Vesalius SCALpel™: Adrenal

Embryology

neural crest cell migration
cortex mesoderm, medulla ectoderm and neural crest
ectopic: organ of Zuckerkandl aortic bifurcation, along aorta, broad ligament, uterus
adrenal mass: 9% of autopsies, incidentaloma on CT most benign cortical adenoma
size is best indicator of malignancy: 92% > 6cm malignant; inhomogeneous, irregular
functional adrenal mass most commonly adrenocortical neoplasm, produce cortisol
30% clinically apparent: 20% androgens, 10% estrogens, 2% aldosterone, 35% mixed
2/3 of adrenocortical carcinoma hormonally active
rapid onset Cushings syndrome with virilization
complete resection only chance of cure

Cortex

glomerulosa: aldosterone, faciculata: cotrisol, reticularis: androgens, estrogens
cortisol: stress -> hypothalamus production CRF -> ant pituitary -> ACTH -> adrenal
diurnal variation, high AM, low PM

Cushing’s
iatrogenic steroid (glucocorticoid) administration most common
syndrome: adrenal adenoma, hyperplasia or, carcinoma, ectopic, iatrogenic
ectopic most commonly (15%) small cell lung Ca, 15% primary adrenal tumor,
bronchial carcinoid, bilateral adrenal hyperplasia
hyperpigmentation most commonly from ectopic
17 hydroxyprogesterone high
disease: pituitary adenoma -> ACTH (70% source of ACTH)

differential diagnosis
24h urine free cortisol most sensitive Dx
lo dose (1 mg) dex to confirm
AM plasma cortisol suppressed < 3 normal; elevated = syndrome
corticotropin releasing hormone (CRH) elevated in disease, normal in ectopic
hi does dex suppression: suppresses pituitary adenoma, not ectopic or adrenal
plasma ACTH differentiates pituitary/ectopic source from adrenal
if other tests inconclusive, inferior petrosal sinus sampling differentiates source
ACTH
aldosterone
decrease in blood pressure stimulates release renin from renal tubules -> liver where
angiotensinogen is converted to angiotensin 1, lung enzyme converts to angiotensin
2, potent vasoconstrictor, stimulates adrenal release of aldosterone which causes
tubular reabsorption Na in exchange for K and H, water reabsorption, increased
vascular volume
aldosteronoma 1% of hypertensive patients
imaging may miss bilateral micronodular disease, idiopathic hyperaldosteronism
if miss on CT 60% bilateral hypersecretion
unilateral adrenalectomy not effective
venous sampling can differentiate
increased BP, hypokalemia, alkalosis, feedback decreases renin
Dx: decreased K (necessary for diagnosis), increased plasma/urine aldosterone,
suppressed plasma rennin (aldosterone/renin ratio), increased Na,
alderosteronoma 65%, idiopathic 35%, rare adrenal carcinoma
adenoma: younger, women, more severe hypertension, hypokalemia
salt loading and postural test no change in aldosterone
CT small,< 2cm, solitary (70%), solitary cortical adenoma, 30%
hyperplasia, rare Ca
primary see decreased plasma rennin
idiopathic: male, older
salt loading drops aldosterone
postural test aldosterone increases
medical Rx: K-sparing diuretics (spironolactone most effective)
secondary hyperaldosteronism from hypovolemia
Na load, captopril decrease aldosterone
spironolactone, ACE inhibitors, diuretics affect aldost measure: withhold
adrenalectomy cures 80% of hypertension, 90% normalization of K
bilateral hyperplasia treated with spironolactone
androgens
<5% of male testosterone
increased in Cushings
congenital adrenal hyperplasia
autosomal recessive defect in cortisol synthesis (21 alphahydroxylase defic.)
causes increase ACTH production -> adrenal hyperplasia -> incr androgen
associated defect aldosterone production, severe electrolyte and fluid loss
adrenal insufficiency
etiologies: cortisol administration, autoimmune, bilateral adrenal hemorrhage,
(Waterhouse-Friedrichson), heparin induced, adrenalectomy, adrenal mets, infection

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symptoms: vascular collapse (no aldosterone, fluid loss), abdominal pain, nausea, weakness, fever, decreased mentation

adrenal crisis, hypotension, responds to cortisol

lab: hyponatremia, hyperkalemia, hypoglycemia (no cortisol), increase BUN/Cr (vol loss), decreased glucocorticoid level

stress dose for surgery in chronic hypothalamus/pit/adrenal axis

major: 100-150mg/d hydrocortisone X 2-3d
(older recommendation 300-400 excessive, unnecessary)

minor surgery: (hernia) 25mg/d, moderate 50-75mg X 1-2d
hydrocortisone short half life: 3 doses/d
no taper necessary

3/4 of septic patients who respond poorly to fluid resuscitation have relative adrenal insufficiency
df: failure to increase cortisol level by at least 9mcg/dl in response to ACTH

supplement with 200mg/d plus mineralocorticoids, benefits sepsis Rx

Medulla

pheochromocytoma

24h urinr VMA, metanephrine (serum catechols elevated in essential HTN)
90% adrenal medullary tumor, 3X as bright as liver on T2 MRI image
MIBG (methyiodobenzylguanidine: norepi analogue) scan for primary sporadic, extra-adrenal, low sensitivity, rarely identifies other sites or changes plan: no advantage over MRI
rule of 10s: 10% extraadrenal, bilateral, child, familial, malignant
most sporadic unilateral
many congenital (MENIIA, B, VonRecklinhausen) bilateral
laparoscopic adrenalectomy for pheo: risk of seeding malignant?
alpha blockade (penoxybenzamine = major alpha blockade; now prefer selective
alpha 1 antagonist [prazosin] or calcium channel blocker) pre-op functional
tumor, subsequent beta blockade if still tachycardic after alpha (no early beta:
chronically volume contracted due to alpha stimulation, beta could cause hypotension), proportional to initial symptoms
pheo during pregnancy: untreated 50% maternal mortality, higher fetal mortality
hypertension, sweating, tachycardia may be mistaken for pre-eclampsia
urine protein elevated in pre-eclampsia, not pheo
treat with alpha block and adrenalectomy first and second trimester
third trimester immediate alpha block (decreased fetal mortality), then controlled C-section
vaginal delivery absolutely contraindicated (BP instability, abruptio)

MS and phenothiazine can precipitate pheo crisis

Incidentaloma

41% benign adenoma, 19% metastasis, 10% adrenocortical malignancy, 9% myelolipoma, 8% pheo
check aldosterone/renin, corticoids, catechols
adrenal met
FNA only of other malignancy present, suspect met and would change plan: 75% positive lymphoma, lung, breast common; also melanoma, renal, leukemia, ovarian
resection of solitary adrenal met can increase survival

References: