Atypical Encephalitis from Ehrliciosis

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Presentation

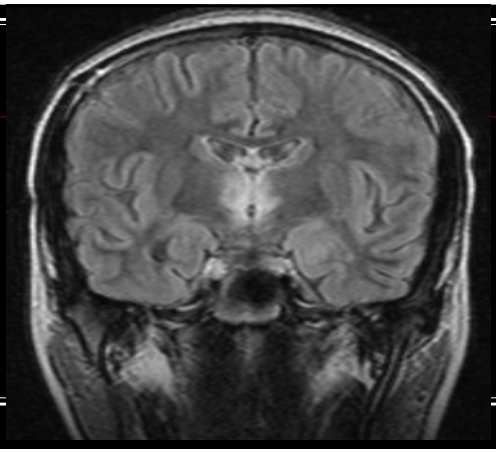
- 17-year old female with 2 month h/o progressive neurological impairment
- Began with episode of fever and malaise in summer
- Over few weeks fever subsided and ptosis appeared in left eye with diplopia

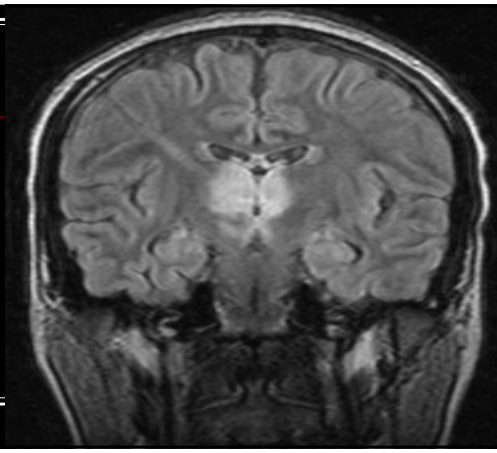
Presentation

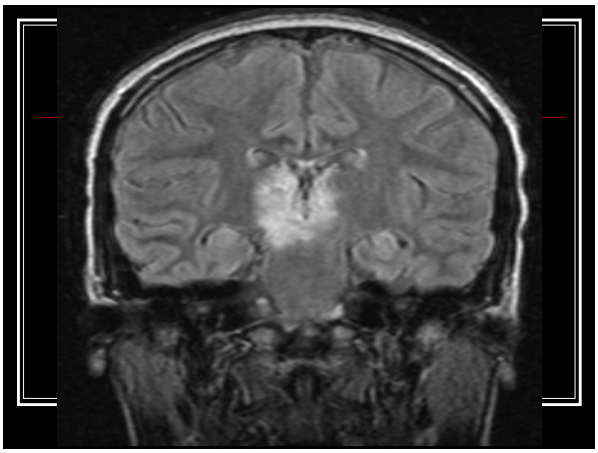
- Progression to right eye, dysconjugate eye\
- Left hemiparesis and numbness requiring maximal assistance with ambulation
- Lethargy, decreased level of alertness prompted ER visit to OSH

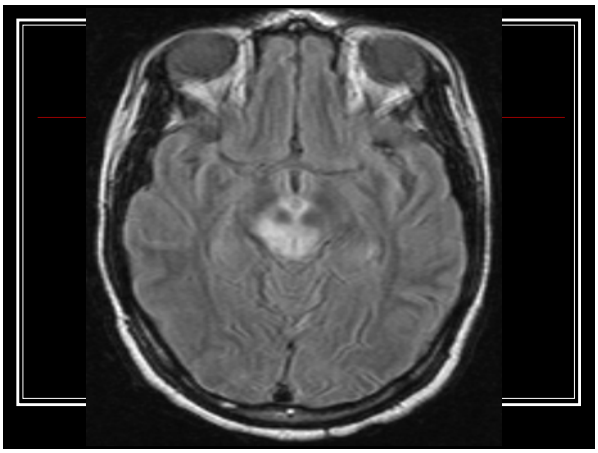
Presentation

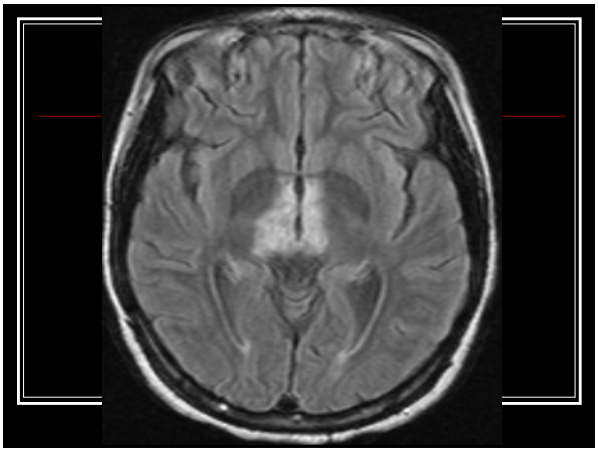
- No PMH (previous illness), PSH nor meds
- MRI obtained at OSH

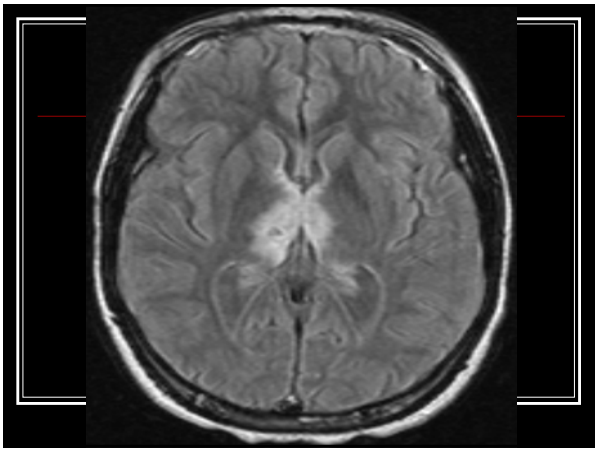


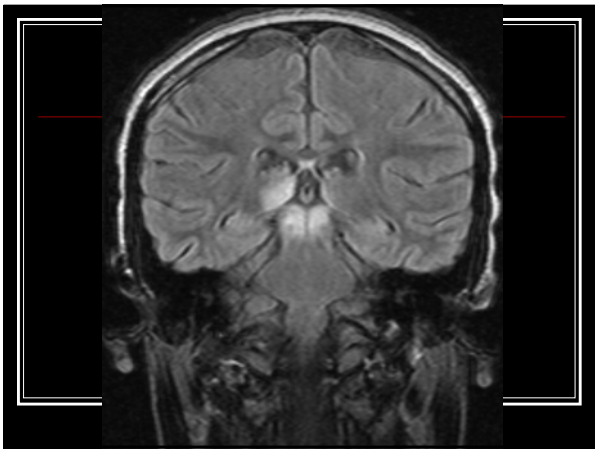












Imaging

- Increased T2 and FLAIR signal within midbrain centering on aqueduct
- Involving R>L tegmental regions and substantia nigra
- Hyperintensity extending into thalamus bilaterally (R>L), extending into genu and posterior limbs of internal capsule (R>L)

Imaging

- Hyperintensity of right posteromedial caudate head
- Pattern affecting deep venous system
- Gliotic reaction consistent with prior chronic DVT of internal cerebral vein (per Neuroradiology)

Physical Examination

- Somnolent and difficult to arouse but able to answer simple questions with slurred speech
- Bilateral ptosis, left gaze deviation in left eye, left facial droop and left facial hemianesthesia

Physical Examination

- F/C x 4, left hemiparesis, left hypertonicity, left-sided decreased sensation
- Afebrile throughout hospital stay

Treatment

- Stereotactic biopsy hospitalization day #2 of right thalamus
- Procedure well tolerated
- Samples sent

Pathology

- Specimen: hypercellular tissue with mildly pleomorphic astrocytes, chronic inflammation
- Perivascular lymphocytic infiltrates, parenchymal infiltration by macrophages
- Rod-shaped microglial cells, one possible microglial nodule

Pathology

- Edematous gray matter
- Gliosis

Laboratory Studies

- Decreased serum absolute lymphocyte count, elevated direct bilirubin
- Otherwise normal CBC, CMP
- CSF drawn on HD #3: no xanthochromia, no rbc's, 23 wbc's, 99% lymphocytes, 1% monocytes,
- Glucose 84, protein 60

Laboratory Studies

- Sexually active, no STD's, no travel, + dog with multiple tick infections
- Testing begun for Lyme, Tularemia, Ehrlichiosis, HSV
- Acyclovir and Doxycycline began empirically HD #5

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Laboratory Studies

- HD #9 PCR: HSV II
- HD #10: + Ehrlichiosis
- Regimen continued for 21 days

Course

- Daily improvement
- HD # 9: more awake, alert
- HD #10: improvement in eye adduction and strength
- HD #12: improvement in ptosis
- Discharged on HD #23 with residual left hemiparesis

Pathology

- Samples sent to CDC in Atlanta
- Perivascular mononuclear infiltrate, scattered macrophages, small foci of hemorrhage
- No intranuclear, intracytoplasmic inclusion bodies, immunohistochemical staining for HSV1 & 2 negative…..Ehrlichiosis

Discussion

- First case of Ehrlichiosis 1986
- Neurological complications rare
- Human monocytic ehrlichiosis: rickettsial infection caused by Gram – obligate intracellular bacteria, Ehrlichia chaffeensis

Discussion

- Infects mononuclear cells
- Major vector is Lone Star Tick
- 25% of cases, no recognizable bite
- 0.6/million, predominance in elderly
- 10% of infections in children
- 3% fatality rate

Discussion

- Most common April – September
- Spectrum of disease broad, mild febrile illness to death
- Most common presentation: undifferentiated fever
- Adult Sx: fever, HA, myalgia, nausea, malaise, rare AMS, varied skin rash

Discussion

- GI, resp, CNS involvement rare
- Peds Sx: rash, fever, HA
- 3.1% of children in SE and SC USA sero+ for HME – most infxn's quiet/mimic viral illness

Discussion

- Signs: elevated LFT's
- Cytopenias (thrombocytopenia, leukopenia)
- CSF: 25% pleocytosis, less commonly in pediatric population

Discussion

- DX: Serum IgG titers > 1:128 in presence of clinical dz
- Morulae (intracytoplasmic inclusions of coccobacilli) can be seen in leukocytes of peripheral smear, marrow aspirate, CSF

Discussion

- Pathology: diffuse perivascular infiltrates, tissue infiltration with monocytes
- Unique case: focal neurological findings
- CNS sx: vasculitis
- Treat with Doxycycline or tetracycline