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 vasoplegia 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 vasoplegia, 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 vasoplegia 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

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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 meningeoma 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 pheocromocytomas 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

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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.