

# Morning Report

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March 23, 2009



Wake Forest University Baptist  
**MEDICAL CENTER**®

# MKSAP: Question #1

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- A 55-year-old man is brought to the office by his wife for evaluation of deteriorating job performance. Over the past 3 years he has become increasingly insensitive to his family's needs, and rarely takes on projects around the house. His mother and a maternal uncle had dementia.
- On examination, he is alert and recalls 3 of 3 words after a delay. His score on Mini-Mental State Examination is 29/30, and his 1-minute verbal fluency score is 6 items. The rest of the neurologic examination is normal.

# MKSAP: Question #1

## Frontotemporal Dementia

- Which of the following diagnostic tests is most appropriate in this patient's evaluation?
  - A. Analysis of CSF for 14-3-3 protein
  - B. MRI of the brain
  - C. Electroencephalography
  - D. PET scan of the brain
  - E. Analysis of blood for presenilin-1 mutation

# Frontotemporal Dementia

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- Characteristic Findings:
  - Personality change
  - Lost initiative
  - Slowing of thought
  - Relative preservation of recent memory
- MRI:
  - Disproportionate atrophy of the anterior frontal and temporal lobes

# MKSAP: Question #1

## Frontotemporal Dementia

- A **55-year-old man** is brought to the office by his wife for evaluation of **deteriorating job performance**. Over the past **3 years** he has become **increasingly insensitive to his family's needs, and rarely takes on projects around the house**. His mother and a maternal uncle had dementia.
- On examination, he is alert and **recalls 3 of 3 words after a delay**. His score on Mini-Mental State Examination is **29/30**, and his **1-minute verbal fluency score is 6 items**. **The rest of the neurologic examination is normal**.

# MKSAP: Question #1

## Frontotemporal Dementia

- Which of the following diagnostic tests is most appropriate in this patient's evaluation?
  - A. Analysis of CSF for 14-3-3 protein
    - CJD is rapidly progressive (weeks / months)
  - B. MRI of the brain
    - To look for atrophy in frontal / temporal lobes
  - C. Electroencephalography
    - Normal / nonspecific mild slowing in degenerative dementias
  - D. PET scan of the brain
    - Can differentiate b/t FTD and Alzheimer's when pts meet criteria for both; consider AFTER structural imaging (MRI)
  - E. Analysis of blood for presenilin-1 mutation
    - (+) w/ familial Alzheimer's; more likely FTD though

# MKSAP: Question #2

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- A 23-year-old woman believes that she has systemic lupus erythematosus and requests treatment for this condition. For the past 2 years, she has had diffuse muscle and joint pain. She wakes up repeatedly during the night and is sometimes barely able to get out of bed in the morning due to muscle pains and fatigue. She also feels tired when she is in the sun. She has difficulty concentrating and periodically feels like she is “in a fog.” She has chronic headaches not relieved with acetaminophen, ibuprofen, or naproxen. She also has facial rashes that develop and resolve within minutes.

# MKSAP: Question #2

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- Recent laboratory studies show that she has mild iron deficiency anemia, and her antinuclear antibody titer is 1:160. Her aunt also was diagnosed with systemic lupus erythematosus.
- Physical examination reveals diffuse soft-tissue tenderness to palpation at multiple sites. Musculoskeletal examination shows no signs of muscle weakness or synovitis.

# MKSAP: Question #2

Fibromyalgia

- Which of the following is the most appropriate next step in this patient's management?
  - A. Low-dose prednisone and hydroxychloroquine
  - B. Amitriptyline; aerobic exercise
  - C. Anti-Smith and anti-double-stranded DNA antibody
  - D. Oxycodone
  - E. Skin biopsy for lupus band test

MKS

## Fibromyalgia:

- Diffuse muscle and joint pain
  - Nonrestorative sleep
  - Chronic headaches
- A 29-year-old female with a history of systemic lupus erythematosus (SLE) and fibromyalgia. PE w/ diffuse soft-tissue tender points.

For the past 2 years, she has had **diffuse muscle and joint pain**. She **wakes up repeatedly during the night and is sometimes barely able to get out of bed in the morning due to muscle pains and fatigue**. She also feels tired when she is in the sun. She has **difficulty concentrating** and periodically feels like she is “in a fog.” She has **chronic headaches** not relieved with acetaminophen, ibuprofen, or naproxen. She also has facial rashes that develop and resolve within minutes.

- Physical examination reveals **diffuse soft-tissue tenderness to palpation at multiple sites**.

Musculoskeletal examination shows **no signs of muscle weakness or synovitis**.



MKS

ANA Testing:

- (+) in 10-15% of healthy young women
- Only indicated if there is a high pretest probability of SLE or CTD

▪ Rec

*mild*

*iron deficiency anemia, and her antinuclear antibody titer is 1:160.* Her aunt also was diagnosed with systemic lupus erythematosus.



# MKSAP: Question #2

## Fibromyalgia

- Which of the following is the most appropriate next step in this patient's management?
  - A. Low-dose prednisone and hydroxychloroquine
    - Presentation not c/w SLE
  - B. Amitriptyline; aerobic exercise
    - Therapy for Fibromyalgia
  - C. Anti-Smith and anti-double-stranded DNA antibody
    - Presentation not c/w SLE
  - D. Oxycodone
    - Not 1<sup>st</sup> line therapy for Fibromyalgia
  - E. Skin biopsy for lupus band test
    - Presentation not c/w SLE

# MKSAP: Question #3

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- A 63-year-old woman has a 3-month history of gradually increasing abdominal distention and fatigue. She has no other symptoms, and medical history is noncontributory.
- On physical examination, the patient has jaundice and evidence of mild muscle wasting. Xanthelasma and spider angiomas are present. Abdominal examination discloses hepatosplenomegaly and moderate abdominal distention consistent with ascites.

# MKSAP: Question #3

## Laboratory Studies

Hemoglobin	12.3 g/dL (123 g/L)
Platelet count	102,000/ $\mu$ L ( $102 \times 10^9$ /L)
Serum aspartate aminotransferase	53 U/L
Serum alanine aminotransferase	47 U/L
Serum alkaline phosphatase	123 U/L
Serum total bilirubin	3.2 mg/dL (54.72 $\mu$ mol/L)
Serum albumin	2.9 g/dL (29 g/L)
INR	1.3



# MKSAP: Question #3

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- Abdominal ultrasonography shows hepatomegaly, a coarse echotexture of the liver, patent portal and hepatic veins, mild splenomegaly, moderate ascites, and no bile duct dilatation. Paracentesis is performed. The ascitic fluid leukocyte count is 80/ $\mu\text{L}$  ( $0.08 \times 10^9/\text{L}$ ), protein is 1.4 g/dL (14 g/L), and albumin is 0.7 g/dL (7 g/L).

# MKSAP: Question #3

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- Which of the following is the most likely diagnosis?
  - A. Peritoneal carcinomatosis
  - B. Cirrhosis
  - C. Budd-Chiari syndrome
  - D. Dilated cardiomyopathy

# MKSAP: Question #3

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- Serum albumin = 2.9
- Ascitic fluid
  - Protein = 1.4
  - Albumin = 0.7
- Serum-ascites albumin gradient =  $2.9 - 0.7 = 2.2$ 
  - SAAG  $\geq 1.1 \rightarrow$  Portal HTN
  - SAAG  $< 1.1 \rightarrow$  Non-Portal HTN

# SAAG $\geq 1.1 \rightarrow$ Portal HTN

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- Etiologies
  - Sinusoidal: Cirrhosis
  - Post-sinusoidal: Budd-Chiari, R-CHF
  - Pre-sinusoidal: portal / splenic vein thrombosis
- Ascites fluid total protein useful if SAAG  $\geq 1.1$ 
  - AFTP  $< 2.5 \rightarrow$  cirrhosis
  - AFTP  $> 2.5 \rightarrow$  cardiac ascites

# SAAG < 1.1 → Non-Portal HTN

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- Etiologies
  - Peritonitis: TB, ruptured viscus
  - Peritoneal carcinomatosis
  - Pancreatitis

# MKSAP: Question #3

SAAG = 2.2  
AFTP = 1.4

- Which of the following is the most likely diagnosis?
  - A. Peritoneal carcinomatosis
    - Expect a SAAG <1
  - B. Cirrhosis
    - Sinusoidal HTN from chronic liver disease
  - C. Budd-Chiari syndrome
    - Expect AFTP > 2.5
  - D. Dilated cardiomyopathy
    - Expect AFTP > 2.5

# Clinical Image of the Day



**Auer Rod: AML**

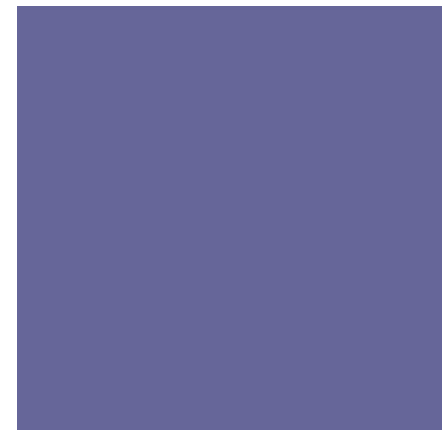
**Myeloblast, ↑N:C,  
cytoplasmic granules,  
rod-shaped inclusion**

# Case Presentation

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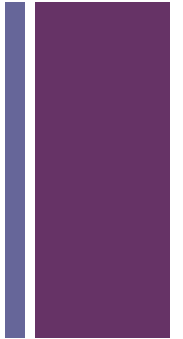
Dr. Zanetta Lamar



# NEUROSARCOIDOSIS

BY BIG Z

# + NEUROSARCOID

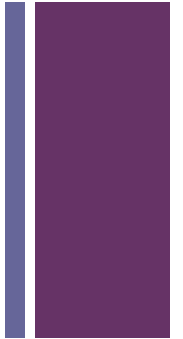


## ■ SARCOID

- Multisystem granulomatous disease
- Noncaseating granulomas
- Older patients presents with systemic symptoms

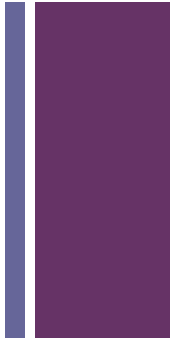


# NEUROSARCOID



- Mortality rate of 10%, twice the overall mortality rate of sarcoidosis.
- Cranial neuropathy and hypothalamic/pituitary lesions tend to occur early and respond favorably to treatment.
- Space-occupying lesions, peripheral neuropathy, and neuromuscular involvement tend to occur later and portend a chronic course.
- Neurosarcoid usually presents with other systemic manifestations.
- Cranial nerve involvement commonly occurs and may be bilateral.
- May present with seizures or meningitis.
- Cerebrospinal fluid (CSF) findings in patients with central nervous system (CNS) involvement may include
  - pleocytosis (mainly lymphocytosis),
  - elevated protein levels or cells (up to 81% of patients), and
  - decreased glucose levels (20% of patients).
  - CSF angiotensin-converting enzyme (ACE) levels may be elevated in 50% of patients, and lysozyme levels, beta2-macroglobulin levels, and
  - CD4/CD8 ratio may be increased.
  - The most common intracranial sites of involvement are the hypothalamus, the third ventricle region, and the pituitary gland

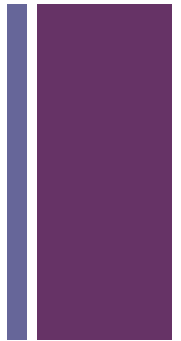
# + NEUROSARCOID



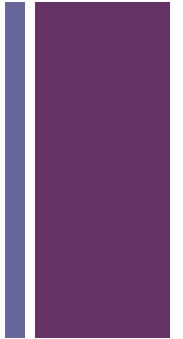
- Differential
  - Cranial nerve disorders
    - Bell's Palsy, Lyme disease
  - Parenchymal lesions
    - Metastasis
  - Peripheral neuropathy
    - Inflammatory, toxic, metabolic
  - Meningeal disease
    - HIV, TB, Syphilis
  - Muscle weakness
    - Polymyositis

# + RANDOM NUMBERS

■ Organ	Number	Percent
■ LUNGS	699	95.0
■ Skin	117	15.9
■ Lymph node	112	15.2
■ Eye	87	11.8
■ Liver	85	11.5
■ Erythema Nod	61	8.3
■ Spleen	49	6.7
■ NEUROLOGIC	34	4.6
■ Parotid/salivary	29	3.9
■ Bone marrow	29	3.9
■ Calcium	27	3.7

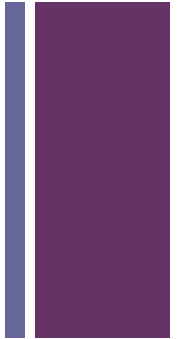


# + NEUROSARCOID TX



- No RCT's
- Corticosteroids
  - mainstay of tx,
  - tx base on response
  - If relapse occurs immunomodulators, radiation may be next step

# + NEUROSARCOID



- 10% die
- 2/3 have can have a monophasic, a relapsing-remitting course, or progressive disease punctuated by episodic deteriorations
- Long-term course of neurosarcoidosis has not been clearly defined.

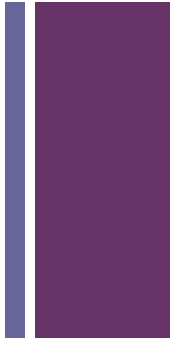
# + References

- Up to Date Online
- Emedicine





# THE END



- QUESTIONS
- COMMENTS
- CRITICISMS