Recurrent tumefactive Multiple Sclerosis: a case report.

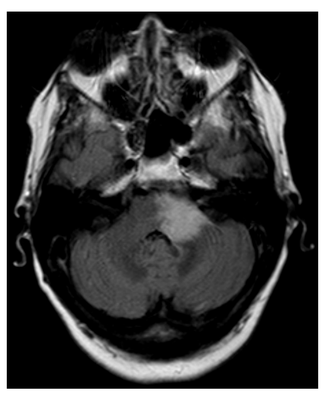
Girelli F., Zagaglia S., Fiori C., Jorio G., Carlini G., Logullo F., Di Bella P., Provinciali L.

Marche Polytechnic University – Department of Experimental and Clinical Medicine

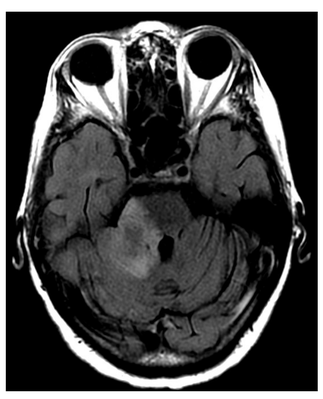
Introduction

Multiple Sclerosis (MS) is a chronic autoimmune inflammatory disease of the central nervous system (CNS) characterized by multifocal demyelination with progressive neurodegeneration.

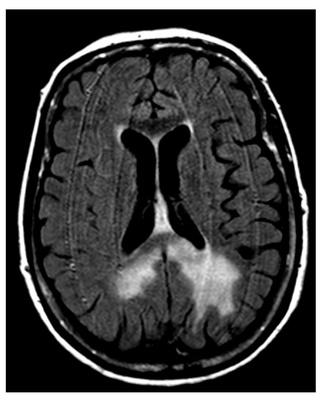
Tumor-like presentation of MS consists of a large single plaque (>2 cm) with edema and mass effect.



2012; MRI: FLAIR sequence. Abnormal signal in the left middle cerebellar peduncule.



2013; MRI: FLAIR sequence. Abnormal signal in the right middle cerebellar peduncule.



2014; MRI: FLAIR sequence, abnormal signal in the splenium of corpus callosum.

Case presentation

We present the case of a 67-years old caucasian right-handed woman who came to our attention in April 2013 for the acute onset of paresthesias in the right half of her tongue and in the right perioral region, with dyplopia and dysphagia. In May 2012 she was admitted to another hospital for a vertiginous syndrome with perioral paresthesias, she was diagnosed with “ischemic stroke”. Several brain scans performed in 2012 showed an area of abnormal signal in the left middle cerebellar peduncule extending to part of the brainstem, suspiscious for a late infarction. While in our Clinic she underwent routine blood tests, immunological screening, factor V Leiden mutation test, all were normal. A CT scan of the brain and a CT angiography ruled out vascular malformations and stenosis. An MRI scan showed hyperintensity in the left middle cerebellar peduncle with no contrast enhancement and a new area of abnormal signal in the right middle cerebellar peduncle extending to the pons and right cerebellar hemisphere suspicious for inflammatory origin. A CSF analysis showed high proteins, damaged blood-brain barrier and polyclonal CNS synthesis of IgG. The patient underwent a high dose steroid therapy, with clinical benefit. An MRI scan performed three months after discharge showed reduced size of the right lesion. Azathioprine was started. In September 2014 she showed new neurological symptoms: cognitive disorders and visual deficits. A new MRI scan showed an area of abnormal sign in the splenium of corpus callosum, extending to the periventricular white matter, with sourrounding edema, two other foci of abnormal signal intensity were present in the cerebellar peduncles, so the patient was admitted again to our Clinic and an MRI performed during her stay confirmed the lesions, suggestive for inflammatory nature or lymphoma. A new CSF analysis showed no new findings. She then underwent a cerebral biopsy in the left occipital lesion with findings consistent with demyelinating disease in acute tumor-like phase. Therapy with mitoxantrone was started.

Conclusion

We described the case of a remitting-relapsing tumefactive Multiple Sclerosis, a diagnostic challenge that can clinically and radiographically mimic several conditions such as ischemic stroke, brain lymphoma or abcess, requiring sometimes a brain biopsy.

References

- Sharmilee Gnanapavan, Zane Jaunmuktane, Kelly Pegoretti Baruteau, Sakthivel Gnanasambandam and Klaus Schmierer12. A rare presentation of atypical demyelination: tumefactive multiple sclerosis causing Gerstmann’s syndrome. BMC Neurology 2014, 14:68 doi:10.1186/1471-2377-14-68 Singer MA. et al. Primary lateral sclerosis. Muscle nerve. 2007 Mar; 35 (3):291-302.
- Goldenberg, MM. Multiple Sclerosis Review. P T. 2012 Mar; 37(3): 175–184.
- Selkirk SM1, Shi J. Relapsing-remitting tumefactive multiple sclerosis.Mult Scler. 2005 Dec;11(6):731-4.