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Neuromyelitis Optica Spectrum Disorder associated to Parsonage Turner Syndrome. Casual or causal connection? A case report.

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Background

Neuromyelitis Optica Spectrum Disorder (NMOSD) is an inflammatory demyelinating disease of the central nervous system (CNS), often associated to *Anti-Aquaporin4-Antibodies* (AQP4-Abs). Recently, *anti-Myelin-Oligodendrocytes-Glycoprotein-Antibodies* (MOG-Abs), an Ab targeting oligodendrocytes rather than astrocytes, has been associated with a broad spectrum of acquired human CNS demyelinating diseases. **Parsonage Turner Syndrome** (PTS) is an idiopathic brachial plexus neuropathy for which immunologic, infectious and genetic etiology have been proposed.

Case Report

A 49 years old male experienced in **1998** and in **2004** a *PTS of the left and right arm respectively*. Both episodes *resolved with steroids*.

In **January 2009** he developed *fever, headache* and later *acute urinary retention, tetraplegia and bilateral amaurosis*. Brain and spine MRI findings were referred to *"encephalitis with leptomeningitis and ependymitis"* associated to a long lesion of the spinal cord. BBB damage and increased lymphocyte count were found at CSF analysis . No virus or bacteria were detected by PCR and Ab testing. Treatment with *acyclovir, i.v. methylprednisolone and Intravenous Immunoglobulin* (IVIG) led to *partial recovery* (sphincteric disturbances, spastic paraparesis).

In **January 2013** the patient developed *fever, gait abnormality and sphincter disturbances*, which *recovered after i.v. methylprednisolone.*

In **November 2016** he developed a new episode of *left PTS*.

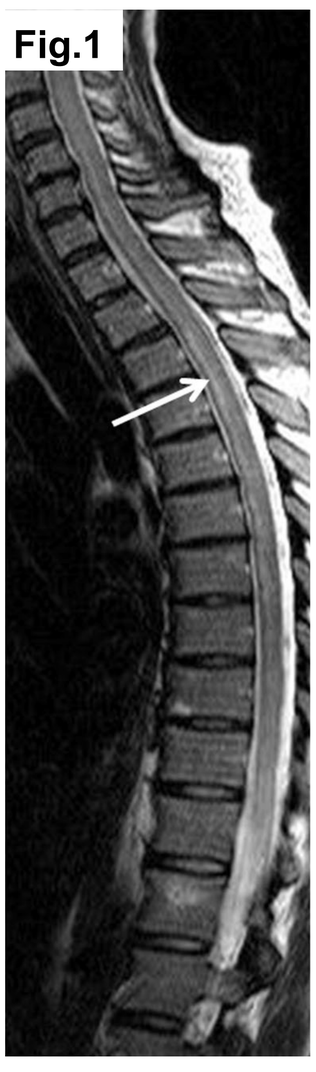
Fig.1

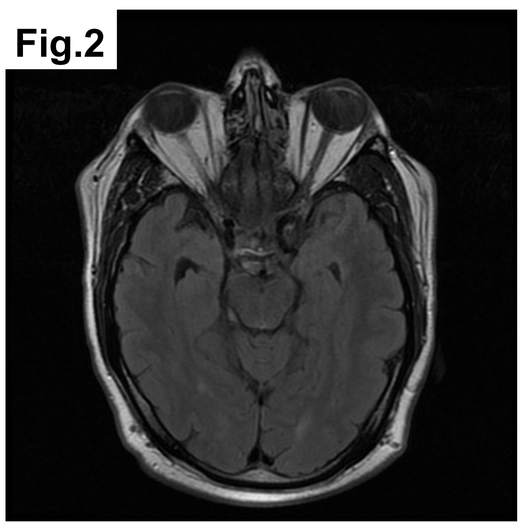
Fig.2

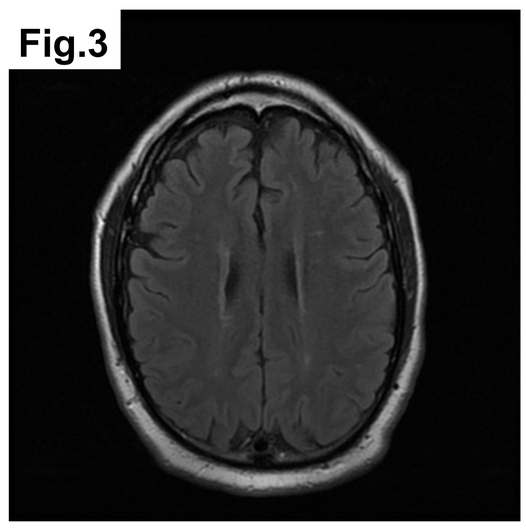
Fig.3

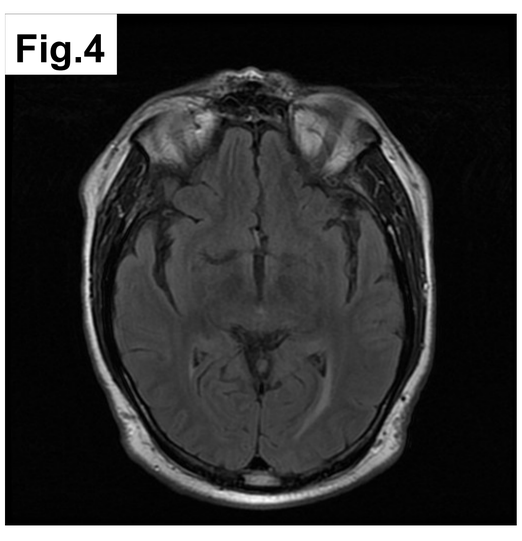
Fig.4

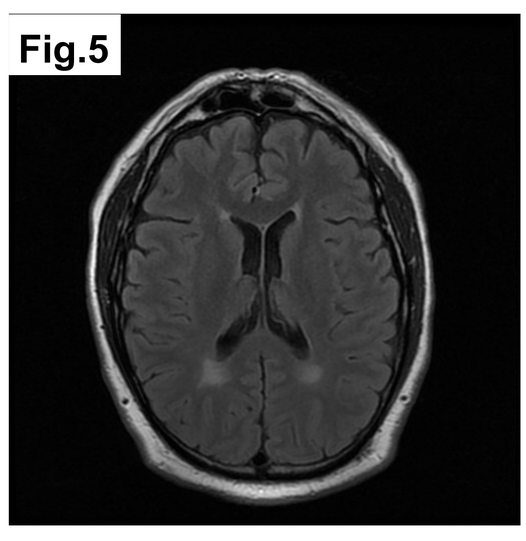
Fig.5

He was then admitted to our Department for further analysis. Neurological examination showed *spastic paraparesis, sphincteric disturbances and mild paresis of the left arm*.

MRI scans reported *atrophy of the optical chiasma, multiple white matter FLAIR-hyperintense lesions in corpus callosum, frontal and parietal periventricular, posterior peritrigonal and temporal pole bilateral, beside a longitudinally extensive spinal cord not enhancing lesion* (Figure 1-5).

Visual evoked potentials were bilaterally abnormal. Electromyography showed signs of previous bilateral brachial plexopathy. A new CSF analysis showed an increase of BBB damage. *Serum MOG-Abs were positive, while Anti-Aq4 and anti-NMO negative*. Patient underwent *Rituximab* treatment (720 mg/week for four weeks); a *slight improvement of spastic paraparesis* was observed.

Conclusion

Diagnosis of our patient remain a clinical dilemma. Actually, *nor criteria for diagnosis of multiphasic ADEM nor that for NMOSD are satisfied*. Furthermore, the association of two rare autoimmune mediated diseases as PTS and NMOSD raises the possibility of a unique spectrum of disease including both. Even if peripheral nerve damage is rarely described in ADEM or NMOSD *a direct implication of MOG autoimmunity or a spreading of the antibodies response towards other antigens (i.e. neurofascin)*, need to be taken into account. Therapy targeting the B cells seemed to us the best choice in this patient.

References

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