Title: Tumefactive Demyelinating Lesion (TDL) of Spinal Cord due to Aquaporin-4 Ab

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Objective: To report a case of Neuromyelitis Optica (NMO) that presented as a tumefactive demyelinating lesion (TDL) in spinal cord.

Background: TDLs are lesions that do not exhibit classic presentation and are indistinguishable from neoplasm on imaging studies. TDLs are rare and typically seen in brain of patients with Multiple Sclerosis (MS). Seven case reports of an isolated spinal cord TDL exist in the literature, but the occurrence of spinal cord TDL in NMO has not previously been reported.

Design: A 43 year old woman with morbid obesity and developmental delay presented with progressive right leg and handgrip weakness of 2 weeks duration. On MR imaging, increased T2 signal was noted in cervical spinal cord from C5 to C7 vertebral level with poorly demarcated boundaries, focal enlargement at C6-C7 and homogeneous enhancement. The imaging features were consistent with a neoplasm such as astrocytoma or ependymoma and myelitis was thought to be unlikely. For definitive diagnosis, she underwent a C5-C7 posterior laminectomy with biopsy of the lesion that showed inflammatory macrophages with myelin and related products, consistent with demyelination. MRI Brain showed an occasional periventricular FLAIR hyperintensity. She was treated with pulse high dose steroids with good improvement and was lost to follow up.

Results: She presented with a relapse involving her cervical spinal cord 2 months after her initial presentation and was treated with high dose steroids and plasmapheresis. Her work up for MS mimics, drawn during her initial admission, was remarkable only for a positive Aquaporin-4 antibody. Rituximab was started for disease modification. We continue to follow the patient.

Conclusions: TDLs are rare and usually occur in the brain of patients with MS. TDLs are more common in women, with an average age of presentation of 37. Their occurrence in spinal cord has been reported in MS but not in NMO. TDLs pose a diagnostic challenge and patients usually undergo a biopsy/excision to obtain a definite diagnosis. Advanced imaging in the form of MR perfusion and MR spectroscopy may be helpful in diagnosis of patients with TDL in the brain and in avoiding surgical intervention. However, in the cord due to decreased tissue volume these tests are not helpful. Relapsing remitting course, presence of periventricular lesions in brain and response to steroids might help in diagnosis. Differentiating these lesions from tumors is important to avoid erroneous surgical procedures and treatment like radiation which can worsen demyelinating disease and puts patients at risk for secondary malignancies.