

# Neurology Case Presentation

Said Ibrahim MD

VCU

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# Chief Complaint

- Acute headache with hemi-paresis and sensory lost.

# Case Presentation

- 25 year old AAM woke up at 5am with acute left side headache, throbbing in nature 10/10, with photophobia and neck pain, no hx of trauma. Patient also had acute hemi-paresis of left arm and leg, with left peripheral facial palsy, and decrease sensation to all modalities involving left arm, left leg and left face. There was also a complaint of b/l blurry vision, but no chest pain.

# Case Presentation

- ROS: As per HPI
- Past medical history: None, other than unprotected sex.
- Social history: Works as security guard. Lives with parents. Associate degree. Drinks socially, no drugs, no alcohol.

# Case Presentation

## ■ Physical Exam

- ◆ T: 98.9, HR: 80, BP: 146/90, RR: 20
- ◆ General: Complaining of headache
- ◆ HEENT: Atraumatic and normcephalic, no meningismus signs, neck supple.
- ◆ C/V: S1 and S2 NSR, no murmurs
- ◆ Chest/Abd/Ext: Normal

# Case Presentation

## ■ Neurological Exam

- ◆ Mental Status: Normal
- ◆ Speech: Normal
- ◆ CN 2-12 normal, except CN 5 and 7 on left side.
- ◆ Motor: 4/5 in left arm and leg. Right side normal.

# Case Presentation

## ■ Neurological Exam

- ◆ Sensation: Decreased to all modalities in left face V1, V2, V3, and left arm, and leg.
- ◆ Reflexes: 2 through out
- ◆ Toes: Down going bilaterally
- ◆ Cerebellum signs normal
- ◆ Gait: Unsteady due to left sided weakness.

# Diagnostic Results

- Non-contrast head CT: Normal other than Right maxillary sinus mucus retention cyst.
- MRI brain without contrast: T1, T2, Flair, DWI normal.
- MRA of head and neck normal.
- 2<sup>nd</sup> MRI brain both pre-contrast T1 and post-contrast T1, and DWI normal.

# Diagnostic Results

## ■ CBC

- ◆ WBC 10.8
- ◆ Hb 15.9
- ◆ Hct 45.6
- ◆ Platelet 285

## ■ BMP

- ◆ Na 138
- ◆ K 4.2
- ◆ Chloride 103
- ◆ Bicarb 28
- ◆ Bun 10
- ◆ Creatine 0.73
- ◆ Glucose 97

# Diagnostic Results

## ■ LP and CSF results

◆ <u>Tube one</u>	<u>Tube three</u>	<u>Glucose</u>	<u>Protein</u>
◆ Clear	Clear	57	43
◆ RBC 1	RBC none		
◆ WBC 22	WBC 12		
◆ 21 lymph	12 lymph		
◆ 1 mono			

# Diagnostic Results

## ■ CSF studies

- ◆ Cryptococcus Ag negative
- ◆ Culture negative
- ◆ Viral culture negative
- ◆ Fungal culture negative
- ◆ HSV type 1 and 2 negative
- ◆ ACE level 1.0 WNL
- ◆ *CSF-VDRL negative*

# Diagnostic Results

- Homocysteine 15.0
- *Vitamin B12 level: 161 pg/ml*
- Cholesterol level: 161
- LDL 88
- HDL 36
- ANA negative
- ACE level 40 WNL
- ESR 1mm in 1 hour
- Lyme ab IgM negative
- Lyme total ab negative
- *HIV type 1 ab POSITIVE*
- *RPR qualitative POSITIVE*
- *RPR titer 1:4*
- *FTA-ABS POSITIVE*
- Ig G index not done
- Oligoclonal Bands negative
- Myelin Basic Protein Negative

# Clinical Summary

- 25yo AAM with hx of unprotected sex, presents with acute left sided headache associated with left peripheral facial palsy and sensory loss with left sided hemi-paresis and left sensory loss. Normal neuro-imaging of the brain. No other risk factors, other than newly diagnosed HIV and Syphilis, with aseptic meningitis.

# Differential Diagnosis Prior to Work-up

- Stroke
- TIA
- Complex Migraine
- Vertebral Artery Dissection
- Vasculitis
- Drug(cocaine)
- HIV
- Vitamin B12 deficiency
- Neurosyphilis
- Hypoglycemia
- Hyperglycemia
- Post-ictal Todds
- Intra-cranial mass
- Neurosarcoid
- Meningitis
- Lyme disease

# Differential Diagnosis Post Work-up

- Vitamin B12 deficiency
- HIV
- Stroke
- Neurosyphilis
- Vasculitis
- Aseptic meningitis

# Clinical Course

- Patient's Vitamin B12 replaced
- Diagnosis of HIV and Syphilis made post-discharge as outpatient, and PCP made referral to ID. Patient received IV regiment of PCN G crystalline aqueous form, and with this there was marked improvement of headaches and hemi-paresis.

# Final Diagnosis Neurosyphilis

- Probable Neurosyphilis causing deficits, based on continual clinical work-up, and course of patient over several months, both as an inpatient and outpatient.
- Localization of deficits difficult, especially with normal neuro-imaging. It is possible that facial palsy caused by meningitis.
- Even though CSF VDRL negative, neurosyphilis still possible in the setting of
  - ◆ Neuro-deficits
  - ◆ positive serum FTA-ABS
  - ◆ CSF pleocytosis above 5
  - ◆ Improvement of clinical symptoms after PCN G IV treatment.

# Final Diagnosis

- Even though CSF-VDRL is considered to be golden standard to diagnosis neurosyphilis, it is imperfect.
  - ◆ Due to low sensitivity of 30-70% and high specificity, an positive CSF-VDRL result confirms diagnosis, but a negative result can not rule out diagnosis.
  - ◆ Therefore CSF-VDRL may be negative in as many as 70% of individuals with neurosyphilis.

# Final Diagnosis

- No universally adopted CSF parameters for the diagnosis of neurosyphilis.
- Some authorities such as Bradley or Adam/Victor state:
  - ◆ CSF pleocytosis  $>5$
  - ◆ protein  $>40-45$ , with positive CSF-VDRL.
  - ◆ Negative CSF-VDRL with the above pleocytosis and protein in combination w/ elevated Ig G index, positive serum FTA-ABS and neuro-deficits.
- Other sources such as a paper overlooking the diagnosis and course of neurosyphilis of 161 pt in Cape Town, South Africa.
  - ◆ positive CSF-VDRL
  - ◆ positive CSF FTA-abs,
  - ◆ pleocytosis(PML orlymp)  $>5/ml$
  - ◆ protein  $>45mg/dl$
  - ◆ Ig G index  $>0.6$ .

# Medlink Neurosyphilis Dx Criteria in the face of HIV infx

- Definite diagnosis
  - ◆ Positive CSF-VDRL
  - ◆ Positive serum FTA-ABS and RPR
- Probable diagnosis
  - ◆ Positive FTA-ABS and RPR
  - ◆ Negative CSF-VDRL
  - ◆ Pleocytosis  $> 20$  cell/mm<sup>3</sup>
  - ◆ Protein  $> 60$ mg/dl
  - ◆ With neurological deficits

# Clinical Question

- With the stroke-like symptoms of this patient, and normal neurological imaging of the brain, a clinical question is raised. What does the neurological imaging of patient's with neurosyphilis and neurological deficits look like?

# Article One

- A retrospective evaluation of six patients who were HIV positive with neurosyphilis, and other opportunistic infx ruled out. Five pt had acute and sub-acute stroke like symptoms involving the BG and MCA, and one had parietal convexity mass mimicking a meningioma with HA and ataxia. Contrast enhanced MR images showed patchy enchancement of the BG and MCA in the five pts, and the convexity mass in the sixth pt, was shown to be a syphilitic gumma.

# Article one (continued)

- The imaging findings in the five pt represented ischemic infarcts caused by meningovascular syphilis. After PCN treatment, serum and CSF VDRL titers dropped, and neurological symptoms and deficits improved in all six pt. An follow up MR of the pt with the gumma, showed complete resolution after PCN tx. In young pt with HIV and stroke like symptoms a diagnosis of neurosyphilis should be considered, and appropriate neuro-imaging should be used to facilitate diagnosis and proper treatment.

# Article Two

- The neuro-imaging studies of 35 patients with document neurosyphilis were reviewed. Of the 35 patients, 32 test positive for HIV, and 3 were HIV negative.

# CT, MR, and angiography findings in the 35 patients

<u>Findings</u>	<u>No(%) of patients</u>
Normal	11(31)
Atrophy	13(37)
-Mild	-7(54)
-Moderate	-5(38)
-Severe	-1(8)
Cerebral Infarct	8(23)
-Corticol/subcortical	-4(50)
-Brainstem	-2(25)
-BG/Thalamus	-5(63)

# CT, MR, angiography findings in the 35 patients (continued)

<u>Findings</u>	<u>No(%) of patients</u>
White matter dz	7(20)
Gummas	2(6)
Extra-axial enhancement	2(6)
Arteritis in 2/4 angio	2(50)

# Therapeutic options for Neurosyphilis

Treponema pallidum is highly sensitive to penicillin.

The treponemicidal level of PCN is 0.03ug/ml.

1. PCN G crystalline aqueous, 4 million units IV every 4 hours or 24 million units, continuous IV infusion daily for 10-14 days. This option achieves treponemicidal levels of PCN in CSF and is most effective.
2. PCN G procaine 2.4 million units IM daily with probenecid 500mg four times daily oral, both for 10-14 days. This level does not achieve treponemicidal levels in CSF.
3. Ceftriaxone 2g IV daily for 10-14 days for PCN allergic patients.

# Resources for patients and family

- Daily Strength

- ◆ [www.dailystrength.org](http://www.dailystrength.org)

- Patient's Association

- ◆ <http://www.patients-association.com>

# Prognosis

- Most patients with earlier forms of neurosyphilis such as asymptomatic meningitis, meningovascular syphilis, gummas or optic syphilis show very good improvement with PCN treatment, and little neurological complications.
- Later forms of the disease such as tabes dorsalis or general paresis do not show good improvement or response to PCN.
- MR findings of medial temporal lobe atrophy, indicative of very poor prognosis.

# Board Questions Q1

- What percentage of syphilis patients develop neurological complications, if not treated appropriately early in the disease course?
  1. 5-10 percent
  2. 25 percent
  3. 50 percent
  4. 70 percent
  5. 50-80 percent

# Answer to Q1

- Choice 1

- ◆ 5-10 percent of patient develop neurological complications if not treated early in the course of the disease.

# Board Question Q2

- How many main forms of neurosyphilis exist, and what are they?
  1. 2; Asymptomatic meningitis, Tabes Dorsalis
  2. 1; General Paresis
  3. 5; Asymptomatic meningitis, meningovascular, central type, tabes dorsalis, general paresis.
  4. 4; Asymptomatic meningitis, meningovascular, tabes dorsalis, general paresis.
  5. 3; Asymptomatic meningitis, tabes dorsalis, general paresis.

# Answer Q2

- Choice 4

- ◆ 4 main types of neurosyphilis exist, which are asymptomatic meningitis, meningovascular syphilis, tabes dorsalis, and general paresis.

# Board Question Q3

- In what forms of neurosyphilis is PCN most effective?
  1. Meningovascular
  2. Tabes Dorsalis
  3. General Paresis
  4. Asymptomatic Meningitis
  5. Choice A and D

# Answer to Q3

- Choice 5

- ◆ PCN most effective in the treatment of meningovascular and asymptomatic meningitis.

# Additional Comments

- The incidence of neurosyphilis has greatly reduced since the advent of antibiotics, and it's natural course has change too. However with the rise of HIV, the incidence of syphilis has started to rise again.

# References

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