

Meet the Professor

CASE 2



Case One

- ID: 60 year old man
- Chief Complaint: Altered sensorium and abdominal distension for 2 days



History of Present Illness

- Over the past 3 months
 - slow mentation
 - inattention
 - word finding difficulties
 - agitation
 - hallucinations
 - delusions of persecution
- Over the past several weeks
 - unable to perform activities of daily living

Summary

- 60 year old male
- altered sensorium
- abd. distension



History of Present Illness

- No additional collateral history

Summary

- 60 year old male
- altered sensorium
- abd. distension



Review of Systems

- General
 - Unknown weight changes
- HEENT
 - No headache or neck pain

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions



Review of Systems

- Heart
 - No chest pain
- Lungs
 - No shortness of breath

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions



Review of Systems

- Abdomen
 - Constipation
 - Distension

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions



Review of Systems

- Genitourinary
 - No dysuria
- Extremities
 - Mild chronic swelling

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation



Review of Systems

- Skin – no rash

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema



Past Medical History

- HTN (diet controlled)
- Seasonal allergies
- No abdominal surgeries

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema



Allergies

- No drug allergies

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries



Medications

- Loratadine 10mg qd prn allergies
- No reported OTC or herbal medications

Summary

- 60 year old male
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- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries



Social History

- Alcohol - none
- Tobacco – none
- Drugs – none (ever)
- Single – married, single partner
- No recent travel history
- Works in a printing press

Summary

- 60 year old male
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- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine



Family History

Father

- HTN, depression

Mother

- COPD, depression

Brother

- Depression

Summary

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- inattention
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- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work



Physical Exam

- Temperature: 97.5 Fahrenheit
- Blood Pressure 110/64
- Pulse 57
- Respirations 12
- BMI = 31

Summary

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- constipation
- edema
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- loratadine
- printing press work
- fam hx depression



Physical Exam

- **General**
 - Appears drowsy
 - Somewhat responsive
 - No oriented to place or time

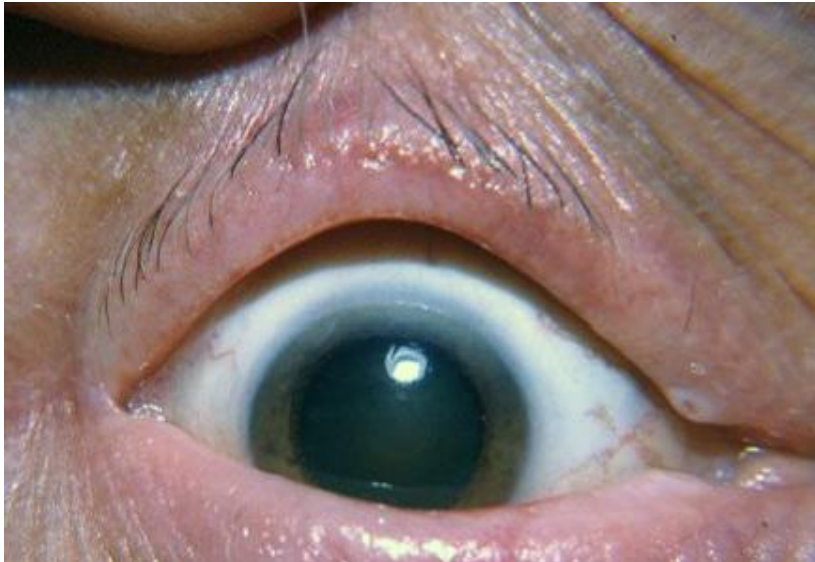
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Physical Exam

- HEENT



Summary

- 60 year old male
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- edema
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- loratadine
- printing press work
- Fam hx depression
- Drowsy
- Not oriented



Physical Exam

- Neck
 - No lymphadenopathy
 - No thyromegaly
 - No bruits
- Heart
 - No murmurs or gallops

Summary

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- drowsy
- not oriented



Physical Exam

- Lungs
 - Clear
- Abdomen
 - Not tender
 - No HSM



Summary

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- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
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- drowsy
- not oriented



Physical Exam

- Extremities
 - 2+ soft pitting edema
 - Cooler distally than proximally

- Skin:



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- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted



Physical Exam

- Neurologic
 - Drowsy but arousable
 - Oriented to self only
 - Agitated and Hallucinating (“hospital staff is out to get me”)
 - Moves all 4 extremities on command, but not able to do formal strength testing
 - Sensation appeared grossly intact
 - DTR’s diminished/sluggish
 - Unable to walk

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Initial Laboratory Exam

A.

- 1. CBC
- 2. Renal
- 3. LFT's
- 4. Blood Culture
- 5. UA
- 6. EKG
- 7. Drug Screen

B.

- 1. CXR
- 2. Abd X-ray
- 3. CK, troponin
- 4. ABG
- 5. Head CT
- 6. ESR
- 7. Folate
- 8. B12 level

To Secondary Tests

Summary

- 60 year old male
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CBC

- WBC 3.9 K/UL
- RBC 4.41 M/UL
- HGB 9.1 G/DL
- HCT 26.2 %
- MCV 100.1 FL
- MCH 25.5 PG
- MCHC 33.1 G/DL
- RDW 14.5 %
- PLT 289 K/UL

■ To initial labs

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Renal

- Sodium 140 MEQ/L
- Potassium 4.1 MEQ/L
- Chloride 100 MEQ/L
- CO2 24 MEQ/L
- BUN 12 MG/DL
- Creatinine 1.1 MG/DL
- Glucose 78 MG/DL

■ To initial labs

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- exam as noted



LFT's

- AST = 22 U/L (11-35)
- ALT = 35 U/L (7-46)
- Alkaline Phosphatase = 129 U/L (46-240)
- Bilirubin, total = 0.8 MG/DL (0.0-1.0)
- Albumin 3.5 GM/DL (3.5-5.0)

■ To initial labs

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Blood Cultures

- Negative

■ To initial labs

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Urinalysis

- Specific Gravity = 1.020
- Yellow
- pH = 6.0
- Protein = 0
- RBC = 0 per hpf
- WBC = 0 per hpf
- No casts
- Nitrite negative
- Leukocyte esterase negative

■ To initial labs

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ABG (on room air)

- pH = 7.41
- pCO₂ = 38
- PO₂ = 89

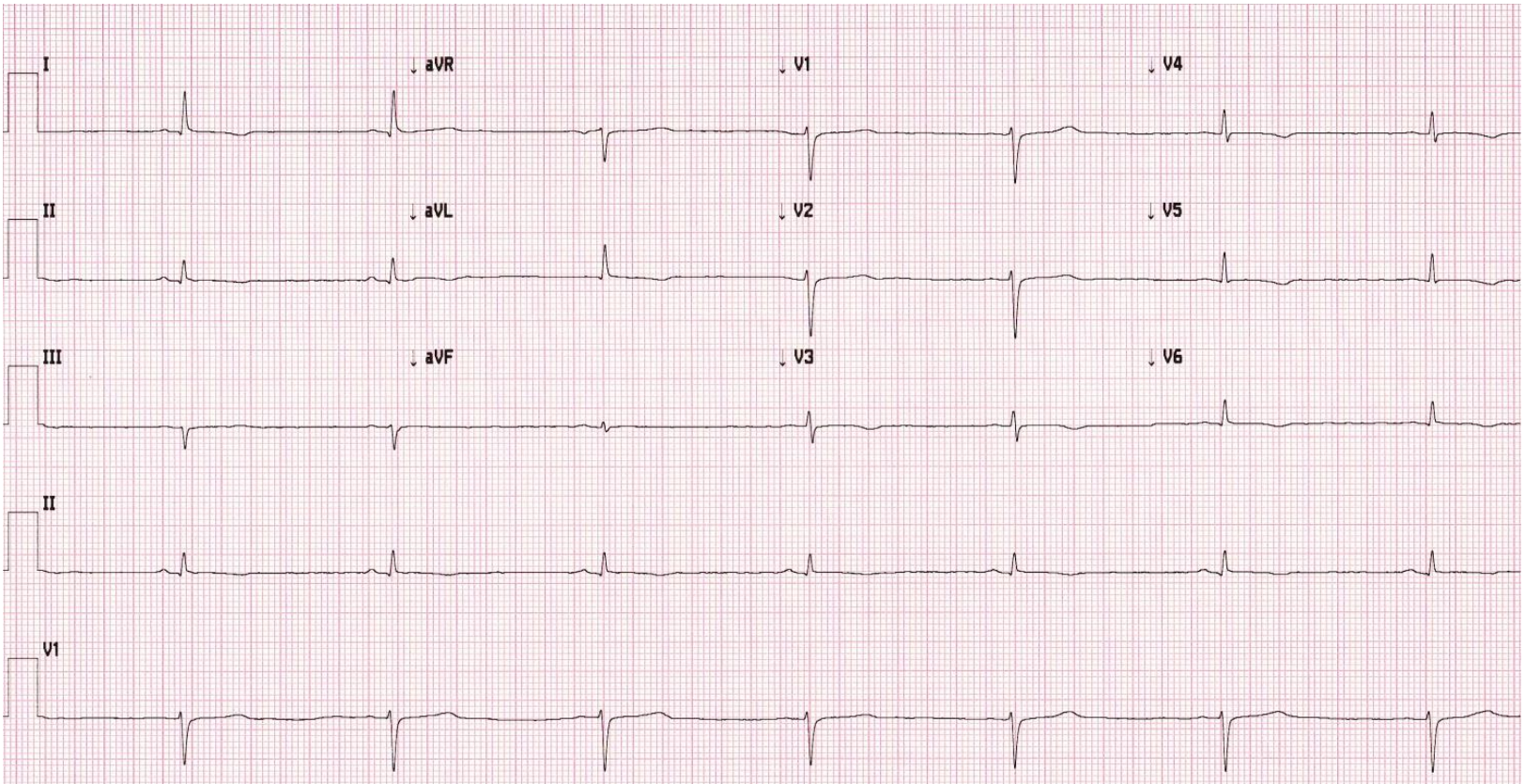
■ To initial labs

Summary

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- exam as noted



EKG



■ To initial labs



CXR



■ To initial labs



AXR



Supine



Upright

Summary

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■ To initial labs



Folate

- Normal

■ To initial labs

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ESR

- 16 mm/hr

■ To initial labs

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Head CT

- Negative (non contrast)

■ To initial labs

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- exam as noted



B12 level

- Normal

■ To initial labs

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- edema
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- loratadine
- printing press work
- fam hx depression
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- not oriented
- exam as noted



CK, Troponins

- CK = 43 mg/dl
- Troponin 0.00 mcg/L

■ To initial labs

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- exam as noted



Toxicology

- Negative

■ To initial labs

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- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted



Secondary Laboratory Exam

- | | |
|---|--|
| <input type="checkbox"/> 1. Abdomen US | <input type="checkbox"/> 11. WNV titers |
| <input type="checkbox"/> 2. CT Abdomen | <input type="checkbox"/> 12. Lead level |
| <input type="checkbox"/> 3. ECHO (TTE) | <input type="checkbox"/> 13. Gut manometry |
| <input type="checkbox"/> 4. MRI Brain | <input type="checkbox"/> 14. Glucose/H ⁺
breath test |
| <input type="checkbox"/> 5. Skin biopsy | <input type="checkbox"/> 15. 14-3-3 protein |
| <input type="checkbox"/> 6. Lumbar Puncture | <input type="checkbox"/> 16. RPR |
| <input type="checkbox"/> 7. Iron Studies | <input type="checkbox"/> 17. HIV |
| <input type="checkbox"/> 8. EEG | <input type="checkbox"/> 18. MMSE |
| <input type="checkbox"/> 9. RF | <input type="checkbox"/> 19. ANCA |
| <input type="checkbox"/> 10. ANA | <input type="checkbox"/> 20. Copper level |

To initial tests

To next steps

Summary

- 60 year old male
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- agitation
- hallucinations
- delusions
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- loratadine
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- exam as noted



Abdominal US

Summary

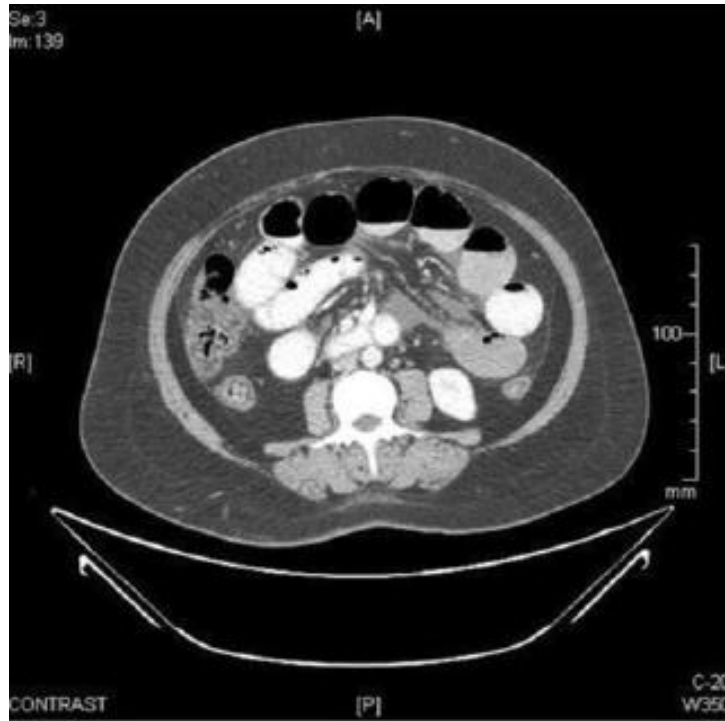
- Gaseous distention with dilated bowel loops.

- 60 year old male
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- word finding prob.
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- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



CT Abdomen



- Dilated bowel loops

■ To Secondary Tests

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- edema
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- loratadine
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- not oriented
- exam as noted



ECHO

- Not done

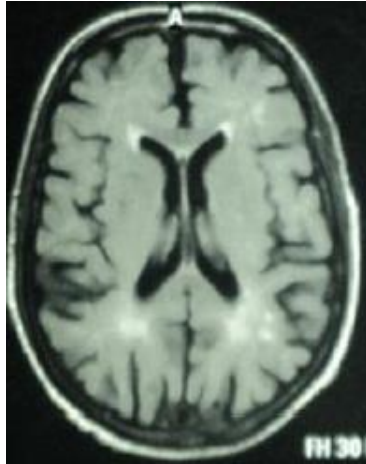
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- drowsy
- not oriented
- exam as noted

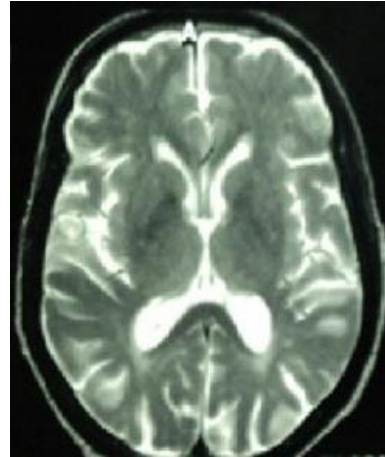
■ To Secondary Tests



MRI Brain



Fluid-attenuated inversion recovery sequence showing prominent bilateral cortical sulci and Sylvian fissures with bilateral periventricular ooze



T2 magnetic resonance imaging image showing hyperintensities in periventricular areas

Summary

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- exam as noted

■ To Secondary Tests



Skin Biopsy

- Not done

Summary

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- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



Lumbar Puncture

- Initial studies:
 - Normal glucose
 - Elevated protein
 - No WBC
 - No RBCs
 - HSV PCR negative

Summary

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- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



Iron Studies

- Normal

Summary

- 60 year old male
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- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



EEG

Summary

- Normal

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



WNV titers

Summary

- Not done

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



Lead level

- 2.19 mcg/dL (nl < 25 mcg/dL)

Summary

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■ To Secondary Tests



Gut Manometry

Summary

- change in the frequency of the slow wave oscillations of smooth muscle electric potential features suggestive of intestinal dysmotility

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- exam as noted

■ To Secondary Tests



Glucose Hydrogen Breath Test

Summary

- Negative

- 60 year old male
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- inattention
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- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



14-3-3 Protein

Summary

- Negative

- 60 year old male
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- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



RPR

Summary

- Negative

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



HIV

Summary

- Negative

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



MMSE

● 14/30

Summary

- 60 year old male
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- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
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- not oriented
- exam as noted

■ To Secondary Tests



Rheumatoid Factor

Summary

- Negative

- 60 year old male
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- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



ANA

- Negative

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
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■ To Secondary Tests



ANCA

- Negative

Summary

- 60 year old male
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- agitation
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- edema
- no abd. Surgeries
- loratadine
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- exam as noted

■ To Secondary Tests



Copper Level

- Not done

Summary

- 60 year old male
- altered sensorium
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- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

■ To Secondary Tests



Next Steps

- Start antibiotics
- Start acyclovir
- Psychiatry consult
- Colonoscopy
- TFT's
- Sleep deprived EEG
- Fat pad biopsy
- Brain biopsy
- CRP
- To Secondary Tests
- To Answer
- Get travel history
- Ophtho consult
- Neuro consult
- Anti-NMDA ab
- Fentanyl screen
- Anti-TTG ab
- Calcium
- Heavy metals
- Observe

Summary

- 60 year old male
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- constipation
- edema
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- loratadine
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- fam hx depression
- drowsy
- not oriented
- exam as noted



Next Steps

Summary

No.

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

- To Secondary Tests
- To Next Steps



Next Steps

- TSH = 76.52 μ IU/mL
- FT4 = 0.1 ng/dL
- FT3 = 0.42 ng/dL

Now what do you want?

- Thyroid CT
 - Uptake Scan
 - Anti-TPO antibodies
 - Thyroid Binding Globulin
 - TRH Test
 - Thyroid Biopsy
-
- To Secondary Tests
 - To Answer

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted



Next Steps

Nope.

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted

- To Thyroid Tests
- To Answer



Next Steps

Blood Anti-TPO antibodies = 580 units/mL (normal: <60 units/mL)

CSF Anti-TPO antibodies positive

- To Thyroid Tests
- To Answer

Summary

- 60 year old male
- altered sensorium
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- word finding prob.
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- drowsy
- not oriented
- exam as noted





Answer

**Mental status changes,
constipation, bradycardia,
madarosis, xerosis,
delayed reflexes due to:**

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted



Answer

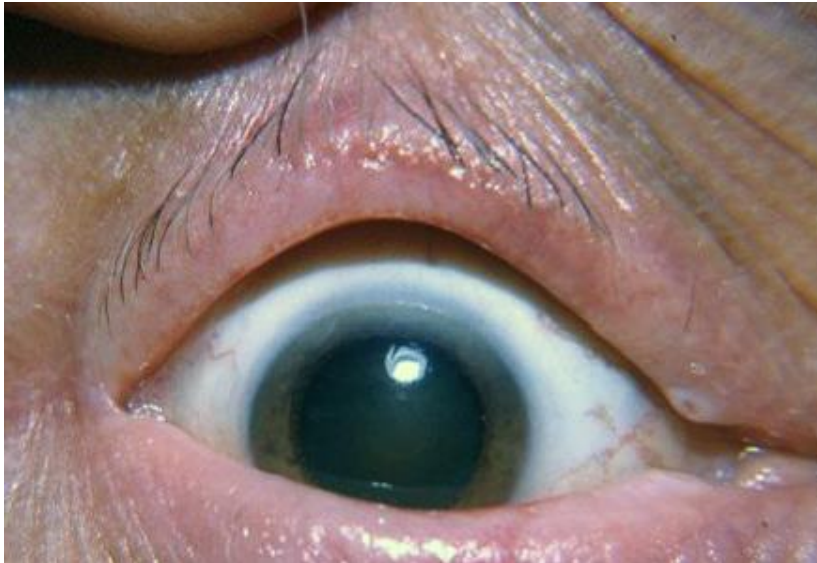
Hashimoto encephalopathy
with gut pseudo-obstruction

Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted



Madarosis



Summary

- 60 year old male
- altered sensorium
- abd. distension
- slow mentation
- inattention
- word finding prob.
- agitation
- hallucinations
- delusions
- constipation
- edema
- no abd. Surgeries
- loratadine
- printing press work
- fam hx depression
- drowsy
- not oriented
- exam as noted



Hashimoto's Encephalopathy

- Lord Brain described Hashimoto's encephalopathy (HE) in a patient with Hashimoto's thyroiditis as characterized by cloudiness of consciousness, tremors, cognitive loss and stroke-like episodes (1966)



Brain L, Jellinek EH, Ball K: **Hashimoto's disease and encephalopathy**. Lancet 1966, 2(7462):512–514.



Hashimoto's Encephalopathy

- Also known as:
 - myxedema madness
 - encephalopathy associated with autoimmune thyroid disease
 - steroid responsive encephalopathy associated with autoimmune thyroiditis

Shera, Irfan A., et al. "Unusual case of Hashimoto's encephalopathy and pseudo-obstruction in a patient with undiagnosed hypothyroidism: a case report." *Journal of medical case reports* 8.1 (2014):



Hashimoto's Encephalopathy

- Controversial diagnosis – not everyone believes in it
- most often characterized by a subacute onset of confusion with altered level of consciousness, seizures, and myoclonus
- believed to be an immune-mediated disorder rather than representing the direct effect of an altered thyroid state on the central nervous system



Hashimoto's Encephalopathy

- bulk of evidence points to an autoimmune vasculitis or other inflammatory process, perhaps associated with immune complex deposition, and possibly disrupting the cerebral microvasculature
- nature of the relationship between Hashimoto thyroiditis (HT) and HE is unclear
 - Some argue that the association is entirely spurious
- symptoms of HE are not paralleled by symptoms of thyroiditis. In addition, changes in antithyroid autoantibody levels do not consistently correspond with neurologic symptoms or improvement with treatment.



Hashimoto's Encephalopathy

- These findings lead some to propose that steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT) is a more appropriate designation for this disorder



Hashimoto's Encephalopathy

Any disease associated with a syndrome of delirium or rapidly progressive dementia may be confused with Hashimoto encephalopathy (HE). These include:

- Creutzfeldt-Jakob disease
- Acute disseminated encephalomyelitis
- Toxic metabolic encephalopathies (table 1)
- Meningoencephalitis
- Psychiatric disease (depression, anxiety, psychosis)
- Carcinomatous meningitis
- Paraneoplastic or autoimmune encephalitis
- Degenerative dementia (Alzheimer disease, dementia with Lewy bodies, frontotemporal dementia)
- Stroke or transient ischemic attack
- Basilar or hemiplegic migraine
- Cerebral vasculitis



Hashimoto's Encephalopathy

Take Home Points

1. It is rare
2. It is an autoimmune disorder, and not due to the thyroiditis
3. Presence of elevated antithyroid antibody titres and the exclusion of other causes of encephalopathy support the diagnosis of HE.
4. If you suspect the diagnosis, steroids are in order
5. With proper treatment most recover



The Patient

- He received hydrocortisone – improved almost immediately
- He started levothyroxine on day 2
- His abdominal symptoms took 1 month resolve
- After 6 weeks of steroids his MMSE was 28/30



Reference

Shera et al. *Journal of Medical Case Reports* 2014, **8**:296
<http://www.jmedicalcasereports.com/content/8/1/296>



CASE REPORT

Open Access

Unusual case of Hashimoto's encephalopathy and pseudo-obstruction in a patient with undiagnosed hypothyroidism: a case report

Irfan A Shera^{1*}, Anurag Vyas¹, Mohd Shafi Bhat² and Qaysar Yousof³

Abstract

Introduction: Hashimoto's encephalopathy is a relatively rare condition associated with an elevated concentration of circulating serum anti-thyroid antibodies, and is usually responsive to steroid therapy. However, hypothyroidism is a rare cause of pseudo-obstruction so here we present a case report of Hashimoto's encephalopathy with gut pseudo-obstruction in an undiagnosed hypothyroid patient.

Case presentation: A diagnosis of unknown aetiology of encephalopathy with gut dysmotility in an undiagnosed profound hypothyroidism case associated with cognitive decline and behavioural disorder was made in a 60-year-old Indian man. The associated clinical and laboratory features led to the final diagnosis of overt hypothyroidism with Hashimoto's encephalopathy with gut pseudo-obstruction.

Conclusions: Hashimoto's encephalopathy is a rare disorder presenting with acute or sub acute encephalopathy of unknown aetiology so there are considerable chances of misdiagnosing it. The unusualness of this case is that since hypothyroidism is a rare cause of intestinal pseudo-obstruction, and presented concomitant with Hashimoto's encephalopathy, that itself is a rare entity. Intestinal pseudo-obstruction is a potentially serious complication that must be recognized and treated promptly with adequate thyroid hormone therapy.

Keywords: Hashimoto's encephalopathy (HE), Overt hypothyroidism, Pseudo-obstruction

Introduction

Hypothyroidism is a disorder caused by hypofunction of the thyroid gland. Iodine deficiency is the most common cause of hypothyroidism worldwide; however, in areas of iodine sufficiency Hashimoto's thyroiditis and iatrogenic causes are most common. Because of its autoimmune nature there is a gradual decline in thyroid function with presentation of a wide range of disease symptoms. Some patients may have minor symptoms, which is called subclinical hypothyroidism, whereas others have a fall in unbound T4 levels and a steep rise in thyroid-stimulating hormone (TSH) > 10 μIU/L, which is referred to as clinical or overt hypothyroidism [1].

Lord Brain in 1966 described Hashimoto's encephalopathy (HE) in a patient with Hashimoto's thyroiditis as characterized by cloudiness of consciousness, tremors, cognitive

loss and stroke-like episodes [2]. Since then HE has gained importance in differential diagnosis of encephalopathy of unknown origin. Shaw [3] in 1991 coined the term HE by describing the constellation of symptoms such as seizure, disorientation, frequent episodes of alternating hemiparesis, high protein levels in cerebrospinal fluid (CSF) and electrocardiogram (ECG) abnormalities. However, these patients also had hypothyroidism and positive anti-thyroid antibodies. Because of the severe neurological complexities the term HE is widely used while some other terms such as myxoedema madness [4], encephalopathy associated with autoimmune thyroid disease [5] or steroid responsive encephalopathy associated with autoimmune thyroiditis [6] have been discarded.

HE is a relatively rare condition; therefore there are considerable chances of misdiagnosing it. HE is generally considered to be an autoimmune encephalopathy; however the pathogenesis is still not clear. Antithyroid peroxidase (anti-TPO) antibodies are found in almost all cases of HE [7] but can also be present in the general population with

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