Rapidly progressive visual worsening in a patient with multiple sclerosis (pwMS): an insight into progressive disease.

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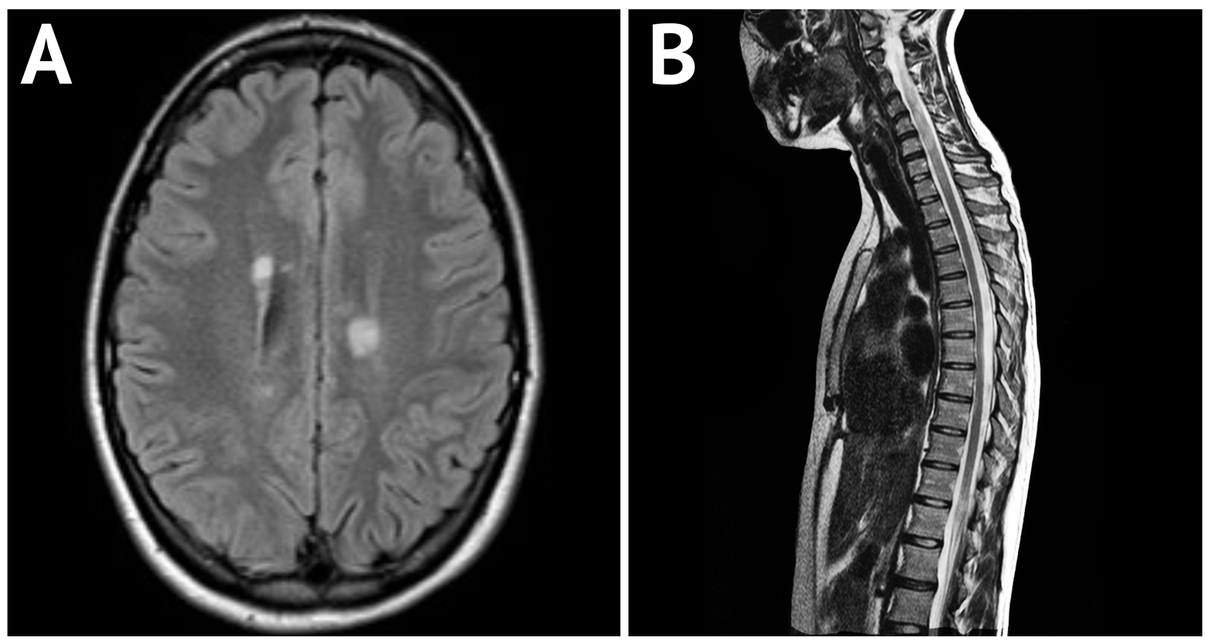
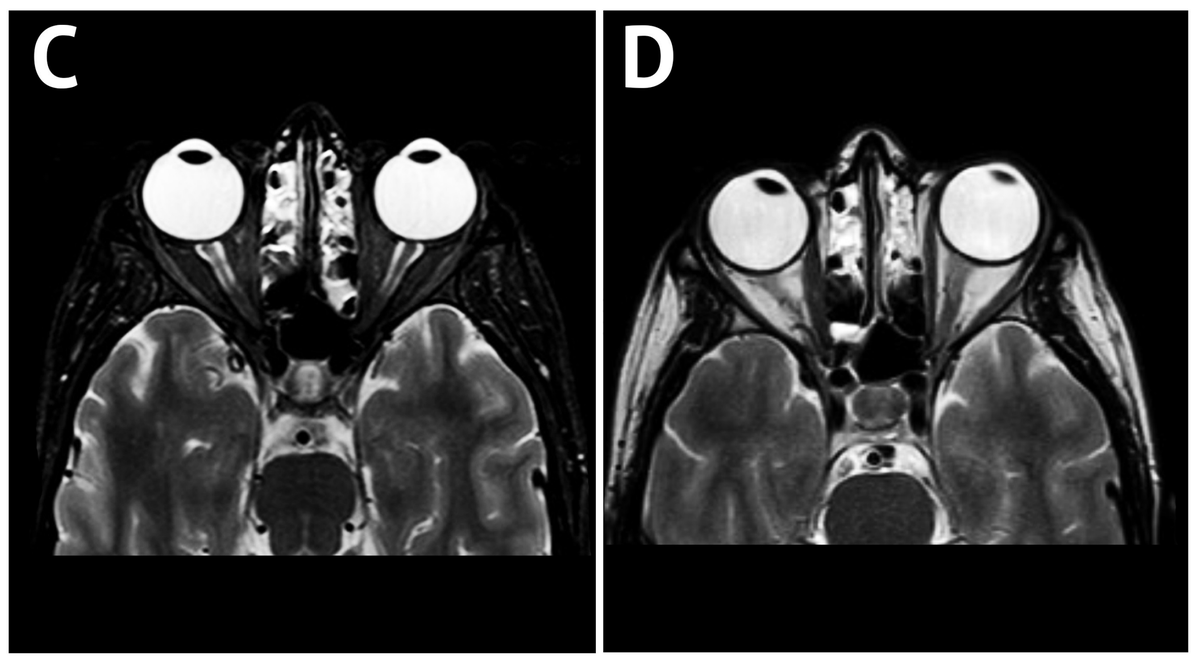
Introduction

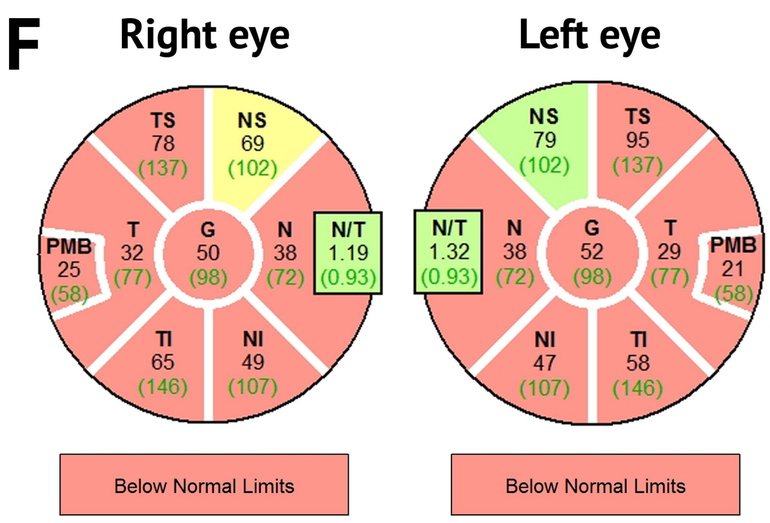
The recent revisions to the clinical course definitions have not defined the transition point, when relapsing-remitting Multiple Sclerosis (MS) becomes secondary progressive. Disease progression in MS is mainly based on motor function impairment. So far, to our knowledge, no cases of rapid disease progression involving the visual system have been reported in pwMS.

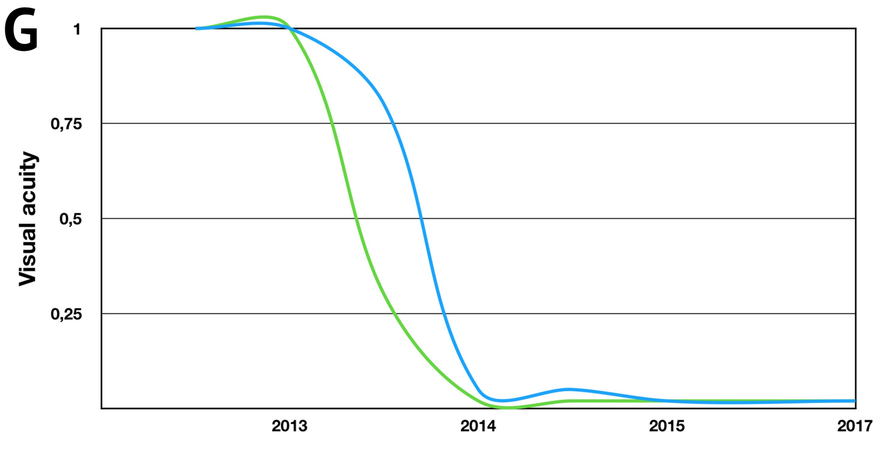
Case report

We report the case of a 27 years-old woman. At the age of 23, she experienced the acute onset of diplopia and dizziness. Magnetic Resonance Imaging (MRI) showed inflammatory demyelinating lesions both in the brain and the spinal cord (fig. A-B). Cerebrospinal fluid analysis was positive for oligoclonal bands. Methylprednisolone was administered, with complete remission. The patient fulfilled diagnostic criteria for MS and she started Interferon Beta and, after a new relapse, Natalizumab. In December 2013 she developed slowly progressive lower limbs spasticity with mild weakness. In May 2014 a subacute progressive visual impairment in both eyes emerged, without ocular pain. Her visual acuity reached 1/20 in the right eye and 1/50 in the left eye in 6 months. Brain and spinal cord MRI showed no signs of disease activity. MRI of the orbits documented T2/FLAIR hyperintensity of central fibers in both optic nerves, with mild atrophy in intraorbital segments (fig. C-D). At Optical Coherence Tomography (OCT) peripapillary Retinal Nerve Fiber Layer and Ganglion Cell Complex thickness were diffusely reduced in both eyes (fig. F). Anti-acquaporin-4 antibodies were negative. Leber Hereditary Optic Neuropathy was excluded by mitochondrial DNA sequencing. Her motor worsening constantly progressed too and EDSS in May 2017 was 5.5. Her visual acuity never recovered and serial MRI controls showed a substantially stable burden of disease (fig. G).









Discussion

Progressive MS involves not only the spinal cord in parallel with the motor function, but also brain grey matter and visual system, as shown by recent MRI and OCT studies, but, from a clinical point of view, motor worsening usually emerges first. However, as we describe in our case, the worsening may be atypical, preferentially affecting one of these systems to a greater extent and thus manifesting with prevalent clinical damage in one specific function. The peculiarity of our case is based on the visual system progressive worsening.

Conclusion

Irreversible worsening in addition to onset of progression is an open topic in MS and its early detection could be important for a specific early therapeutic approach. This case may contribute to shed light on definition of progressive MS and on the underlying different pathogenetic mechanisms.