

Med C PCC

Mike Simonson PGY 3

Jordan Lockhart PGY 2

Jedi Master Joey Jose PGY 1

Adam Woodyard PGY 1

Riju Dasgupta PGY 1

Clinical Case “I felt a Disturbance in the Force” - Obi Wan

Case: 55 year old AA female

Smoldering MM, Hep C, hx tobacco use, HTN, CAD, seizures, TI

P/W: Painful, Worsening skin lesions

HPI: Diffuse rash continued to increase in number and discomfort. Lesions throughout body draining and painful to touch. Pinching sensation, but non-pruritic. No aggravating or alleviating factors. Denies new soaps, sick contacts, hx of similar rash, med changes, animal exposures and insect bites.

Previous workup: G/C, RPR, VZV, HSV, ANCA, blood and wound cx all negative

Bx of lesion



ED Course “Do. Or do not. There is no try” - Yoda

ED: Significant Electrolyte Derangements/acidosis and pain

Initially mildly tachycardic and hypertensive

BMP notable for K 2.7, CO₂ 13, AG 18. Given oral and IV K. Given dose of vanc and zosyn. Also given morphine, then norco for pain control

UNINTENTIONAL tylenol overdose- N-acetylcysteine

Admitted for pain control and continued disease management



ROS “I find your lack of faith disturbing” - Darth Vader

Complains of inguinal/vulvar lesions. Lesions on arms, chest, legs, back

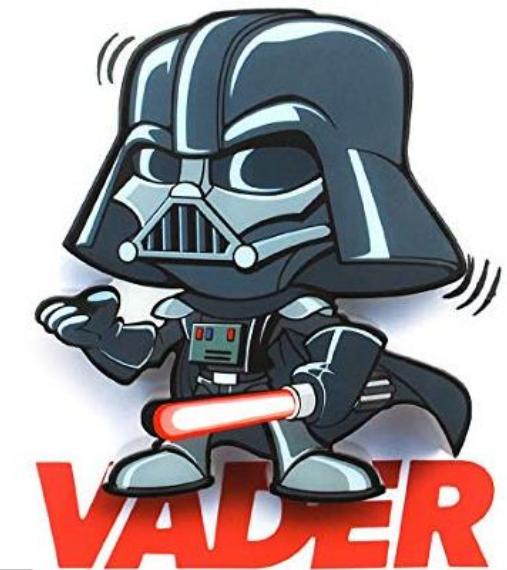
Increasing dyspnea w/ URI sx, chronic cough sputum change from clear to yellow/green

Chest heaviness, resolves w/ rest, lasts 15-30min, assoc w/ diaphoresis/N

Dysuria and abdominal pain related to lesions

Denies oral/eye lesions, difficulty breathing/swallowing

Denies emesis, drug/EtOH use for >10 years, bleeding issues



Physical Exam "In my experience, there is no such thing as luck" -Obi Wan

Vitals: **BP 149/85** **Pulse 121** **Temp 98.8 °F (37.1 °C) (Temporal)** **Resp 18**
Ht 5' 1" **Wt 140 lb (63.5 kg)** **SpO2 98%** **BMI 26.45 kg/m²**

Extensive nodular lesions with central umbilication or ulceration. Located throughout upper/lower extremities including hands/feet as well as scattered lesions on thorax/abd and extensive lesions in inguinal folds and outer labia. Some present with bloody or yellow drainage. Lesions anywhere from 0.5cm-3.5cm in diameter. Extremely tender to palpation

PE otherwise unremarkable

Physical Exam (pictures) “I’ve got a bad feeling about this” -...basically everyone



Hospital Course “Stay on Target” -Gold Five

Patient started initially on topical steroids, transitioned to oral prednisone for suspected *insert disease that shall not be named yet*. Recent bx (at previous workup) of lesion with neutrophilic infiltrates w/out leukocytoclastic vasculitis. HAGMA resolved with N-acetylcysteine. NAGMA likely related to underlying RTA. ESR, CRP and Procalcitonin elevated. Heme/onc following but deferred to outpatient for tx of MM. Nephro consult, concern for RTA (likely acquired) given hypok, acid-base abn, and failure to acidify urine in distal tubule. Started D5W w/ bicarb, aldactone was switched to amiloride. TTKG 12, K+ wasting. Electrolytes stabilized and lesions appeared smaller/less painful with steroids. Follow up with derm/rheum...and Jordan.

Sweet Syndrome

Neutrophilic dermatoses

- Histology containing neutrophilic infiltrate
- No signs of infection (of the affected area) or true vasculitis

Acute febrile neutrophilic dermatosis

- Rash formed of erythematous pustules or plaques
- Dermal neutrophilic infiltrate
- Fever
- Neutrophilia

Subtypes and Pathogenesis

Subtypes of Sweet Syndrome

- Classical (idiopathic, can be seen in association with infection, IBD, pregnancy)
- Malignancy-associated (more common in hematologic cancers, esp AML)
- Drug-induced (Granulocyte-colony stimulating factor, G-CSF most common)

Pathogenesis

- Hypersensitivity
- Cytokine dysregulation
- Genetic

Symptoms/Findings

Systemic Symptoms

- Fever
- Malaise
- Myalgia
- Arthralgia
- Headache

Lab Findings

- Leukocytosis
- Elevated ESR

Rash

- Erythematous, tender papules
- Plaques with vesicular surfaces
- Painful and burning, not pruritic
- Less common manifestations:
 - Bullous
 - Subcutaneous
 - Neutrophilic dermatosis of the dorsal hands

Sweet Syndrome Rashes



© 2010 Logical Images, Inc.



© 2012 Logical Images, Inc.



© 2012 Logical Images, Inc.

Diagnosis: 2/2 major and 2/4 minor criteria

Major Criteria

- Acute onset of erythematous, painful nodules/plaques
- Neutrophilic infiltrate on biopsy (with no evidence of vasculitis)

Minor Criteria

- Fever
- Association with underlying malignancy, inflammatory disease, or pregnancy OR preceded by GI infection, URI, or vaccine
- Excellent response to systemic steroids or potassium iodide
- Abnormal lab values: (3 of 4 of: ESR >20, elevated CRP, WBC >8000, >70% neutrophils)

Differentials

Other causes of rash

- Cutaneous infection
- Vasculitis
- Urticaria
- Drug eruptions
- Subcutaneous sarcoidosis
- Erythema nodosum

Other neutrophilic dermatoses

- Pyoderma gangrenosum
- Behcet's Syndrome
- Pustular psoriasis
- Reactive arthritis (esp when lesions in the genital area)
- And more!

Treatment

First line

- Systemic glucocorticoids: oral prednisone at 0.5 to 1 mg/kg daily
 - Within 48 hours: symptoms improve
 - Within 1-2 weeks: skin lesions resolve
 - Once disease controlled, taper over 4-6 weeks
 - In cases where disease recurs with tapering prednisone, 10 mg/day may be continued for 2-3 months
- Alternatives:
 - Colchicine (1 to 1.5 mg/day for average 15 days, usually rapid response)
 - Dapsone (begin at 25 mg/day then uptitrate to 100-200 mg/day for 1-3 week course)
 - Potassium iodide (supported by case series)

Treatment: Refractory disease

Pulse glucocorticoid therapy

High-dose methylprednisolone (IV 500 to 1000 mg per day for 3-5 days)

Methotrexate, cyclosporin, oral retinoids

Rituximab, Adalimumab

Azacitidine has been shown to be effective in treating refractory disease in patients with MDS

The Force?

Prognosis

Spontaneous resolution may occur in weeks - months

Lesions typically heal without scarring but may leave hyperpigmentation that resides for months

Recurrence

- 30% in patients with Classical Sweet Syndrome
- Up to 69% in patients with underlying hematologic malignancies

Question 11 ★

This patient is a febrile, middle-aged woman who was recently diagnosed with acute myeloid leukemia.



MKSAP question

MKSAP question

Question 11 ★

This patient is a febrile, middle-aged woman who was recently diagnosed with acute myeloid leukemia.



Which of the following is the most likely diagnosis for the skin findings shown?

- A Acute febrile neutrophilic dermatosis (Sweet syndrome)
- B Erythema nodosum
- C *Mycobacterium marinum* infection
- D *Pseudomonas* folliculitis
- E Pyogenic granuloma

Answer

Which of the following is the most likely diagnosis for the skin findings shown?

73%

A

Acute febrile neutrophilic dermatosis (Sweet syndrome)

10%

B

Erythema nodosum

1%

C

Mycobacterium marinum infection

11%

D

Pseudomonas folliculitis

5%

E

Pyogenic granuloma

References

- Star Wars
- UpToDate
- MKSAP

GI PCC

Indraneel Reddy
Davis Mann

HPI

- 24 y/o M with previous history of epistaxis, HTN, tobacco abuse, and EtOH abuse w/ EtOH hepatitis
- CC: epigastric abdominal pain with nausea and non-bloody vomiting for 1 day

ROS and more history

- No fever, chills, or sweats. **Decreased appetite.** No HA, visual disturbance, eye pain, jaundice, sore throat or mouth ulcers. No skin rash or itching. No chest pain. **Mild shortness of breath, dyspnea on exertion,** cough or wheeze. No urinary frequency, urgency, hematuria, or dysuria. No myalgia, arthralgia, or joint swelling. No numbness, or confusion **but endorsed dizziness and weakness.** **Abdominal pain, N/V,** no constipation, diarrhea No polyuria, polydipsia, heat or cold intolerance.
- Less than $\frac{1}{2}$ ppd, denied illicit drug use
- Of note, he drank a $\frac{1}{2}$ -1 750 ml bottle of vodka daily for 3 years, last drink was before pain started
- No surgical history, no relevant family history

In ED

- Vitals: BP 99/62, T 97.7, HR 129, RR 18, O₂ 97%
- Labs showed K 2.5, Cr 2.51, Anion gap 37, Bicarb 15, Mag 1.1, BG 201, Phos 5.9, AST 315, ALT 52, alk Phos 132, Bilirubin 9.7, lipase 5804, Beta hydroxybutyrate 25.70, ETOH level 0.345, platelet 58, pH 7.292
- CT and U/S abdomen showed findings suggestive of acute pancreatitis
- Physical exam: Diffuse tenderness in abdomen with distention, icteric sclera
- While there, patient was given 2 liters NS, pepcid, zofran, morphine, folate,, and thiamine
- Patient was admitted to ICU for pancreatitis, 2/2 to alcohol abuse and GI was consulted for alcoholic hepatitis and pancreatitis

GI Consult

- Consulted for pancreatitis and alcoholic hepatitis
- After seeing him, ordered PT/INR, and patient was started on solu-medrol 40 mg for acute on chronic alcoholic hepatitis
- Patient acutely worsened and needed intubation overnight
- MELD score 30, Maddrey's discriminant factor 36.9
- At this point, liver transplant became a possibility
- After 3 days extubated, but LFTs were up and down
- After 7 days of steroids, Lillie model score was 0.025, and steroids were planned for a full 28 day course

Post consult

- GI signed off after
- Latest values: AST 164, ALT 108, Alk phos 189, Bilirubin 7.5, INR 1.4
- MELD 16 and Discriminant 16.2
- Still in ICU, currently on PO prednisolone 40 mg

Alcohol-Induced Liver Disease

- 2nd most common reason for liver transplantation
- 25% of heavy drinkers develop cirrhosis
 - 100g alcohol daily for 20+ years
 - 10g in 12oz beer
- Increased MCV
- AST:ALT at 2:1 ratio, <500
- Elevated GGTP
- Advanced cases: elevated INR and thrombocytopenia

Alcoholic Hepatitis

- Fever, Jaundice, Tender hepatomegaly, leukocytosis
- Clinical diagnosis
- Severity
 - Maddrey Discriminant Factor Score
 - $= 4.6 \times (\text{PT} - \text{Control PT}) + \text{Total Bilirubin}$
 - Severe > 32, or presence of hepatic encephalopathy
 - Prednisolone
 - Pentoxifylline
- Lillie Model: risk stratification calculator for patients receiving steroids to predict which will not improve and should be considered for alternative therapy, predicts mortality at 6 months
 - Scores >0.45 predict 6 month survival of 25% , <0.45 predicts a 6 month survival of 85%

STOPAH

- Prospective multicenter randomized, double-blind trial, 2015, >1000 patients
 - 2x2 Factorial Trial
- Primary Outcome: All cause mortality at 28 days
- Neither Prednisolone or Pentoxifylline reduced all cause mortality at 28 days
 - 2011 Meta-analysis showed a mortality benefit with Prednisolone (46%)
 - 2000 Single Trial, Pentoxifylline in Severe Alcoholic Hepatitis, showed improved mortality

A 50-year-old man is seen for preoperative medical evaluation before left shoulder arthroplasty. History is significant for alcohol-related cirrhosis and osteoarthritis. Medications are lactulose, furosemide, and spironolactone. The patient stopped drinking alcohol 5 months ago but has difficulty with medication adherence. He reports increasing ascites and lower extremity edema.

On physical examination, vital signs are normal. There is no jaundice or scleral icterus. Spider telangiectasias are noted on the face and chest. The abdomen is distended with flank dullness. There is 1+ pitting edema to the knees bilaterally. Mental status examination is normal.

The calculated Model for End-stage Liver Disease (MELD) score is 22.

The patient is instructed to increase his furosemide.

Which of the following is the most appropriate preoperative management?

- A Cancel surgery and refer for liver transplant evaluation
- B Delay surgery until after patient achieves 1 year of sobriety
- C Delay surgery until after placement of a transjugular intrahepatic portosystemic shunt
- D Proceed to surgery

Which of the following is the most appropriate preoperative management?

- 41%  A Cancel surgery and refer for liver transplant evaluation
- 8%  B Delay surgery until after patient achieves 1 year of sobriety
- 18%  C Delay surgery until after placement of a transjugular intrahepatic portosystemic shunt
- 34%  D Proceed to surgery

References

- MKSapp 18
- Medscape
- Uptodate
- Thursz MR, et al. “Prednisolone or pentoxifylline for alcoholic hepatitis”. The New England Journal of Medicine. 2015. 372(17):1619-1628





Endocrinology PCC



**Jared Kusar PGY I
Branavan Ragunanthan PGY I
August 20th, 2019 Noon Conference**

Patient History

HPI: 47 year old African American male presents to the ED with dizziness.

PMHx: type 2 DM, neuropathy, gastroparesis, anxiety disorder, major depression with h/o suicide attempt, secondary AI, pituitary microadenoma (5mm on MRI 1/19), thyroglossal duct cyst

ROS:pertinent positives: fatigue, weakness, anorexia, orthostatic hypotension, nausea

Meds: Effexor, Admelog, Zofran, Cortef, lantus, humalog Reglan

PSHx: Excision of external laryngocele right side

Social: denies tobacco,or Alcohol use. Endorses Marijuana use. Maternal Grandmother DM2

Vitals: BP 152/99, Pulse 96, T 97.2 F, RR18, wt 141lb, BMI 22

Physical Exam: Thin male, awake, alert, not in distress; note of scar on R upper neck, no thyromegaly, no LAD, RRR, no murmur, intact pulses, clear breath sounds, no rales, no wheezes, abdomen soft, non-tender, no masses, no open skin lesions, no edema, no focal deficits, flat affect, memory is mildly impaired.

Labs: BMP wnl, A1c 8.7, Cortisol 19.3, Prolactin 104.8, TSH 1.404

Imaging: MRI Brain W/ WO contrast: Findings: Dynamic postcontrast imaging confirms the presence of an area of circumscribed, relative hypoperfusion in the right anterior pituitary gland, best appreciated on series 1401, image 68. This area measures approximately 5 x 5.5 x 4 mm (AP x transverse x craniocaudal), similar dimensions to the previous examination.

Assessment and Plan

For his AI:

- his presenting symptoms are possibly related to AI
- recent cortisol level of 19.3 done @1314 would not be helpful since pt has already received steroid a few hrs prior
- given his n/v, will switch to IV steroids and gave stress doses
- started Hydrocortisone 30-10 mg BID IV
- switch back to oral hydrocortisone @ 15-5 BID once more stable

For his pituitary microadenoma:

- TSH would not be helpful in the setting of pituitary dysfunction; will check free T4
 - elevated PRL could be related to metoclopramide; will DC metoclopramide for now and repeat PRL on 8/19
 - check testosterone outpatient
- repeat prolactin after holding reglan 104.8->8.6 (ref 3.7 - 17.9 ng/mL)

f/u outpatient free T4 pending, ACTH pending, testosterone pending,

Primary Adrenal Insufficiency

Clinical Presentation:

- Fatigue, weakness, anorexia/weight loss, salt craving
- Low serum cortisol levels with markedly elevated ACTH levels
- Hypotension (volume depletion due to aldosterone deficiency)
- Hyperpigmentation (due to increased ACTH secretion with resultant increased melanocyte stimulating hormone levels)
- Hyponatremia
- Hyperkalemia (due to mineralocorticoid deficiency)
- Eosinophilia (unclear, possibly due to effects on bone marrow and on cell migration)
- GI symptoms

Primary Adrenal Insufficiency

Common causes of PAI:

- Autoimmune adrenalitis (responsible for greater than 80% of cases)
 - In addition, approximately 50% of patients with autoimmune PAI have other autoimmune diseases involving other endocrine glands
 - Number of infections can cause PAI including CMV, fungal, and TB
 - Malignant infiltration
 - Patients can have adrenal hemorrhage with acute stress (sepsis)
 - Hemorrhagic necrosis of adrenal glands seen in meningococcemia or pseudomonas

Secondary Adrenal Insufficiency

Clinical presentation:

- Fatigue
- Anorexia
- Hypoglycemia
- Eosinophilia
- Pale skin
- Hypercalcemia
- No Hyperkalemia (intact RAAS)

Secondary Adrenal Insufficiency

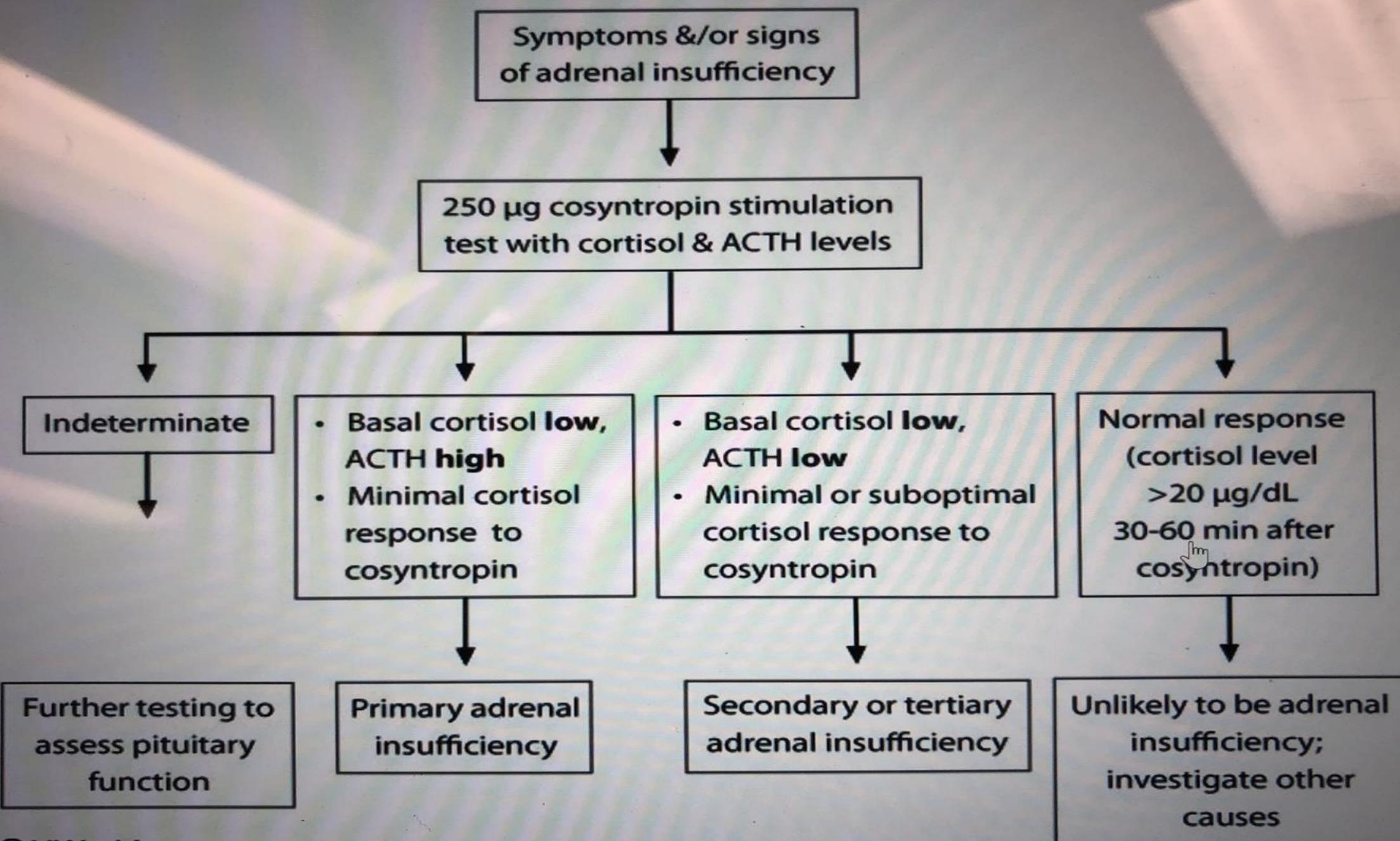
Etiologies (any process that interferes with ACTH secretion)

- Panhypopituitarism - pituitary tumors, infectious diseases such as TB or histo, head trauma, sheehan syndrome, pituitary apoplexy, mets.
- Isolated ACTH deficiency - Autoimmune induced, genetic induced

Diagnostic Workup

Patients in shock:

- Check random cortisol assay
- Give IV fluids and dexamethasone
- Perform ACTH stimulation test after patient stabilizes.



Treatment

- Long-term glucocorticoid therapy
- Will need mineralocorticoid replacements with fludrocortisone if Primary AI is diagnosed

MKSAP Q49

A 65 yo F is admitted to the hospital for fatigue and weakness over the last 1 to 2 weeks. Medical history is significant for HTN, T 2 DM, and RA. For the past 3 months, the patient's RA has been treated with MTX and Prednisone. Because of inadequate control, etanercept was added 2 weeks ago. At that time, the patient decided to D/C prednisone due to bruising. Current medications are MTX, etanercept, amlodipine, folic acid, metformin, and ASA.

PE: BP = 110/68 sitting and 90/64 standing. Pulse is 102 sitting and 110 standing. Symmetrical synovial bogginess is noted in the MCP joints and wrists bilaterally.

Studies show 8AM cortisol of 2ug/dl (normal = 5-25)

MKSAP Q49

A 65 yo F is admitted to the hospital for fatigue and weakness over the last 1 to 2 weeks. Medical history is significant for HTN, T 2 DM, and RA. For the past 3 months, the patient's RA has been treated with MTX and Prednisone. Because of inadequate control, etanercept was added 2 weeks ago. At that time, the patient decided to D/C prednisone due to bruising. Current medications are MTX, etanercept, amlodipine, folic acid, metformin, and ASA.

PE: BP = 110/68 sitting and 90/64 standing. Pulse is 102 sitting and 110 standing. Symmetrical synovial bogginess is noted in the MCP joints and wrists bilaterally.

Studies show 8AM cortisol of 2ug/dl (normal = 5-25)

Which of the following is the most appropriate management?

- A. Initiation of Hydrocortisone**
- B. Initiation of Hydrocortisone and fludrocortisone**
- C. Performance of an ACTH stimulation test
dexamethasone**
- D. Performance of an ACTH stimulation test after**

MKSAP Q49

Correct Answer is A

- Patient has secondary adrenal insufficiency.
- PO and occasionally topical glucocorticoids can suppress ACTH secretion.
- Glucocorticoids prescribed at doses above physiologic replacement for longer than 3 weeks should be tapered when discontinued allowing recover of Pituitary-adrenal axis
- Diagnosis of Adrenal insufficiency is based on demonstrating low serum cortisol levels.
- **Choice B:** Fludrocortisone is needed only in primary adrenal insufficiency. There is no mineralocorticoid deficiency in secondary adrenal insufficiency
- **Choice C and D:** ACTH not necessary since cortisol level less than 3 is diagnostic of adrenal insufficiency.

MKSAP Q5

A 22 yo F is evaluated for loss of appetite and fatigue and 4.5 kg weight loss over the past 6 mo. Medical history is otherwise unremarkable and she takes no medications.

On PE, BP = 110/70 and HR = 94. Other VS are normal. BMI = 22. Patient looks tanned, even in areas not exposed to the sun, and has buccal and palmar hyperpigmentation.

K+ = 5.3 mEq/L (3.5 - 5.0)

Na+ = 134 mEq/L (136-145)

ACTH = 650 pg/ml (9-52)

Cortisol = 2.8 ug/dl (5-25)

MKSAP Q5

A 22 yo F is evaluated for loss of appetite and fatigue and 4.5 kg weight loss over the past 6 mo. Medical history is otherwise unremarkable and she takes no medications.

On PE, BP = 110/70 and HR = 94. Other VS are normal. BMI = 22. Patient looks tanned, even in areas not exposed to the sun, and has buccal and palmar hyperpigmentation.

K+ = 5.3 mEq/L (3.5 - 5.0)

Na+ = 134 mEq/L (136-145)

ACTH = 650 (9-52)

Cortisol = 2.8 ug/dl (5-25)\

Which of the following is the most appropriate treatment?

- A. Dexamethasone BID**
- B. Hydrocortisone BID**
- C. Hydrocortisone BID and Fludrocortisone once daily**
- D. Prednisone BID**
- E. Prednisone BID and fludrocortisone once daily**

MKSAP Q5

Correct Answer is C

- Treatment of primary adrenal insufficiency. Patient has all layers of adrenal cortex affected and thus requires both glucocorticoid and mineralocorticoid therapy. Primary adrenal insufficiency is confirmed by low cortisol and high ACTH levels.
- Preferred glucocorticoid for adrenal insufficiency is hydrocortisone because of shorter duration of action and can be prescribed BID or TID to mimic circadian rhythm. Prednisone once daily may be substituted for patients having difficulty adhering to BID regimen (**Answer choice E**).
- **Answer Choice A:** Dexamethasone has long duration of action with potential to cause comorbidities from excess steroid exposure
- **Answer Choice B and D:** Only glucocorticoid therapy is required for treatment of secondary adrenal insufficiency. Fludrocortisone replacement not required as aldosterone secretion is not under the control of ACTH.

Key Points

- The most common cause of adrenocorticotrophic hormone deficiency is iatrogenic following administration of exogenous glucocorticoids and suppression of adrenocorticotrophic hormone production.
- Glucocorticoids prescribed in greater than physiologic doses for 3 weeks or longer should be tapered off to allow recovery of the pituitary-adrenal axis.
- Glucocorticoid dosing must be adjusted in the setting of physiologic stress or acute illness.