up to 3.2 cm without lymphadenopathy nor metastatic liver lesions. After extensive discussion regarding management, the patient opted for a surgical approach.