

with expression of other genes and *Mkrn3* decreased progressively through puberty and adult ages. **Conclusions:** Different types of loss-of-function *MKRN3* mutations were associated with premature sexual development in both sexes. Their phenotypes were relatively uniform, regardless of the mutation type. Clinical features of children with *MKRN3* mutations were similar to the idiopathic CPP group.

Adrenal

ADRENAL CASE REPORTS I

An Adrenal Incidentaloma with Extramedullary Hematopoiesis

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Introduction: Adrenal adenomas are incidentally noted during nonadrenal disease imaging at a rate up to 4%. Frequency of incidentalomas increases with age, most being adrenocortical adenomas. This case highlights an uncommon etiology of such an adrenal mass finding. **Case Description:** A 37-year-old with a history of hypertension, hypothyroidism, and tobacco use was admitted after post-operative complication. Prior to surgery, she had been experiencing right upper quadrant pain along with 70 lb weight loss and diarrhea. She otherwise had frequent palpitations. Patient had undergone cholecystectomy and common hepatic duct injury was noted. Imaging revealed a 5.3 x 3.5 cm right adrenal mass previously unknown prior to surgery. Hormonal workup was negative for overproduction of aldosterone, cortisol, DHEA-S or metanephrines. **Discussion:** Patient underwent successful resection of adrenal mass, revealing adrenal adenoma with osseous metaplasia and hematopoiesis. Extramedullary hematopoiesis is a usually discovered incidentally as in this case. Typical sites are the spleen and liver. Cases of adrenal gland manifestations have been reported in the presence of hemoglobinopathies (thalassemia, hereditary spherocytosis) or myelofibrosis. At four-month follow-up, laboratory testing on this patient didn't suggest any erythrocytic or leukocytic disorder. **Conclusion:** This case highlights an uncommon finding of hematopoiesis in an adrenal incidentaloma without any underlying hematologic defect or disease. **References:** Motta, I., Boiocchi, L., Delbini, P., Migone De Amicis, M., Cassinerio, E., Dondossola, D., Rossi, G. and Cappellini, M. D. (2016), A giant adrenal myelolipoma in a beta-thalassemia major patient: Does ineffective erythropoiesis play a role?. *Am. J. Hematol.*, 91: 1281-1282. doi:10.1002/ajh.24446. Stewart P, Newell-Price J. (2016). The Adrenal Cortex. In Melmed S, Polonsky K, Larsen P, Kronenberg H, *Williams Textbook of Endocrinology*. (13th ed., pp 489-555). Philadelphia, PA: Elsevier

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS II

Etomidate - an Under Utilized but Safe and Efficacious Drug to Treat Acute Severe Cushing's Syndrome- Case Reports of Ectopic ACTH Syndrome from Neuroendocrine Malignancies

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Background: Intravenous etomidate infusion, in non-hypnotic doses, rapidly lowers cortisol levels by blocking 11-beta hydroxylation of deoxycortisol to inhibit cortisol production. It is an underutilized drug due to concerns of excess sedation and ICU monitoring.

Clinical Case 1: A 44 year-old female with HTN, diabetes, and recently diagnosed pancreatic neuroendocrine tumor with liver metastases presented with altered mentation. Labs showed severe hypokalemia, metabolic alkalosis, serum AM cortisol >60.2 ug/dL (n 6.7-22.6 ug/dL), ACTH of 698.1 pg/mL (n 7.2-63.3 pg/mL), 24-hour urine free cortisol (UFC) of 5791 ug/24hr (n 5-64 ug/24hr), midnight salivary cortisol of 8.04 ug/dL (n <0.01-0.09), and abnormal low dose (LDDST) and high dose (HDDST) dexamethasone suppression tests each with cortisol >60.2 ug/dL. She developed worsening psychosis, likely secondary to hypercortisolism. After ICU transfer, etomidate infusion was initiated at 2.5 mg/hr and titrated upward, leading to rapid drop in cortisol levels and concomitant improvement in mentation. No respiratory or airway difficulties developed. Ketoconazole and metyrapone were started and etomidate was weaned off. Steroids were added once cortisol levels fell below 10 ug/dL as part of "block and replace." The patient eventually underwent bilateral adrenalectomy with improvement in hemodynamic and blood glucose parameters. She was discharged on physiologic replacement doses of hydrocortisone and fludrocortisone, with no reported issues two months later.

Clinical Case 2: A 51 year-old man with one month of hemochezia presented with hypertension, severe hypokalemia, metabolic alkalosis, and QTc prolongation. Colonoscopy was unremarkable; however, labs revealed a cortisol of 43.1 ug/dL, ACTH of 83.6 pg/mL, and 24-hour UFC of 7,494 ug/L, with an abnormally elevated LDDST. Imaging showed a pancreatic mass and multiple hypodense liver lesions. The overall presentation was suggestive of ectopic ACTH syndrome due to metastatic neuroendocrine carcinoma, which was confirmed on biopsy. Chemotherapy, ketoconazole, and metyrapone inadequately lowered cortisol. Etomidate drip was initiated at 3 mg/hr in the ICU, with rapid reduction in cortisol levels to <20 ug/dL without respiratory compromise. Attempts to wean off etomidate were unsuccessful and the patient underwent bilateral adrenalectomy. The surgery was compromised due to extensive liver and pancreatic enlargement, and follow up imaging revealed incomplete resection. 8am cortisol level (off etomidate) was >60.0 ug/dL. Metyrapone and ketoconazole were resumed and hydrocortisone was later initiated for "block and replace". The patient remains in critical condition.

Conclusion: Etomidate-in non- hypnotic doses is useful for the rapid lowering of cortisol levels.

Diabetes Mellitus and Glucose Metabolism

DIABETES TECHNOLOGY AND ADVANCES IN CLINICAL TRIALS

Racial Differences in Technology Use Among Type 1 Diabetes in a Safety-Net Hospital

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