[1]VANDERBILT UNIVERSITY MEDICAL CENTER, Nashville, TN, USA, [2]Vanderbilt University, Nashville, TN, USA.

Adrenal myelolipomas (AMLs) are rare benign adrenal tumors containing adipose and hematopoietic tissue, with a reported incidence of 0.08 to 0.4% on autopsy. AMLs are the second most common primary adrenal incidentaloma. Congenital adrenal hyperplasia (CAH) is associated with 10% of analyzed AML cases, half of which are bilateral in CAH patients.

This is a 40-year old male with CAH diagnosed shortly after birth, due to 21-hydroxylase deficiency. He was doing well on a maintenance dose of hydrocortisone 20mg PO qAM and 10mg PO qPM and fludrocortisone 0.2mg PO daily until two years ago when he was incidentally found to have large bilateral AMLs while undergoing abdominal MRI and CT scans. These measured 6.6x3.6x7.7cm on the right (R) and 12.3x8.4x6.8cm on the left (L) at the time. He was asymptomatic, denying flank and abdominal pain. Follow up adrenal CT a year later revealed his AMLs increased in size to 8.7x4.2x6.6cm (R) and 13.9x6x8cm (L). Repeat CT another year later showed further rapid enlargement of his AMLs, measuring 11.1x6.1x7.9cm (R) and 17.1x7.8x10.8cm (L). He also exhibited a rising 17-hydroxyprogesterone level of 11,547ng/dL, despite an increased hydrocortisone dose (20mg BID). Although he remained asymptomatic, due to the precipitous growth of the masses and his increasing steroid requirement, a surgical approach was recommended. Open bilateral adrenalectomy was performed by an experienced endocrine surgeon and patient was discharged from the hospital with maintenance hydrocortisone and fludrocortisone therapy as well as strict sick day instructions.

AMLs were first described in 1905 by Gierke. In the past, they were often discovered on autopsies, but more recently, due to the increase in imaging, have been incidentally diagnosed on more patients. Mostly, they occur unilaterally and are small (<4 cm) in size. Individuals with hormonal dysfunction such as those with Cushing's Syndrome, Conn's syndrome and CAH, particularly with difficult-to-control corticotropin levels, may be at a greater risk of developing AMLs concurrently, however this phenomenon is still not well understood. Small asymptomatic AMLs can be monitored with serial imaging over time. Spontaneous rupture of AMLs was found in 4.5% of cases, mostly occurring in tumors > 10cm, some resulting in retroperitoneal hemorrhage or even hemorrhagic shock. Although there is no clear consensus on surgery, development of symptoms or significant growth (to >10cm), as in our case, is a reason to pursue surgical evaluation especially in a young, otherwise relatively healthy adult.

Adrenal

ADRENAL CASE REPORTS

Large Invasive Adrenal Cortical Carcinoma in a Patient With Neurofibromatosis Type I

Brenda G. Sandoval, MD[1], Maria Del Mar Felix Morales, MD[1], Joy Wortham, MD[1], Maureen Koops, MD[2], Ramona Granda-Rodriguez, MD[1], Jan M. Bruder, MD[1].

[1]University of TX HSC at San Antonio, San Antonio, TX, USA, [2]Audie L. Murphy Veterans Hospital, San Antonio, TX, USA.

Background: Adrenal cortical carcinoma is a rare neoplasm. Adrenal cortical carcinoma has previously been reported in less than 10 cases of patients with Neurofibromatosis Type I. **Clinical Case:** A 44-year-old Hispanic man with Neurofibromatosis Type I (NF1) presented with progressive hypoxia, tachypnea and abdominal pain. Intubation was required with 100% FiO2. CT angiogram of the chest and abdomen showed a 9.5 cm left adrenal mass with tumor thrombus in the left adrenal vein, distal left renal vein, IVC extending into the right atrium, suggestive of an adrenocortical carcinoma. Café-au-spots, neurofibromas and Lisch nodules were seen on physical exam but no signs suggestive of a functional adrenal cortical carcinoma. Transthoracic echocardiogram demonstrated a large well circumscribed echo-dense mass filling the right atrial cavity. Hemodynamically he was unstable with labile blood pressures due to right atrium thrombus burden. He was supported pending pheochromocytoma investigation. Biochemical work up revealed an elevated 24-hour free cortisol concentration of 95.9 ug/d (<=60ug/d) with a normal 24-hour urine metanephrine and normetanephrine [140ug/d (55-320ug/d) and 448 ug/d (114-865ug/d]. Plasma free metanephrine level was normal 0.43 (0.00-0.49nmol/L) and the normetanephrine levels was increased at 2.7 (0.00-0.89nmol/L) consistent with stress response. Serum aldosterone level and renin activity with potassium and DHEA-S levels were normal. Once pheochromocytoma was ruled out, he successfully underwent a left nephrectomy, left adrenalectomy, IVC thrombectomy, right atrium thrombectomy under a multidisciplinary team of surgeons. Surgery was well tolerated and he was discharged home 11 days later. Pathology confirmed adrenal cortical carcinoma, stage 4, with involvement of endocardium and left renal vein with mitotic count greater than 20/50 high-power fields, consistent with a high-grade tumor. **Summary:** Patients with NF1 have a higher predilection to develop tumors including pheochromocytoma, paraganglioma, gastrointestinal stromal tumor, and pancreatic neuroendocrine tumor. NF1 association with adrenal tumors is well known, most commonly pheochromocytomas which occur in 0.1–5.7% of patients with NF1. However, based on a literature search there have been less than 10 case reports which postulate an association between NF1 and ACC. Genetic analysis of these reported cases suggest a loss of heterozygosity at the NF1 locus as a possible explanation of development of ACC in patients with NF1. **Conclusion:** Adrenal cortical carcinoma is rare but should be considered in a patient with NF1 and adrenal mass when plasma/urine metanephrines are not suggestive of pheochromocytoma.

Adrenal

ADRENAL CASE REPORTS

Levetiracetam Treatment Causing False Negative Screening Test in a Woman With Aldosterone Producing Adenoma

Ahmed Sawah, MD[1], abdullah mallisho, MD[1], muneera abd ulmalik alshareef, MD[2], amirah husseian alzahrani, Fellow[3].

[1]Detroit Medical Center/Wayne State University Endocrine Fellowship Program, west bloomfield, MI, USA, [2]King Fahd