Paraneoplastic limbic encephalitis in Hodgkin’s Lymphoma

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Our patient’s CC: Disorientation and insomnia

HPI: 27 year old man AW with no PMH presents with 6-8 week history of progressive confusion and insomnia. Cannot recall day’s events, and is frequently confused and frustrated. No recent travel history, illnesses, or toxic/animal exposures.

PE: NAD, VS wnl, Aaox1. Can recall US presidents back to FDR, his childhood pet’s name. 0/4 objects at 2 minutes.

LP: clear, colorless, 1 RBC, 4 WBCs in 4th tube, Gram stain negative. (Does not recall LP 45’ later.)
Menu of Tests

1. CT w/out contrast  
   -to assess acute hemorrhage, mass lesions

1a. CT w/contrast  
   -to assess mass lesions, infarcted tissue

2. MRI  
   -anatomy, soft tissue, inflammation
Our patient: Axial Cranial MRI

T1
- Pre-contrast

T2
- Pre-contrast

Post-contrast

FLAIR

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Anatomy review

- Suprasellar cistern
- Chiasmatic cistern
- Basilar artery
- Lateral ventricle
- Substantia nigra
- Quadrigeminal cistern
- Cerebellar folia
- Midbrain
- Temporal lobe
- Hippocampus
- PCA
What’s in the differential?

1. Viral encephalitis
   -HSV1, HHV6
2. Bacterial encephalitis
   -Bartonella (CSD), Lyme, TB
3. Autoimmune
4. Paraneoplastic
Companion patient 1: HSV encephalitis on axial MRI

- Asymmetric lesions
- Associated hemorrhage
- Fever in >90% of patients, frequent seizures
- Patient had no CSF lymphocytosis, Negative CSF HSV PCR

www.utdol.com, ‘HSV1 encephalitis’
Companion patient 2:
HHV6 encephalitis on axial MRI

HHV6 causes limbic encephalitis with anterograde amnesia and temporal lobe MRI abnormalities

Disease seen only in setting of profound Immunocompromise: “PALE”=post-transplant acute limbic encephalitis

Patient had negative HHV6 CSF PCR

Narrowing the differential

1. Viral encephalitis
   - HSV1, HHV6
2. Bacterial encephalitis
   - Bartonella (CSD), Lyme, TB
3. Autoimmune
4. Paraneoplastic

No systemic manifestations
No known exposures
Negative PPD
Negative CSF cultures and serologies

Lesions atypical for MS
Presentation atypical for MS
No oligoclonal CSF banding
Normal ESR, ANCA, ANA
Paraneoplastic Syndrome

Anti-cancer immune response aberrantly targets self
- myasthenia gravis and thymoma
- antibody-mediated

Most common cancers in 15-29 year old males:
1. Testicular
2. Lymphoma
3. Melanoma --> no skin lesions
4. CNS --> already performed MRI
5. Leukemia --> normal CBC

Radiologic menu of tests to search for cancer in 27 year old
-testicular ultrasound --> normal
-CT, PET
Our patient:
Anterior mediastinal mass on axial chest CT

Oval shaped anterior mediastinal mass, 2.5 cm in largest diameter, approximately 50 Hounsfield units, no hilar, or axillary lymphadenopathy

DDx Ant. Mediastinal Mass?
1. Thymoma
2. Lymphoma
3. Germ Cell Tumor
4. Ectopic thyroid tissue

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Our patient: Coronal PET Scan

PET uses F18-fluoro-deoxyglucose (FDG) to label metabolically-active tissue as “FDG-avid”

Intensely FDG-avid mass in the region of the thymus

Moderate FDG-avid lymph nodes in right para-tracheal regions

No FDG-avid masses below the diaphragm, or FDG-avid destructive bone lesions

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Sternotomy was performed and the mass was resected
Our patient: microscopic histopathology

Pathology: Multiple CD15+CD30+ Reed-Sternberg cells, with prominent mixed lymphocytic infiltration and dense fibrous septae within normal thymic tissue. 

Consistent with diagnosis of intrathymic nodular sclerosis Hodgkin’s lymphoma.

Image courtesy of Dr Kevin Long, BWH Pathology
Use of PET to Stage Hodgkin’s Disease (HD)

+/− “B symptoms”: night sweats, fever, unexplained >10% weight loss
Companion patient 3: Stage IV HD on coronal PET

40 year-old man with HD

Image courtesy of Dr. Patrick Donohoe, PACS, BIDMC
Companion patient 3:
Stage IV HD on axial C-CT

Image courtesy of Dr. Patrick Donohoe,
PACS, BIDMC
Companion patient 3: Stage IV HD on axial PET/CT

FDG-avid para-aortic lymphadenopathy

Images courtesy of Dr. Patrick Donohoe, PACS, BIDMC
Companion patient 3:
Stage IV HD on axial C-CT

Image courtesy of Dr. Patrick Donohoe,
PACS, BIDMC
Companion patient 3: Stage IV HD on axial PET/CT

Images courtesy of Dr. Patrick Donohoe, PACS, BIDMC

FDG-avid destructive bone lesion
Use of PET to stage Hodgkin’s

Patient PET scan; Centricity, BWH

Staging in HD

www.Lymphomation.org

+/- “B symptoms”: night sweats, fever, unexplained >10% weight loss

Patient AW therefore has stage IA Hodgkin’s --> 87% 10 year overall survival
Personal Paper

THE OPHelia SYNDROME:
MEMORY LOSS IN HODGKIN'S DISEASE

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Jane did it at school. It took a month before a conversation with a school teacher made me wonder, and ask to see some homework.

A golden summer crisped into fall. Jane and I sat night by night, with homework. If she brought it home, if she brought the right books, if she brought any books. By the middle of October we knew there was something seriously wrong. We realised when we looked at her work and talked to worried teachers that there was a serious problem — a teenage neurosis, perhaps drugs. The evening walk became more bizarre, like talking to an elderly person whose conversation
Summary

1. Presentation of patient AW, subacute onset of anterograde amnesia
2. Neuroanatomy at level of midbrain
3. Radiologic differential for temporal lobe MRI lesions
4. Discussion of Paraneoplastic Syndromes
5. Radiologic differential for anterior mediastinal mass
6. Use of PET for Hodgkin’s Disease Staging
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