Tumefactive MS

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• No conflicts of interest
Case

- 17-year-old female
- No significant PMH except for bilateral otitis media requiring antibiotics 1 month prior to onset of neurological symptoms
• Early May 2015: Consulted to a local hospital for 2 weeks of progressive severe headache, nausea/vomiting, mild gait impairment, mild right hemiparesis and difficulty with vision focus that even led to a fender-bender

• Exam: Normal alertness. Rest of exam also normal except for suspected mild field cut on right visual field and reflexes brisker on right side. Normal alertness
• 3.8 x 2.4cm predominantly cystic parietooccipital lesion with open-ring enhancement and surrounding vasogenic edema

• IV steroids and transferred to Children’s Hospital
Hospitalization Children’s 5/7 – 5/14

- MRS: in favor of a demyelinating etiology (elevated lactate peak, mild decrease in NAA and mild elevation choline/creatinine ratio within enhancing wall)

- MRI total spine: unremarkable

- CSF: OP 26 cm H₂O, protein 61, 1 RBC / 2 WBC, IgG index 0.53, no OCB.

- Blood work: vitamin D level 12
• Discussion with neurosurgery and neuroradiology and decision to hold off biopsy

• Hospital course notable for full resolution of headache and improvement of visual symptoms (treated with IV steroids then oral)

• Discharged home with oral prednisone taper for 14 days. Headache reoccurrence just after stopping prednisone; restarted, slower taper

• Short term follow-up with Dr. Wundes
On follow-up in June and early July, she continues to improve and has rapidly become fully asymptomatic.

Normal neuro-ophthalmology exam on 7/2 (including HVF).

Diagnosis at this point: presumed CIS.

Possible lower risk CDMS in context of negative CSF and spine MRI?

Started on vitamin D supplementation and decision to hold off DMT.

FU early September with same day MRI.
Tumefactive demyelinating lesions (TDL)

- Definition: large (>2 cm) acute lesion(s), often associated with edema and/or ring enhancement

- Other terms: tumefactive multiple sclerosis
Presentation

- TDL are very rare in MS and occur in the early stage

- Variable presentation

- Impairment may be very slight even in a patient with a massive lesion, while confusion, hemiparesis, or neglect syndrome can be seen in another patient with a lesion that appears no different

- Rare cases have required an emergency craniotomy
Largest series

- 168 patients with biopsy-confirmed TDL
- Median age at onset 37 yo
- Presentation typically polysymptomatic: motor, cognitive, sensory, and cerebellar symptoms most frequent
- Brain MRI: Lesions often multifocal. Median size of the largest lesion 4 cm. Enhancement > 50% of the lesions (ring, heterogeneous, and homogeneous patterns)
- Clinical course at last follow-up: Monophasic 24%, relapsing remitting 51%
- Final diagnosis at last follow-up: Multiple sclerosis in 79% and an isolated demyelinating syndrome in 14%

Canadian retrospective study

- 30 patients with TDL over 15 years
- 70% female; mean age at onset 41
- 82% polysymptomatic onset; median EDSS of 4
- 98% of lesions enhanced; mean size of well-defined lesions was 2.7 x 2.4 cm
- 87% treated with high-dose steroids and 75% improved. 23% received PLEX and 13% cyclophosphamide due to poor steroid response
• 73% partial and 27% complete recovery. Median EDSS at follow up 1 in those with a single event, but 6 in those with recurrent TDLs

• Radiologically, 89% of lesions decreased in size and 82% had resolution of enhancement

• 1st demyelinating event in 83%. Total 45% MS (after mean follow-up 2.7 years)

• 20% had recurrent TDL episodes
TDL and fingolimod

• TDLs might be seen more frequently during fingolimod treatment in particular, in patients with previous natalizumab treatment; 16 cases published

• Cause: fingolimod-induced redistribution of immune cells in susceptible individuals (decrease in regulatory cells circulating in the blood + an unusual population of effector cells in the CSF)

• Mostly upon starting fingolimod; estimated risk 1/10,000

Tumefactive MS lesions under fingolimod; http://www.ncbi.nlm.nih.gov/pubmed/24097813
Biopsy proven tumefactive multiple sclerosis with concomitant glioma: case report and review of the literature

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We report a case of pathologically confirmed tumefactive multiple sclerosis (MS) followed shortly thereafter by the diagnosis of an oligoastrocytoma. The complexity of diagnosis and management of concomitant presence of tumefactive MS and glial cell tumors is discussed.
Take-home message

• TDL is rare but important to consider it in the right time.

• Not all cases will develop CDMS (45-75%).

• Tumefactive lesion(s) at MS onset not predictive of a more severe or protracted MS course.

• TDL recurrence is possible (20%).

• Increased risk with fingolimod?