

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 Cushing's syndrome with virilization

complete resection only chance of cure

Cortex

glomerulosa: aldosterone, fasciculata: cortisol, reticularis: androgens, estrogens

cortisol: stress -> hypothalamus production CRF -> ant pituitary -> ACTH -> adrenal

diurnal variation, high AM, low PM

glucocorticoids

stimulate hepatic gluconeogenesis

inhibit protein synthesis (inhibit wound healing)

inhibit fibroblast activity (inhibit wound healing)

lipolysis

increase response to gluconeogenic hormones

increase glycogen synthesis

increase uptake glucose

inhibit bone formation (osteoporosis)

negative calcium balance

anti-inflammatory

inhibit leukocyte mobilization

decrease migration of inflammatory cells

decrease production inflammatory mediators (interleukins, leukotrienes,

bradykinins)

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

low 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,

aldosteronoma 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 alpha hydroxylase def.)

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

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 (methyoiodobenzylguanidine: 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:

Pasieka J. What's new in general surgery: endocrine surgery. JACS 199(3), Sept. '04: 437-445.