of steroid synthesis elsewhere. Extra adrenal sources for glucocorticoid production are known such as skin, gonads and thymus. However, the levels are insufficient to mount a significant stress response. There is evidence of adrenal regeneration in adrenalectomized animals. The regeneration is primarily of the adrenal cortex and does not involve the medulla. There has been one case report in literature of a 11 year old German boy who had adrenal regeneration detected on adrenal scintigraphy (Bilateral normal adrenal glands with normal activity) 13 years after adrenalectomy for Cushing's disease.

References: Gotlieb N, Albaz E, Shaashua L, et al. Regeneration of Functional Adrenal Tissue Following Bilateral Adrenalectomy. *Endocrinology*. 2018;159(1):248–259. doi:10.1210/en.2017-00505

Taves MD, Gomez-Sanchez CE, Soma KK. Extra-adrenal glucocorticoids and mineralocorticoids: evidence for local synthesis, regulation, and function. *Am J Physiol Endocrinol Metab*. 2011;301(1):E11-E24. doi:10.1152/ajpendo.00100.2011

Adrenal

ADRENAL CASE REPORTS

Cardiac Arrest in a Child with Non-classic Lipoid Congenital Adrenal Hyperplasia Associated with a New STAR Gene Mutation

Yun Yan, MD[1], Isabelle Thiffault, PhD[1], Sarah Tsai, MD[1], Luke He, MD[2], Francesco De Luca, MD[1].

[1]Children's Mercy, UMKC, School of Medicine, Kansas City, MO, USA, [2]Creighton University, School of Medicine, Phoenix, AZ, USA.

Introduction: Steroidogenic acute regulatory (STAR) protein regulates steroid hormone synthesis by transporting cholesterol into mitochondria. *STAR* gene mutations lead to lipoid congenital adrenal hyperplasia (LCAH), the rare but most severe form of congenital adrenal hyperplasia in children. We present an unusual case with an episode of cardiac arrest in a young girl during an acute febrile illness and later she was diagnosed with adrenal insufficiency secondary to a non-classic LCAH. **Case:** 2-year 11-month-old previously healthy white female was brought to an urgent care clinic due to severe lethargy and a seizure-like activity during a fever illness. She was found to have an undetectable blood glucose level and went into cardiac arrest shortly after arrival. CPR was performed for approximately 11 minutes. She then developed sever respiratory distress and was intubated. She was transferred to the PICU with IV sodium bicarbonate given en route. On admission, her body weight was 13.26 kg (36.80^{th} percentile), height 90 cm (17.56^{th} percentile), and BMI 16.17 (62.88^{th} percentile). Her physical exam revealed normal external female genitalia and normal skin pigmentation. Lab evaluation revealed normal sodium and potassium but elevated anion gap, hyperuricemia, elevated creatinine kinase, abnormal liver function tests and abnormal coagulation profile. Brain MRI revealed findings consistent with hypoxic-ischemic encephalopathy. Renal function improved within 24 hours and hepatic function returned to normal after 20 days. Due to her severe hypoglycemic event, a high-dose ACTH stimulation test was performed. The results were consistent with adrenal insufficiency: baseline cortisol level, 7.3 µg/dL; 30 minutes cortisol, 7.8 µg/dL; 60 minutes cortisol, 9 µg/dL (normal response, ≥18 mcg/mL at 30 or 60 minutes). The baseline ACTH level was significantly elevated, 1688 pg/mL (0–46) as well as the renin activity, 24.3 ng/hour (1.7–11.2). Genetic testing revealed a 46 XX karyotype. *STAR* gene analysis identified compound heterozygosity; a novel deletion (c.811delC, p.Leu271Cysfs*50) and a previously reported missense mutation (c.661G>A, p.Gly221Ser). The girl is now 11 years old and exhibits normal growth, normal cognitive development, and she has developed early signs of puberty (Tanner stage 2 for breast). She takes daily hydrocortisone, fludrocortisone and stress dose hydrocortisone as needed. **Conclusion:** In non-classic LACH, the onset is generally late or not acute. Initial clinical features are variable and nonspecific. For this reason, non-classic LCAH may be overlooked. Adrenal crisis is a life-threatening complication, and it is important that clinicians are aware of the clinical features of non-classic LCAH and consider it in the differential diagnoses. Genetic testing for *STAR* should be considered in individuals with non-autoimmune primary adrenocortical insufficiency.

Adrenal

ADRENAL CASE REPORTS

Careful Evaluation of Cosyntropin Dose in the Diagnosis of Adrenal Insufficiency

Clio Musurakis, MD, Solab Chitrakar, MD, Randa Eldin Sharag, MD, Ekta Shrestha, MD, Gauri Pethe, MD, Faisal Qureshi, MD

Amita Health Saint Francis Hospital, Evanston, IL, USA.

Introduction: The use of the 250µg cosyntropin dose or otherwise called high-dose ACTH test is the gold standard test for diagnosis of primary adrenal insufficiency. The 1µg dose test or the low-dose test is mostly reserved for diagnosis of secondary adrenal insufficiency. Careful consideration of the results produced during the diagnostic process is imperative to avoid mislabeling of patients with a disease that requires lifelong treatment.

Case Report: This is the case of a 45-year-old female with a history of asthma and psoriasis who presented with emesis. Home medications included monthly TNF-alpha inhibitor injections for psoriasis, triamcinolone acetonide topical spray and budesonide-formoterol inhaler. On admission, she also had nausea, chills and diaphoresis, as well as palpitations, lightheadedness, and shortness of breath. When she arrived at the ER, vitals were remarkable for low blood pressure. Labs were unremarkable except for CMP concerning for anion gap metabolic acidosis, hyponatremia, and hypokalemia. A random serum cortisol was 6.4 mcg/dL, which was relatively low. ACTH was within normal range. Due to concern for adrenal insufficiency, a 1µg cosyntropin test was performed which showed a peak cortisol concentration of less than 18 mcg/dL. As the response was assessed as suboptimal, endocrinology was consulted to offer a treatment plan for steroids. However, the test was repeated using the gold standard 250µg cosyntropin dose and the patient then showed an adequate response and she was not started on steroids.

Conclusions: This is a case that demonstrates how the 250 µg ACTH or high-dose stimulation test should be used

for diagnosis of primary adrenal insufficiency (AI), as it is the gold standard. The 1 μg ACTH or low-dose stimulation test can be used for diagnosis of primary AI but only when the high dose test is not available. On the other hand, the 1 μg ACTH stimulation test has been shown to be more sensitive than the 250 μg test in diagnosing secondary adrenal insufficiency. When using the most appropriate test correctly, the clinician can only then offer the patient the best treatment strategies. Our patient did not require chronic replacement therapy. The steroids in this case could have harmed the patient as chronic administration could cause adrenal gland suppression.

Adrenal

ADRENAL CASE REPORTS

Catastrophic Presentation of a Ruptured Pheochromocytoma

Mohammad Khair Ahmad Ibraheem Hamad, MD, Salma Bashir Abdelrahman, MBBS, MRCP (UK), Mohammed AlKhatib, MD, Ibrahim Obeidat, MD, Marwa Gomaa Mokhtar, MB, BCh, Zaina Abdelhalim Alamer, MD, Dabia Hamad Al-Mohanadi, MBBS, FRCPC.
Hamad Medical Corporation, Doha, Qatar.

Background: Pheochromocytoma is a catecholamine-secreting tumor, encountered in less than 0.5% of patients with hypertension and around 4% of patients with adrenal incidentaloma.[1] It classically presents with episodic headache, sweating, and hypertension but rarely can present with serious complications such as hypertensive, pheochromocytoma crisis, and hemorrhagic shock.
Clinical Case: We report a 49-year-old male patient with a history of hypertension for five years on Amlodipine but not compliant. Presented to the Emergency Department with sudden onset left severe flank pain associated with one episode of vomiting for six hours. Vital signs were stable, and basic labs were within normal. Bedside ultrasound couldn't appreciate any abnormality due to obesity, urinary tract CT-scan showed a large heterogeneous non-enhancing mass, possibly hematoma arising from left adrenal gland. Afterward, his condition deteriorated rapidly, he became hypotensive, and hemoglobin level dropped from 14 to 8gm/dl within a few hours. He became agitated with a worsening level of consciousness, so he was admitted under the medical intensive care unit (MICU) and required intubation and started on mechanical ventilation. Abdominal CT-angiography was done to look for any active bleeding, showed re-demonstration of the same lesion in the left adrenal without contrast enhancement or extravasation. He underwent urgent explorative laparotomy that showed ruptured large left adrenal hematoma, which was entirely evacuated with the adrenal tissue, his vital signs were maintained stable during the surgery. Postoperative course showed uncontrolled blood pressure requiring three antihypertensive medications; otherwise, he had gradual improvement until successful weaning, and returned to his baseline. The histopathology result was suggestive of pheochromocytoma with extensive bleeding.
Conclusion: Ruptured pheochromocytoma is an extremely rare presentation of hemorrhagic shock and needs a high index of suspicion. In our patient, the lack of classical presentation and imaging features of pheochromocytoma combined with the condition rarity, made the diagnosis very challenging. Urgent surgical intervention may be warranted, and multidisciplinary perioperative preparation is the key to a favorable outcome.
References: [1] Bravo EL. Pheochromocytoma: new concepts and future trends. Kidney Int. 1991 Sep;40(3):544–56. doi: 10.1038/ki.1991.244. PMID: 1787652.

Adrenal

ADRENAL CASE REPORTS

Catecholamine-Producing Adrenal Schwannoma: A Rare Pheochromocytoma Imposter

Christine E. Chiu, MD[1], Trevor E. Angell, MD[2].
[1]LAC/USC MEDICAL CENTER, Los Angeles, CA, USA, [2]Keck Medicine of USC, Los Angeles, CA, USA.

Background: Adrenal schwannoma is a tumor of the adrenal medulla that poses a pre-operative diagnostic challenge due to its non-specific imaging findings and rarity. Even more uncommon are functional catecholamine-producing adrenal schwannomas (CPAS) that can lead to the clinical presentation of catecholamine excess. Here, we present a case of biochemically and histopathologically confirmed CPAS and provide a review of the existing cases in the literature.
Clinical Case: A 43-year old woman with a history of hypertension was incidentally found to have a 4 cm left adrenal lesion. Computed tomography of the mass demonstrated indeterminate enhancement and washout characteristics. Upon evaluation, she had episodes of palpitations and fatigue without headache or flushing. Her medications were notable for metoprolol, vortioxetine, and lisdexamfetamine. Laboratory results were notable for elevated 24-hour urine metanephrine and normetanephrine of 551 mcg/24hr (normal: 58–203 mcg/24hr) and 927 mcg/24hr (normal: 88–649 mcg/24hr) respectively. Due to the possibility of pheochromocytoma, the patient elected for surgery and underwent laparoscopic left adrenalectomy after pre-treatment with an alpha-1-antagonist. Pathology demonstrated a 4.8 cm well-circumscribed tumor arising from the adrenal medulla that showed mostly hypercellular areas of spindled cells with wavy nuclei and focal nuclear palisading along with hypocellular areas with loose edematous stroma with focal areas of calcification and microcyst formation. IHC of the tumor cells was diffusely positive for S-100 and SOX-10, consistent with adrenal schwannoma. Laboratory results approximately 2 months post-operative on the same medications demonstrated normalization of 24-hour urine metanephrine and normetanephrine, confirming CPAS. A search of PubMed was performed using the search terms "adrenal schwannoma," which returned 41 reports, of which 2 confirmed CPAS. Of the 3 cases, including ours, patients had a mean age of 44 years old and M:F ratio of 1:2. Each of the 3 tumors were left-sided with sizes ranging from 4.8 cm to 7.7 cm, which were all surgically removed. In the two previously published reports, the reported pre-operative laboratory markers were serum catecholamines and urine catecholamines, respectively, but neither compared levels to a reference range. In all cases, catecholamine levels normalized following surgical removal.