



## Case Vignettes

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### Tumefactive Demyelinating Disease:

66 year old gentleman with recurrent cerebellar lesions



## History

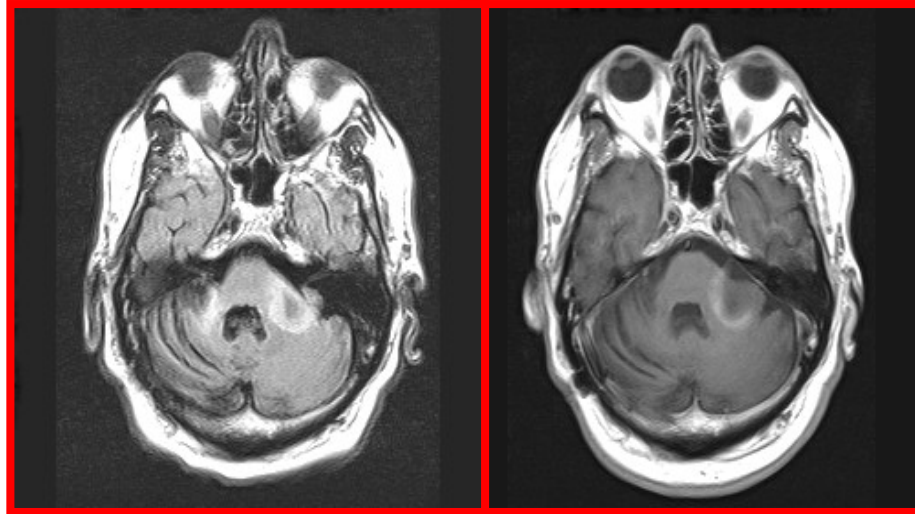
- 1995:
  - Numbness R face; dysarthria; difficulty writing due to R arm ataxia
  - MRI: large, isolated middle cerebellar peduncle lesion
  - Biopsy and radioRx initiated (2 Rx)
  - Aborted after path dx at Mayo of demyelinating disease



## History (cont)

- 1995-2004:
  - residual hemiataxia
  - no new symptoms
- 12/2004:
  - Identical symptoms on the left
    - facial numbness, dysarthria and left hemiataxia
    - MRI: new solitary tumefactive lesion





FLAIR

T1 with gad



## Course

- Attempted resection of new lesion
  - path “inconclusive”
- Review of radiology and clinical assessment at Mayo
  - probable demyelinating disease



## Questions

- Is diagnosis of tumefactive demyelinating disease correct?
- Can tumefactive disease of the CNS present in any location...supratentorial, brainstem, cerebellar?
- What is the risk of relapse?
- Does relapse tend to occur in the same location?
- Is the long remission in this case typical/atypical?
- Maintenance treatment?
  - which and why?



## Take Home Message

- Tumefactive MS may pursue a variable course:
  - no recurrence
  - typical MS
  - recurrent tumefactive presentation
- Consider tumefactive MS:
  - minimal mass effect
  - open ring sign
  - subacute presentation
  - young person
  - ANY mass lesion!



## NMO:

Do I have MS, NMO or both?

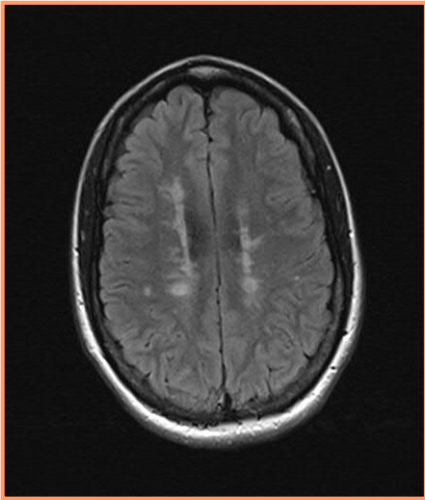


## 38 y woman Af Am

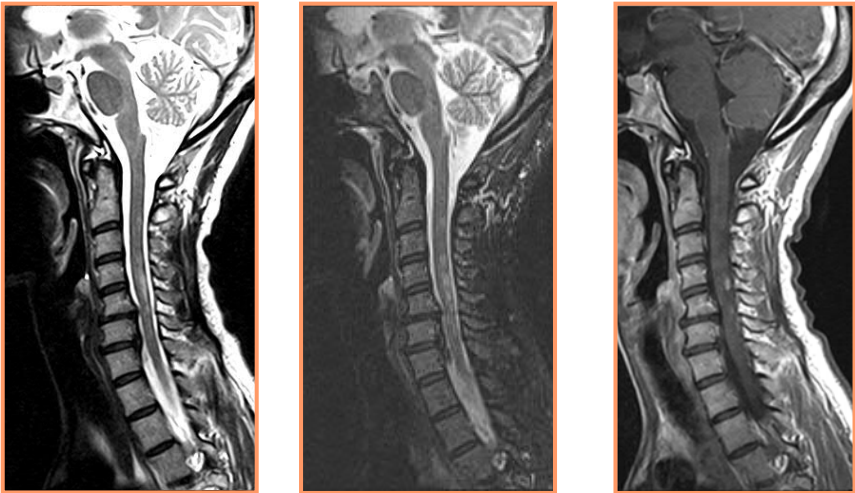
- Fall/2008: numb thighs X1wk—full recovery
- Dec/2008: after ice fishing, numb below breasts and severe weakness both legs
  - Good response to IVMP
  - Mild residual numbness and R leg weakness
- Jan/2009:
  - CSF 62 WBC, 93% polys; MBP 22 (nl <4); IgG index 1.44 (nl <0.8); 8 OCB
- PI: Breast ca 2000-in remission; ?SC trait
- FH: p uncle with MS



MRI brain: FLAIR



MRI Spine—T2, STIR, T1gad



The best diagnosis in this patient is:

1. Multiple sclerosis
2. Recurrent transverse myelitis
3. Neuromyelitis optica
4. Sickle cell-associated spinal cord infarction
5. None of the above



Serology for NMO

NMO-IgG+



## Differential diagnosis

### MS

- MRI head looks like MS
- CSF IgG index and OCB positive
- Never optic neuritis
- Younger than average NMO??

### NMO

- Symptoms confined to TM
- Long spinal cord lesion
- CSF pleocytosis with polys
- NMO-IgG+
- Very high MBP
- Af Am??



## The diagnosis is

1. MS
2. NMO
3. Both
4. Neither





## Solitary Tumefactive Lesion

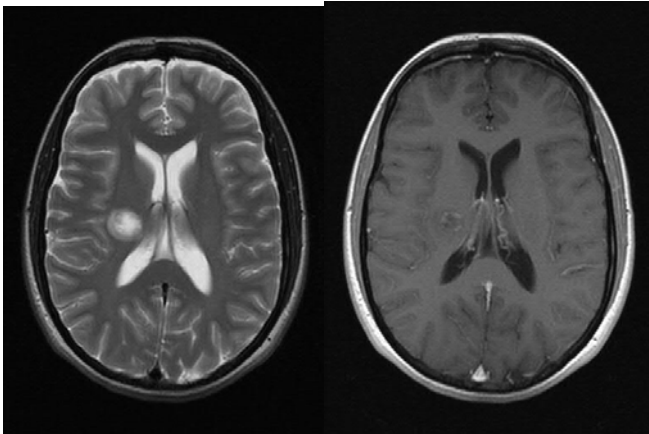


### 41 y woman

- 10/2010: subacute clumsiness L foot and hand followed by L facial droop
  - Symptoms fluctuate with level of fatigue
  - Good response to IV corticosteroids, followed by rapid return of symptoms
- No remote history of demyelinating disease events
- CSF: negative; nl IgG index/OCB
- PI: negative
- Exam: mild L hemiparesis and L Babinski sign

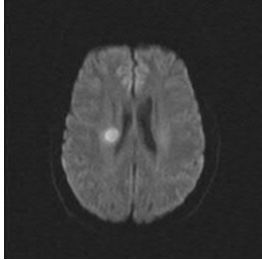


MRI 11/10/10

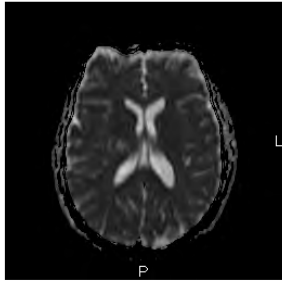


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DWI



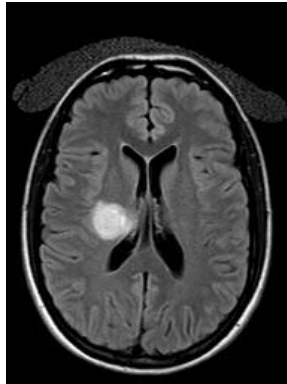
ADC



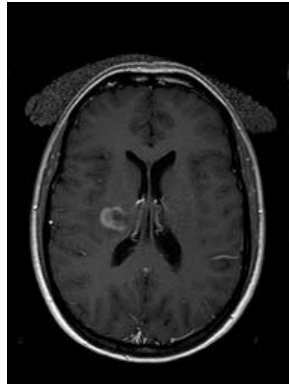
Worsening hemiparesis and persisting enhancement...



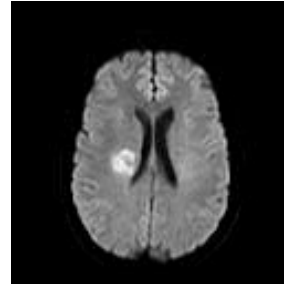
## MRI 12/6/10



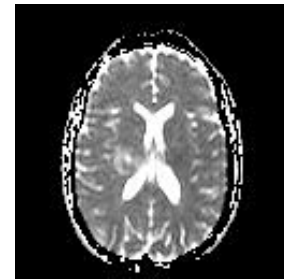
FLAIR



T1 with gad



DWI



ADC

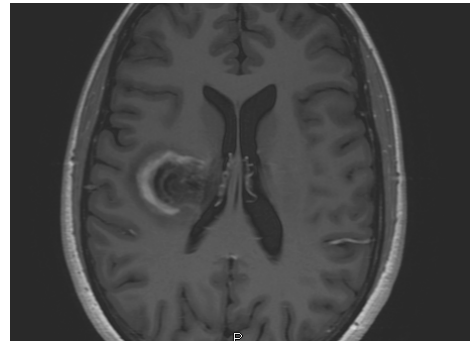
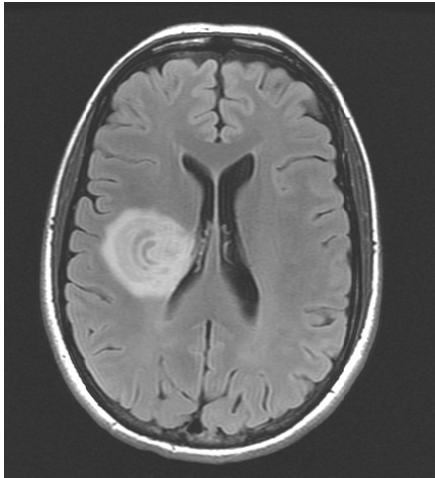


### At this point...

- Diagnosis?
- Workup?
- Treatment?



## MRI 1/4/11:



### At this point...

- Diagnosis: Is it certain?
- Treatment:
  - Acute?
  - Maintenance?
- Prognosis?



## Followup

- Treated with PLEX followed by cyclophosphamide IV X1 cycle
- No further treatment
- Followup MRI's:
  - Lesion shrank
  - No new enhancement
- Stable hemiparesis on no maintenance treatment

