A rare imaging finding in CNS demyelinating disease : Wallerian degeneration of corticospinal tract.

Nidhiben Anadani MBBS, Megan Hyland MD, Andrew Goodman MD, Lawrence Samkoff MD. University of Rochester, New York

Background

Wallerian degeneration (WD) in the central nervous system (CNS) is a well known phenomenon though the process is slow in the CNS as compared to the peripheral nervous system. WD has been documented commonly after stroke but rarely in multiple sclerosis (1).There have been few cases reported of WD after acute demyelinating event (2) in patients at risk of having MS presenting with a clinically isolated syndrome.

Discussion

- Exact mechanism of WD in demyelinating disorder is not known, axonal damage can be one factor (3).
- Corticospinal tract lesions have also been described in neuromyelitis optica spectrum disorder (4). These are generally thought to present with clinical symptoms .
- Although our patient did not have worsening of symptoms she tested positive for anti AOP4 antibody. We suggest anti AQP4 and anti MOG antibody should be tested in patients with longitudinally extensive corticospinal tract lesion.
- WD is thought to be a poor prognostic marker for recovery (1) though our patient-1 had no deficits.

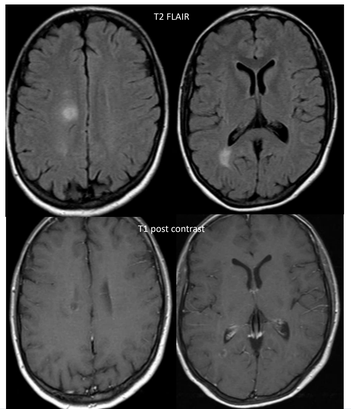
References

1)MRI demonstration of Wallerian degeneration in various intracranial lesions and its clinical implications vijay Sawlania et al, Journal of the neurological Science. March 10,1997 Volume 146, Issue 2, Pages 109-116
2) A Wallerian degeneration pattern in patients at risk for MS J.H. Simon, R.P. Kinkel, L. Jacobs, L. Bub and N. Simonian. Neurology March 14, 2000; 54 (5)
3) Wallerian Degeneration: A Major Component of Early Axonal Pathology in Multiple Sclerosis Tomasz Dziedzic,Imke Metz,obias Dallenga,Fatima Barbara König, Sven Müller, Christine Stadelmann, Wolfgang Brück. Brain pathology. Volume 20, Issue 5 Pages 877–998
4) MRI characteristics of neuromyelitis optica spectrum disorder An international update. Ho Jin Kim,MD, PhD et al, Neurology. 2015 Mar 17; 84(11): 1165–1173

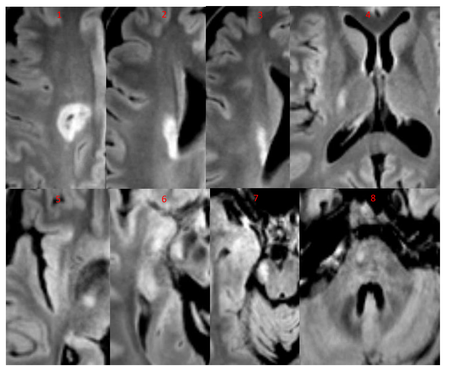
CASE 1

A 32-year-old female woke up with pulling sensation on left side and left facial droop. Examination showed subtle left sided weakness. Brain MRI showed 2 periventricular T2 hyperintese lesions, one in the right centrum semiovale which showed restricted diffusion and subtle contrast enhancement and another in the right parietal lobe which showed open ring enhancement. Cerebrospinal fluid (CSF) was negative for oligoclonal bands. Serum AQP-4 antibody was negative by cell based assay (CBA), ANA was positive (1:80). She did not have lesions in spinal cord. She was started on treatment for MS with Interferon beta-1b. Follow up MRI 7 months after presentation showed T2 hyperintensity along right corticospinal tract from the centrum semiovale to the pons. Examination was normal. One year after initial presentation serum AQP4 antibody was positive by ELISA but repeatedly negative by CBA.

MRI at presentation



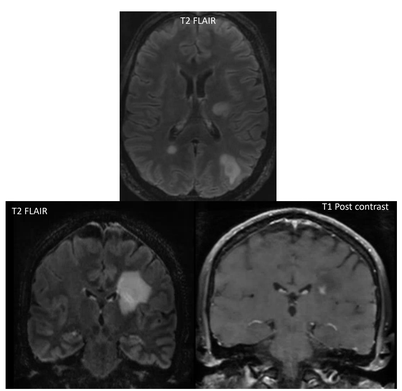
7 months after initial presentation



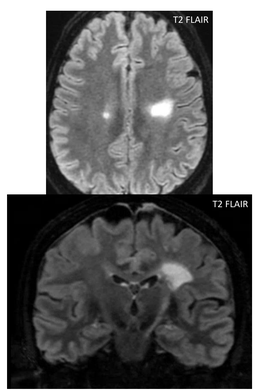
Sequential T2 FLAIR images

CASE 2

A 46-year-old male with history of high blood pressure presented with acute onset right sided weakness and dysarthria. Examination showed right facial droop, right arm plegia and right leg paresis. MRI of brain showed 3 large ovoid T2 hyperintense lesions located in left corona radiata, left occipito-parietal lobe junction and right occipital lobe with patchy contrast enhancement. 2 of the lesions showed concentric ring appearance. CSF was negative for oligoclonal bands. Serum AQP4 antibody was negative. His weakness improved after 5 days of high dose intravenous steroid and plasma exchange. Follow up scan in 9 months showed one new periventricular lesion without enhancement and T2 hyperintense signal along the left corticospinal extending to the ponto-medullary junction. His examination at follow up was significant for right spastic hemiparesis with a spastic gait. With dissemination in time and space he was diagnosed of MS and started on glatiramer acitate.



MRI at presentation



9 month after initial presentation