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Introduction: A composite pheochromocytoma (PC) is an adrenal tumor that is often diagnosed post-operatively on histopathology. PCs are unique in that it is a combination of typicalpheochromocytoma and neural crest derived tumors. The incidence is reported to be less than 3%of adrenal neoplasms. The most common co-existing tumor within a PC is a ganglioneuroma. Wepresent a rare case of PC containing ganglioneuroblastoma (PC-GNBL) in a woman withoutsignificant biochemical manifestation of excess catecholamine production. **Case Report:** A 63-year-old woman with a prolonged history of uncontrolled hypertension on 4 oral anti-hypertensive medications (Amilodipine 10mg daily, Valsartan/HCTZ 320/25mg daily andClonidine 0.1mg/24h patch) and uncontrolled type 2 diabetes on insulin was diagnosed with a1.5x1.8x1.5cm right adrenal incidentaloma 2 years prior on CT imaging for abdominal pain. Hormonal evaluation was notable for plasma free metanephrine of 66 (<57pg/ml),normetanephrine 229 (<148pg/ml), and total metanephrines of 295 (205 pg/ml). However, 24-hour urine metanephrine evaluation was normal on two occasions: metanephrine 121 and 168mcg/24h (90-315), norepinephrine 237 and 336 mcg/24h (122-676) and total metanephrines 358and 504mcg/24 h (224-832). Hyperaldosteronism and hypercortisolism were ruled out. Follow-up CT scan 14 months later demonstrated growth of the right adrenal nodule to 1.7x2x2cm with49% washout. She underwent laparoscopic right adrenalectomy without perioperativecomplications. Pathology was consistent with a PC-GNBL. The PASS score was 7, consistentwith malignant pheochromocytoma. Within weeks of surgery, she had marked clinicalimprovement. Blood pressure was controlled on one anti-hypertensive and Hgb A1c decreased to7.1% from 11% without requiring insulin. CT abdomen/pelvis 6 months post-operatively did notshow evidence of metastasis. She was referred for genetic testing. **Conclusion:** This case highlights an unusual presentations of pheochmocytoma. It's important to recognizethat resistant hypertension can present without episodic headaches, diaphoresis, palpitations, andwithout biochemical evidence of catecholamine excess. Composite PCs are indistinguishableclinically or radiologically from ordinary pheochmocytomas. These exceedingly rare mixedtumors are only diagnosed via surgical pathology. To date, there are only a few cases reported inthe medical literature of co-existing PC-GNBL tumors. Due to the scarcity of composite PCscases, important information regarding its presentation and prognosis are unknown. It remains tobe seen whether the GNBL part of the tumor changes the prognosis of the tumor. However, inour case, the clinical status of our patient improved.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS

Description of a SDHD c.129G>A (p.W43X) Mutation With Variable Presentation in Multiple Family Members.

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Approximately 40% of paragangliomas and pheochromo cytomas are attributed to hereditary mutations. *SDHD* mutations account for 7% of inherited mutations (PGL1 syndrome), is maternally imprinted and has variable penetrance. *SDHD* pathogenic variants (PV) have been previously described extensively in Dutch pedigrees, with a varying lifetime risk for tumor development. Here we report a large family (Fig 1) displaying a *SDHD* c.129G>A (p.W43X) variation in 12 family members, 10 of whom had screening or tumor history available. The presentation and age of diagnosis in family members showed variable penetrance. Age at first diagnosis of a pheochromocytoma/paraganglioma ranged from 10 - 45 years. Family members displayed bilateral pheochromocytomas, bilateral carotid body tumors, and paragangliomas of the head, neck and trunk with variable recurrence rates (none to multiple). No malignant lesions were detected to date. Pheochromocytomas were norepinephrine producing. Paragangliomas ranged from non-functional to dopamine and norepinephrine producing. Compared to previous reports of other *SDHD* mutations, the *SDHD* c.129G>A (p.W43X) variation displayed an earlier age at first diagnosis with a highly variable phenotype ranging from one benign, non-secreting paraganglioma to bilateral pheochromocytomas and recurrent parganagliomas along the parasympathetic chain from head to abdomen. This report contributes to the evolving understanding of the phenotypic presentations of various genetic mutations. We propose that expert guidelines that suggest screening family members with the *SDHD* c.129G>A (p.W43X) variation at the age of 8 for early detection of pheochromocytomas and paragangliomas is beneficial.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS

Development of Delayed Paraneoplastic ACTH Syndrome in Small Cell Lung Carcinoma

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Background: Paraneoplastic/ectopic ACTH syndrome (EAS) is a rare but well-recognized syndrome in small cell lung carcinoma (SCLC), occurring in 1-5% of cases (1) and most often at diagnosis. We present a patient with SCLC who developed paraneoplastic ACTH syndrome 9 months after initiation of treatment. **Case:** A 73 year old female with long history of smoking and COPD (on home O_2) and SCLC seen during hospitalization with severe muscle cramps, generalized weakness, hypertension and hypokalemia. Eleven months prior, she was diagnosed with SCLC after presenting with left superior mediastinum, supraclavicular and neck lymphadenopathy. She received 4 cycles of chemotherapy and immune checkpoint inhibitor