Adrenal Pathology
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Adrenal Path Lecture Outline

- Introduction
- A disease with too much hormone: Cushing syndrome
- A disease with too little hormone: Addison disease
- Tumors of the adrenal

Adrenal gland anatomy

Adrenal gland histology and hormones
Cushing Syndrome

- Elevated serum cortisol
- Clinical features:
  - Hypertension
  - Characteristic pattern of weight gain
  - Hyperglycemia
  - Thin skin
  - Depression
- Most common cause: exogenous steroids

Cushing syndrome: buffalo hump

Cushing syndrome: moon facies

Causes of Cushing syndrome

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Addison Disease

- Also called primary chronic adrenal insufficiency
- Decreased serum cortisol and mineralocorticoids
- Most common cause: autoimmune attack on adrenal cortex
- Eventually fatal if not treated
Symptoms of Addison Disease

- Early: signs are vague (weakness, fatigue)
- Later: skin bronzing/hyperpigmentation
- Eventually: serious complications (hypotension, electrolyte imbalances)
- Misdiagnosis is not uncommon

Why is there hyperpigmentation in Addison disease?

Cortisol ▼
ACTH
↓ Cortisol

Normal
Addison Disease

33 year-old female with no prior medical issues.

Day 1 prior to admission
- Headache

Day 2 prior to admission
- Very fatigued, vomiting

Day of admission
- Husband brought to ER
- Vision loss right eye
- BP 100/60
- Given IV fluids

Day 2 post admission
- Right-sided paralysis
- Oxygen levels dropping
- CT: brain swelling

Day 3 post admission
- Coma
- BP 70/33 despite IV fluids
- 40 pound weight gain

Day 4 post admission
- No brain activity
- Acute renal failure
- “Nothing else can be done.”

Day 5 post admission
- Different doctor
- IV Na+, hydrocortisone, dextrose

Day 6 post admission
- Opened eyes

Day 10 post admission
- IV removed
In ensuing weeks
• Feeding tube removed
• Intensive speech, physical, occupational therapy

Two months later
• Back to work
• Feeling normal
• Must take steroids daily for the rest of her life

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Pheochromocytoma

• Derived from adrenal medulla cells
• Cells produce epinephrine
• Patients present with hypertension
• Usually benign
• Diagnosis: epinephrine breakdown products in urine

A couple good pheo mnemonics

The 10% tumor
• 10% occur outside the adrenal
• 10% bilateral
• 10% familial
• 10% malignant

Pheo symptoms
• Paroxysms of...
• Pressure (hypertension)
• Perspiration
• Palpitations (tachycardia)
• Pallor

Neuroblastoma

• Derived from neural crest cells
• Relatively common childhood tumor
• Prognosis better in:
  • Children < 18 months
  • Lower stage/grade
  • Hyperdiploid tumors
  • Fewer copies of \( N\)-\( myc \)

MEN Syndromes
Brad Pitt vs. John Cleese
MEN Syndromes

- Inherited disorders that predispose patients to getting endocrine tumors
- MEN-1 and MEN-2 (A and B)
- MEN tumors are much more aggressive than sporadic tumors

MEN Syndromes Ric dulously Simplified

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MEN 2A Genetics

- RET gene mutation
- Proto-oncogene
- Encodes tyrosine kinase receptor
- Mutation turns gene on

MEN 2B

2B has most of the features of MEN 2A
- Medullary thyroid carcinoma
- Pheochromocytoma
- RET gene mutation

But it's slightly different
- No parathyroid involvement
- Patients are “marfanoid”

MEN-2

Cleese-cell hyperplasia
bRETone gene
one of a kind
always turned on