SAT-146

BACKGROUND: Ectopic ACTH secretion (EAS) is a rare cause of Cushing's syndrome. Olfactory neuroblastoma (ONB) is a malignant tumor derived from the olfactory epithelium and can rarely be a source of ectopic hormone production. There are only 19 reported cases of EAS from ONB. We report a case of severe ectopic Cushing's due to ONB. Interestingly, the patient also presented with Graves disease, which is an unusual pathophysiologic combination since supraphysiologic levels of glucocorticoids suppress the immune system, thereby ameliorating autoimmune processes. Remarkably, Graves disease improved following the removal of the source of ectopic ACTH. **CASE PRESENTATION**: A 41year old male presented with epistaxis, anosmia, and headaches. He also reported recent weight gain, muscle weakness, and new onset hypertension. Cross-sectional imaging revealed a right nasal cavity mass with intracranial extension. Endonasal biopsy was consistent with a diagnosis of ONB. Biochemical evaluation demonstrated hypokalemic alkalosis, hyperglycemia, and severe hypercortisolism [ACTH 734 (9-46 pg/ml), am cortisol 110 (2-15 ug/ml), late night salivary cortisol 9.8 and 22.53 (<0.09 ug/dl), 24 hour UFC 41,337.3 (4-50 mcg) and non-suppressed cortisol 110 (1.8 mcg/dl) by 1 mg dexamethasone]. Pituitary MRI showed no sellar pathology. Following a period of cortisol suppression with Ketoconazole then with Etomidate infusion, the patient underwent resection of the ONB, followed by chemo- and radiotherapy, which resulted in improvement of severe hypercortisolism. Histopathology showed positive ACTH immunostaining. Pre-operative evaluation also demonstrated hyperthyroidism: TSH <0.010 (0.3-5uIu/ml), FT4 3.11 (0.89-1.76 ng/dl), normal TT3 0.81 (0.6-1.81 ng/ml). Thyroid auto-antibodies were negative; however, radioiodine scan and uptake demonstrated diffusely increased uptake in the enlarged thyroid gland. Pre-operative euthyroidism was achieved with a combination of methimazole, SSKI and cholestyramine. After 6 months of methimazole therapy the patient presented with hypothyroidism (TSH 72.37, FT4 0.1). Methimazole was discontinued and he achieved euthyroidism on subsequent evaluations. **CONCLUSION**: EAS due to ONB is a very rare cause of Cushing's syndrome. To our knowledge, this is the first reported case of a male presenting with EAS and concurrent Graves hyperthyroidism. An extensive review of the literature and seeking expert opinion did not provide a convincing pathophysiologic explanation to this unusual concurrence. We therefore hypothesize that, while the two endocrine conditions were simultaneous, they were likely unrelated.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS I

Functioning Abdominal Paraganglioma Presenting as Acute ST-Elevation Myocardial Infarction

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SUN-924

Background:

Functioning paraganglioma is a rare catecholamine-producing tumor that arises in the sympathetic nervous system. It usually presents with sustained or paroxysmal hypertension accompanied by episodes of its classic triad of headache, palpitation, and diaphoresis. However, a wide range of signs and symptoms may be present. We report an unusual manifestation as acute myocardial infarction, which accidentally diagnosed by the trigger response from metoclopramide injection.

Case presentation

A 66-year-old woman with medical history of well-control hypertension, dyslipidemia and type 2 diabetes mellitus for 8 years presented with typical angina pain for 1 hour. She denied history of chest pain or triad symptoms of pheochromocytoma/paraganglioma (PCC/PGL). Her physical examination was unremarkable except severe hypertension, 206/89 mmHg, and occasional sinus tachycardia. Acute inferior wall myocardial infarction was proposed by an electrocardiogram study, acute ST elevation in lead II, III and aVF, and highly elevated cardiac enzymes. Echocardiogram and coronary angiography showed preserved left ventricle function (LV ejection fraction 70%) without regional wall motion abnormality. No evidence of coronary artery disease was found. During the catheterization, the cardiologist raised the possibility of the presence of PCC/PGL from her fluctuating blood pressure, 73/42 to 206/113 mmHg, after 10-mg metoclopramide injection to control her vomiting. Computer tomography of the abdomen showed a lobulated heterogeneous enhancing left para-aortic mass with internal necrosis, 6.1x4.9x4.1 cm in size, abutting left anterolateral aortic wall and encasing celiac trunk, superior mesenteric artery, and left renal arterial wall. Her hormonal study showed 24-hour urine fractionated metanephrine/normetanephrine levels of 2,924 nmol (<1,777 nmol)/4,328 nmoL(< 3,279 nmol), respectively, and plasma free metanephrine/normetanephrine levels of 93.66 pg/mL (0-96.6 pg/mL)/233.61 pg/mL (0-163.05 pg/mL). She underwent surgical tumor removal with uneventful outcome and the pathology confirmed the diagnosis of functioning PGL. During 2-years follow-up, the patient remained asymptomatic and her hormonal and functioning imaging study showed no recurrence. The genetic testing for PCC/PGL panel was negative.

Conclusion:

We present an unusual manifestation of PCC/PGL as acute coronary syndrome. The clinician should remind this tumor as the differential diagnosis, especially in a patient with negative coronary angiogram.

Pediatric Endocrinology

PEDIATRIC OBESITY, THYROID, AND CANCER

Endocrine Complications Following Cancer Treatment in Survivors of Pediatric Solid Tumors: 18 Years' Experience of a Single Academic Center

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MON-096

Background: Survival rates of pediatric cancer have been significantly improved over recent decades because of advances in chemotherapy and radiotherapy. The endocrine consequences of cancer treatment have become the