Dementia Revealing Multiple Sclerosis
Salma Sakka, Nouha Farhat, Amal Bouhamed*, Hanen Hadj Kacem, Olfah Hdiiji, Mariem Damak Chokri Mhiri
Department of Neurology, Habib Bourguiba Hospital, Sfax, Tunisia

Abstract
Cognitive abnormalities are frequently observed in advanced stages of Multiple Sclerosis (MS). Their frequency increases from clinically isolated syndromes, to relapsing-remitting and secondary progressive MS. The most frequently impaired functions are information processing speed, attention and memory. Dementia is uncommon but may reveal a MS. We reported a 47-year-old woman suffering since 8 months of headache with a progressive amnesia, temporo-spatial disorientation and behavioral disorders. On examination, we found dementia syndrome and a frontal syndrome. Brain MRI showed a T2 hyperintensity multifocal ovoid lesions with enhancement in the periventricular white matter and in the corpus callosum. Diagnostic hypotheses were ADEM or MS. Isoelectric focusing revealed oligoclonal bands and elevated IgG index. She was treated with intravenous methylprednisolone 1 g daily for five consecutive days. There was a regression of the symptomatology but 4 months after diagnosis, the patient presented with a postural instability, gait difficulty, terminal intention tremor and dysmetria. A diagnosis MS was considered and she was given interferon. The concept of MS being a dementing illness is not novel. Neurologists are well aware that patients with MS may present with cognitive impairment. But the fact that this disease is revealed by dementia is rare and new.

Keywords: Multiple sclerosis; Acute disseminated encephalomyelitis; Dementia; Syndrome frontal; Brain Perfusion by Single photon emission computed tomography

Introduction
Cognitive abnormalities are frequently observed in advanced stages of Multiple Sclerosis (MS). Their frequency increases from clinically isolated syndromes, to relapsing-remitting and secondary progressive MS. The frequency of dementia in MS remains indeterminate in the absence of validated diagnostic criteria and prospective study on its prevalence [1]. Indeed, it does not appear explicitly in the dementia criteria of the DSM-5 [1]. So that dementia remains little known in MS but may inaugurate this disease. We reported a case of MS revealed by dementia.

Case Report
A 45-year-old woman with no significant medical history, particularly infectious or recent vaccination, was admitted in April 2011 for eight months history of episodes of temporal and spatial disorientation, behavioural disorders and rapidly progressive anterograde amnesia which affects her daily life.

The neuropsychological assessment showed a dementia syndrome with a Mini Mental State Examination (MMSE) score of 22/30, a temporal-spatial disorientation, episodic memory disorder unimproved by indication and objectified by the 16-item Free and Cued Recall (RL/RJ16), severe dysexecutive syndrome (Frontal Assessment Battery (FAB)=4/18), verbal fluency with an important attention deficit disorder. The rest of the examination showed a frontal syndrome without sensorimotor deficit.

Brain MRI showed multiple hyperintense and oval lesions on T2 and FLAIR- weighted sequences. These lesions were present in the periventricular and juxtacortical white matter, and corpus callosum with the largest in the right frontal lobe, measuring 3 cm in diameter (Figure 1).

Cerebrospinal Fluid (CSF) analysis showed an inflammatory profile with intrathecal synthesis of IgG: 2 white blood cells, total protein concentration=0.23 g/L, gamma globulin elevation=13.8%, IgG index=0.92, detection of Oligoclonal Bands (OCB).

The standard blood test and the sedimentation rate value were normal. Immunoassay (Antiphospholipid antibodies, Anti-native DNA antibodies, Anti Neutrophil Cytoplasmic Antibody, Antithyroid antibodies) were negative. HIV, Lyme, VDRL-TPHA, serology and sputum specimens for Tuberculosis testing were negative.

Acute Disseminated Encephalomyelitis (ADEM) was suspected. The presence of an oligoclonal profile of the CSF and the absence of gray matter involvement had not allowed to retain this first diagnosis. The second suggested diagnosis was the multifocal Clinically Isolated Syndrome (CIS). The patient received intravenous corticosteroids (1 g daily for 5 consecutive days). There was an improvement in cognitive impairment (MMSE score=29/30) and radiological aspect (Figure 2). The patient was not treated by immunomodulatory therapy later.

In September 2011, she had admitted for a postural instability, gait difficulty, terminal intention tremor and dysmetria. The examination showed an MMSE score again at 20/30, a quadripyramidal syndrome and a static and kinetic cerebellar syndrome. The EDSS score had increased to 6. A second MRI brain and cervical spine showed new demyelinating lesions with enhancement in the corpus callosum, cerebellum and cervical spinal cord. The diagnosis of relapsing-remitting MS was retained. The patient received methyl-prednisolone (1 g daily) for 5 days, which again improved clinical signs except for memory disorders. Then the patient was treated with interferon beta-1b. Faced with the persistence of memory difficulties, a requested Brain Perfusion by Single Photon Emission Computed Tomography (SPECT) had objectified a moderate hypoperfusion of the left parietal association cortex extending towards the parieto-temporal crossroads.

*Corresponding author: Amal Bouhamed, Department of Neurology, Habib Bourguiba Hospital, Sfax, Tunisia, Tel: +216-74106047; E-mail: amal_bouhamed@hotmail.fr

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Figure 1: Brain MRI (04/2011): Axial fluid-attenuated inversion recovery (FLAIR) (a,b) and post contrast T1 weighted (c,d) images of hyperintense and oval lesions in the periventricular. Sub-cortical and corpus callosum structures with the largest in the right frontal lobe. Measuring 3 cm in diameter and annular enhancement of most lesions.

Figure 2: Brain MRI (09/2011): Axial (FLAIR) (a,b) and post contrast T1w (c,d) images showing a regression of hyper intense lesions of the white matter with absence of enhancement.

Figure 3: Brain Perfusion SPECT showing hypo perfusion with thinning of the right parietal association cortex.

(Figure 3). Since May 2012, there has been a progressive deterioration of cognitive impairment, balance disorders and EDSS score (=5). The diagnosis of secondary progressive MS was therefore retained.

Discussion

Cognitive impairment in the "multinodular sclerosis" was described in 1868 by Charcot. These disorders affect 40 to 60% of patients with MS. They are present from the beginning of the disease and seem to develop faster than the motor disorders [2]. The main cognitive disorders detected concern attention, working memory, speed of information processing and verbal fluency [3]. Dementia is rare (<2% of patients followed for MS). Severe cognitive impairment inaugurating MS with sometimes acute or subacute onset as in our case have been reported [4]. These patients are usually older than those with early
motor signs. The evolution of cognitive forms of MS is marked later by the appearance of a pyramidal syndrome, cerebellar syndrome as in our case or a cranial nerves lesion [5]. MS starting with cognitive impairment may progress to a relapsing-remitting or progressive form. However, physical disability remains weakly correlated with the degree of cognitive decline. The cognitive profile is often subcortical. Aphasia, agnosia, alexia and apraxia (characteristics attributed to cortical dysfunction) are exceptional in MS [6]. The main factors causing cognitive dysfunction are the existence of a diffuse brain injury and later the evolution of cerebral atrophy. The morphological and functional imaging can detect these factors [7]. The different studies performed in conventional MRI (lesion load) and Magnetization Transfer Imaging (MTI) show a significant correlation between cortico-subcortical cerebral atrophy, the volume of cortical lesions and cognitive impairment than between this one and the white matter lesion load [8]. Functional imaging such as Brain Perfusion SPECT and functional MRI have their places. According to studies, Brain Perfusion SPECT will show cortical and white matter hypoperfusion. Cortical hypoperfusion results from neuronal death after axonal damage or vascular steal syndrome. It is more important in progressive forms of MS. A predominance of hypoperfusion in the temporal lobes (like our patient) and frontal lobes with asymmetric involvement (more severe in the left hemisphere) have been reported [9]. Brain Perfusion SPECT can also differentiate between true cognitive disorders and an impairment of performance objectified by neuropsychological tests but caused by depression or fatigue [10]. Functional MRI studies have shown early cerebral compensation phenomena, which allow the use of additional cortical areas to correctly perform certain cognitive tasks, but can then be limited by the extent of the lesion and by the complexity of the tasks requested. To date, the results are contradictory concerning the potential effects of Disease Modifying Drugs (DMDs) on cognitive impairment. This therefore requires a cognitive remediation therapy [11]. The results may be unsatisfactory because of a late diagnosis and treatment of these forms with dementia onset and greater resistance to treatment.

Conclusion

MS revealed by dementia is rare and can be under diagnosed. The evolution of MS starting with cognitive impairment may be mediocre due to diagnostic and therapeutic delay and greater resistance to treatment.

References