ADRENAL CYSTS AND PSEUDOCYSTS

ADRENAL CYSTS AND PSEUDOCYSTS

DESCRIPTION A rare (0.064–0.18% on autopsy studies) condition, more often detected on imaging. Most are asymptomatic. These cysts can cause GI discomfort, pain if large, and even an acute abdomen with rupture or infection. Four major types are recognized: Endothelial, pseudocyst, epithelial, and parasitic, in order of decreasing incidence. Parasitic cysts arise primarily from *Equinococcus granulosus* infection. Adrenal pseudocysts are thought to result from infarction or hemorrhage of a cyst or tumor.

TREATMENT

- >3.5 cm: Aspiration for fluid analysis and cytology to rule out malignancy
- <3.5 cm: Observe with serial imaging (US or CT or MRI).

REFERENCE

Sebastiano C, Zhao X, Deng FM, et al. Cystic lesions of the adrenal gland: Our experience over the last 20 years. *Hum Pathol*. 2013;44(9):1797–1803.

ADRENAL CYTOMEGALY

DESCRIPTION Found infrequently in children and adults and considered a benign mass lesion, the condition is seen often in Beckwith–Wiedemann syndrome. Other possible associations include hemolytic disease of the newborn, erythroblastosis fetalis, and congenital rubella. It is characterized by the presence of large polyhedral cells with eosinophilic granular cytoplasm and enlarged nuclei in the adrenal cortex. Adrenal cytomegaly rarely forms cysts. This condition is thought to be a degenerative process but not a malignancy, possibly caused by a physiologic condition that demands increased functional capacity and proliferation of adenocytes.

REFERENCE

Noguchi S, Masumoto K, Taguchi T, et al. Adrenal cytomegaly: Two cases detected by prenatal diagnosis. *Asian J Surg*. 2003;26(4):234–236.

ADRENAL HEMORRHAGE

DESCRIPTION Adrenal hemorrhage (AH) is a collection of blood producing a mass effect in 1 or both adrenal glands, with or without adrenal necrosis and insufficiency. It occurs in up to 30% of selected neonatal intensive care patients, 14–22% of newborns at autopsy, and up to 15% at autopsy of adult patients dying in shock. Signs and symptoms include fever, flank or abdominal pain, tachycardia, nausea, vomiting, respiratory distress, and weakness. Unilateral AH may be an incidental finding during imaging. AH may result from multiple mechanisms: Stress, sepsis (Waterhouse–Friderichsen syndrome), anticoagulation-related hypotension, vascular spasm, adrenal venous thrombosis, or heparin-associated thrombocytopenia.

Workup may show dropping hemoglobin and electrolyte abnormalities (hyponatremia, hyperkalemia in 56% of bilateral AH).

TREATMENT

Includes replacement of fluids, electrolytes, and blood if anemia is significant. Patients should be started on steroid replacement if adrenal insufficiency is suspected. Surgical exploration may be necessary for uncontrollable hemorrhage, uncertain diagnosis, or if abscess formation is suspected.

REFERENCE

Simon DR, Palese MA. Clinical update on the management of adrenal hemorrhage. *Curr Urol Rep*. 2009;10(1):78–83.

ADRENAL HYPOPLASIA

DESCRIPTION Reduced ACTH production can result in hypoplasia of the adrenal gland (secondary adrenal hypoplasia); this can occur as a result of lack of pituitary trophic signaling, such as in pituitary agenesis. Congenital adrenal hypoplasia (primary) is an inherited disorder, with several forms identified. The major form of adrenal hypoplasia is X-chromosome linked and traced to the DAX-1 (AHCH) gene. This gene is in close proximity to other genes encoding for glycerol kinase and Duchenne muscular dystrophy (both associated with adrenal hypoplasia). Hypogonadotrophic hypogonadism is also a common finding. It typically presents in the neonatal period or with adrenal crisis (dehydration, hyponatremia, hyperkalemia, hypotension, hypoglycemia). Disorders of the external genitalia may include micropenis, undescended testes, or hypospadias. It can be detected by biochemical testing (serum cortisol, corticotropin-releasing hormone (CRH) stimulation test, etc.). Antenatal maternal estriol screening can also detect adrenal hypoplasia. Treatment is replacement of adrenal hormones.

Diagnosis must be made early, or it can be fatal secondary to salt wasting.

REFERENCE

Ferraz-de-Souza B, Achermann JC. Disorders of adrenal development. *Endocr Dev*. 2008;13:19–32.

ADRENAL INCIDENTALOMAS

DESCRIPTION Incidentally discovered adrenal lesions – called "adrenal incidentalomas" – are by-products of increased availability and use of advanced imaging. Adrenal masses are found in approximately 4% of patients undergoing abdominal CT scans, and the prevalence increases with age. Most are nonfunctional, benign adenomas. It is important to consider 2 questions in the evaluation of an adrenal incidentaloma: Whether it is functioning and whether it is malignant. Differential diagnosis includes benign nonfunctioning adenoma; cyst/pseudocyst; hormonally active tumors such as pheochromocytoma, primary hyperaldosteronism, and Cushing disease (nodular hyperplasia); myelolipoma and malignancies including adrenocortical carcinoma; or metastasis from lungs, breast, colon, kidney, melanoma, or lymphoma. Incidentaloma <4 cm are likely benign. A 1-mg dexamethasone suppression test and measurement of plasma-free metanephrines is recommended for all patients with an adrenal incidentaloma, as well as a serum potassium and plasma aldosterone concentration-plasma rennin activity ratio for patients with hypertension. (See also Section I: "Adrenal Mass.") (Image ☼)

TREATMENT

- Surgical removal is indicated with hormonally active tumors, as well as any tumors >6 cm.
- Observation is warranted for any mass <4 cm and nonfunctioning. A repeat CT 6–12 mo after the initial study is reasonable for follow-up.

REFERENCE

Grumbach MM, Biller BM, Braunstein GD, et al. Management of the clinically inapparent adrenal mass (incidentaloma). *Ann Intern Med*. 2003; 138(5):424–429.

ADRENAL METASTASES

DESCRIPTION The 4th most common site of metastatic tumor spread. Common metastases include breast (most common), lung, kidney, stomach, pancreas, and melanoma. (See also Section I: "Adrenal Mass.")

REFERENCE

Gittens PR Jr, Solish AF, Trabulsi EJ. Surgical management of metastatic disease to the adrenal gland. *Semin Oncol*. 2008;35(2):172–176.

ADRENAL MYELOLIPOMA (ADRENAL MYOLIPOMA)

DESCRIPTION Referred to as *myolipoma* and *myelolipoma* in the literature, this rare, usually nonfunctioning lesions are composed of adipose and hematopoietic cells may represent extramedullary hematopoiesis. It is rarely metabolically active (Cushing or Conn syndrome) and usually asymptomatic, except when very large or if hemorrhage occurs. They mostly occur in the adrenal glands, but extra-adrenal myelolipomas have been reported (presacral, retroperitoneum). It can be diagnosed radiographically and is more typically incidentally discovered at imaging or autopsy. Ultrasound shows a highly echogenic mass. CT demonstrates focal densities near that of fat (Hounsfield units of –30 to –115). MRI T1-weighted images demonstrate high signal intensity, whereas T2-weighted images are moderately intense. The main diagnostic similarity is well-differentiated liposarcoma. (See also Section I: "Adrenal Mass.")

TREATMENT

Excision if symptomatic or if diagnosis cannot be confirmed radiographically or on needle biopsy.

REFERENCE

Nabi J, Rafiq D, Authoy FN, et al. Incidental detection of adrenal myelolipoma: a case report and review of the literature. *Case Rep Urol*. 2013;2013:789481. doi: 10.1155/2013/789481.

ADRENAL ONCOCYTOMA

DESCRIPTION Oncocytic neoplasms of the adrenal gland, unlike that of the kidney, are rare with only 147 cases described. 80–90% of lesions are nonfunctional and only 10–20% of lesions show malignant elements. Typically occurs from 27–72 yr of age. More common in women (2.5:1 compared to men) and the left adrenal gland (3.5:1 compared to the right). Histologically, lesions are highly granular and eosinophilic due to an abundance of mitochondria. Grossly, they are large, well-rounded, and encapsulated with an average diameter of 8 cm (2–20 cm). When cross sectioned, they have a brown, yellow, or mahogany appearance. All tumors >6 cm should be excised. Percutaneous biopsy of an indeterminate mass has 73% sensitivity. Resection of

the adrenal lesion can be performed either laparoscopically or using an open technique. If benign, the prognosis is excellent; if malignant, there is a 20–35% 5-yr survival rate. (See also Section I: "Adrenal Mass.")

REFERENCE

Mearini L, Del Sordo R, Costantini E, et al. Adrenal oncocytic neoplasm: A systematic review. *Urol Int*. 2012;1–9.

ADRENALITIS

DESCRIPTION An inflammation of the adrenal gland that can lead to primary adrenal insufficiency (Addison disease), which accounts for 80% of cases. Tuberculosis is the 2nd leading cause, with the balance made up by fungal infections, hemorrhage, metastatic neoplasms, sarcoidosis, amyloidosis, and adrenal leukodystrophy. Autoimmune adrenalitis can be associated with thyroiditis, diabetes mellitus, pernicious anemia, vitiligo, hypoparathyroidism, and mucocutaneous candidiasis (autoimmune polyendocrine syndrome type 1, also known as candidiasis-hypoparathyroidism-Addison disease-syndrome), or with autoimmune polyendocrine syndrome type 2 (also known as Schmidt syndrome). HIV with opportunistic CMV adrenalitis accounts for an increasing number of cases. (See also Section I: "Adrenal insufficiency, acute (adrenal crisis) and "Addison Disease.")

TREATMENT

- Replacement of adrenal and other hormones, as necessary
- Treatment of underlying cause

REFERENCE

Perry R, Kecha O, Paquette J, et al. Primary adrenal insufficiency in children: Twenty years experience at the Sainte-Justine Hospital, Montreal. *J Clin Endocrinol Metab*. 2005;90(6):3243.

ADRENOCORTICAL DISEASE, PRIMARY PIGMENTED NODULAR

DESCRIPTION Primary pigmented nodular adrenocortical disease (PPNAD) is a rare ACTH independent form of Cushing syndrome, accounting for <1% of Cushing syndrome patients. Hypercortisolism is resistant to a dexamethasone suppression test. Typically, bilateral adrenal glands are involved with gross appearance of multiple nodules of varying sizes and pigmented colors. Histologically, the nodules are circumscribed, unencapsulated, and comprised of polygonal cells with an eosinophilic appearance. 25% of patients manifest Carney complex, which includes spotty skin pigmentation, endocrine tumors, and neuroendocrine tumors. Treatment requires bilateral adrenalectomy as unilateral and partial adrenalectomy has resulted in recurrence. (See Section I: "Cushing Disease and Syndrome.")

REFERENCE

Manipadam M, Abraham R, Sen S, et al. Primary pigmented nodular adrenocortical disease. *J Indian Assoc Pediatr Surg*. 2011;16(4):160–162.

ADRENOGENITAL SYNDROME

DESCRIPTION This is the most common cause of disorders of sexual development (DSD) (formerly ambiguous genitalia), caused by an inborn error of metabolism involving cortisol synthesis. At fault is a defect in any 1 of 5 enzymes involved in the cortisol biosynthetic pathway (21-hydroxylase, 11-hydroxylase, 3-hydroxysteroid dehydrogenase, 20.22-desmolase, or 17-hydroxylase), which may result in CAH. Usually presents with an autosomal recessive inheritance. (See also Section I: "Disorders of Sexual Development [DSD]"; Section II: "Congenital Adrenal Hyperplasia.")

SYNONYMS

- CAH
- Female pseudohermaphrodite
- Male pseudohermaphrodite

COMPLICATIONS

- For untreated females:
  - Premature pubic and axillary hair development
  - Rapid somatic maturation, premature epiphyseal closure, short adult stature
  - No breast development or menstruation until excessive androgen production is suppressed
- For untreated males:
  - Sexual and somatic precocity within 1st 2 yr of life
  - Premature epiphyseal closure, short adult stature
- Untreated males and females with salt-losing variant:
  - Progressive weight loss, dehydration within 1st few weeks of life

TREATMENT

- Early diagnosis with ascertainment of correct sex and prevention of salt wasting and metabolic consequences
- Steroid replacement with cortisone, fluorohydrocortisone as needed
- Surgical genital reconstruction may be necessary early in life, based on specific findings

REFERENCES

Newman K, Randolph J, Anderson K. The surgical management of infants and children with ambiguous genitalia. Lessons learned from 25 years. *Ann Surg*. 1992;215:644–653.

New MI, Abraham M, Yuen T, et al. An update on prenatal diagnosis and treatment of congenital adrenal hyperplasia. *Semin Reprod Med*. 2012; 30(5):396–399.

ADRENOLEUKODYSTROPHY

DESCRIPTION Rare, X-linked recessive metabolic disorder occurring in boys, and characterized by adrenal atrophy and widespread, diffuse cerebral demyelination. It produces mental deterioration, corticospinal tract dysfunction, and cortical blindness. There is lab evidence of adrenal cortical dysfunction. 2 phenotypes, with onset in childhood or young adulthood, exhibit hypogonadism. Death inevitably occurs within months of onset. A defect is theorized in peroxisomes, which handle long-chain fatty acids. *Lorenzo's oil* (a mixture of glyceryl trioleate and glyceryl trierucate oil) has been tried in this disease, with some delay in neurologic symptoms. Bone marrow transplantation is under study.

SYNONYM

Formerly Schilder disease

REFERENCE

Moser HW, Moser AB, Hollandsworth K, et al. "Lorenzo's oil" therapy for X-linked adrenoleukodystrophy: Rationale and current assessment of efficacy. *J Mol Neurosci*. 2007;33(1): 105–113.

AGING MALE SURVEY

DESCRIPTION The Aging Male Survey (AMS) is a questionnaire developed to detect hypogonadism in adult men. It has 3 domains: Psychological, Somato-vegetative, and sexual. The minimum and maximum scores are 5 and 25, respectively, for the Psychological and Sexual domains and 7 and 35 for the Somato-vegetative domain. The higher the score, the more severe the symptoms. The AMS has been shown to have a sensitivity (83%) and specificity (39%) similar to those of the shorter ADAM Survey. (See also Section I: "Andropause [Late Onset Male Hypogonadism]" and Testosterone, Decreased [Hypogonadism; Section II: "Androgen Deficiency in the Aging Male [ADAM] and ADAM Survey.")

REFERENCE

Moore C, Huebler D, Zimmermann T, et al. The Aging Males Symptom Scale (AMS) as outcome measure for treatment of androgen deficiency. *Eur Urol*. 2004;46:80–87.

AL GHORAB CORPORAL SHUNT

DESCRIPTION A surgical treatment for the management of priapism refractory to penile irrigation. A small transverse incision is made on the dorsum of the glans. A section of septum between the glans spongiosa and the corpora cavernosa is removed to create a shunt. (See also Section I: "Priapism" and Section II "Al Ghorab Corporal Shunt With Burnett "Snake" Maneuver.)

REFERENCE

Benjelloun S, el Mrini M, Aboutaieb R, et al. [Priapism. Apropos of 10 cases]. *J Urol (Paris)*. 1993;99(2): 91–93.

AL GHORAB CORPORAL SHUNT WITH BURNETT "SNAKE" MANEUVER

DESCRIPTION A modification of the Al Ghorab distal corporal-glanular shunt for priapism. The Burnett "snake" modification involves passing a 7/8 Hegar dilator through the amputated distal tips of the corpora cavernosa bilaterally. The dilator is passed to the proximal limit of the corpora cavernosum laterally on each side to avoid urethral injury. Milking of ischemic blood and clot is performed until bright red blood is visualized. A study of 10 patients with a mean follow-up of 7 mo reported that 8 men had no recurrence of priapism. Of the 6 men who had normal erectile function preoperatively, 2 had partial erectile function postoperatively.

REFERENCE

Segal R, Readal N, Pierorazio PM, et al. Corporal Burnett "snake" surgical maneuver for the treatment of ischemic priapism: Long-term followup. *J Urol*. 2013;189:1025–1029.