NEURO-BEHÇET'S DISEASE PRESENTING AS AN ISOLATED PROGRESSIVE COGNITIVE AND BEHAVIORAL SYNDROME

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Objectives

Behçet's disease (BD) is a chronic disease manifesting as a vasculitis that affects arteries and veins of any size. Neurological dysfunction, defining neuro-Behçet's disease (NBD), can be present in up to 44% of patients. It is usually characterized by a progressive or relapsing multifocal neurological syndrome, similar to other acquired leukoencephalopathies, often associated with impairments in cognitive functions (memory, language, executive functions) and behavior. In this report, we describe an elusive case of NBD presenting exclusively with progressive cognitive and behavioral deterioration.

Subjects and Materials

Patient's clinical history subacutely started at age 42 with memory disturbances, non-fluent speech and tendency to disinhibition, with relentlessly progressive course, thus severely impairing social skills and daily life activities in the following months. During the entire disease course, the patient underwent extensive clinical, instrumental and laboratory evaluations, including neuropsychological examinations, brain MRI, cerebral FDG-PET, lumbar puncture and, finally, cerebral biopsy.

Results

The first MRI scan showed T2/FLAIR hyperintense lesions in right medial thalamus and right cortico-spinal tract, with enhancement after gadolinium administration. Follow-up MRI scans evidenced a progression of lesion burden, with involvement of bilateral supratentorial white matter including the genu of corpus callosum, as seen in Figure 1. FDG-PET revealed hypometabolism in right mesiofrontal and left temporal cortex, bilateral caudate nuclei, left putamen and both cerebellar hemispheres. CSF examination showed mild pleocytosis. All microbiological assays resulted negative. A new comprehensive neuropsychological examination was performed. (Table 1). The evaluation disclosed deficits of selective, divided and sustained attention skills and executive functions (cognitive flexibility, working memory and inhibitory control). Memory was significantly impaired. According to Frontal Behavioral Inventory questionnaire, our patient manifested anhedonia, inflexibility, indifference, disorganized and socially inappropriate behavior, thus displaying both apathetic and disinhibited conducts. Only partial and transient response was achieved after steroidal treatment. The cerebral biopsy revealed gliosis and perivascular inflammatory infiltrates. After a thorough interview, the patient revealed that he had been affected by erythema nodosum and painful, relapsing oral and genital ulcers up to three years before the onset of neurological symptoms. The presence of HLA-B51 allele and the positivity of the skin pathergy test corroborated the diagnosis of BD.

Discussion and Conclusions

Cognitive and behavioral features of NBD usually develop simultaneously to focal neurological signs and systemic manifestations of BD, reflecting disease activity. In our case the typical neurocognitive profile, associated with highly suggestive imaging findings, developed largely independently from the other BD features, thus hampering the correct diagnosis. Therefore, we recommend to include BD among the possible alternative diagnoses of acquired leukoencephalopathies, in order to promptly start an appropriate treatment.

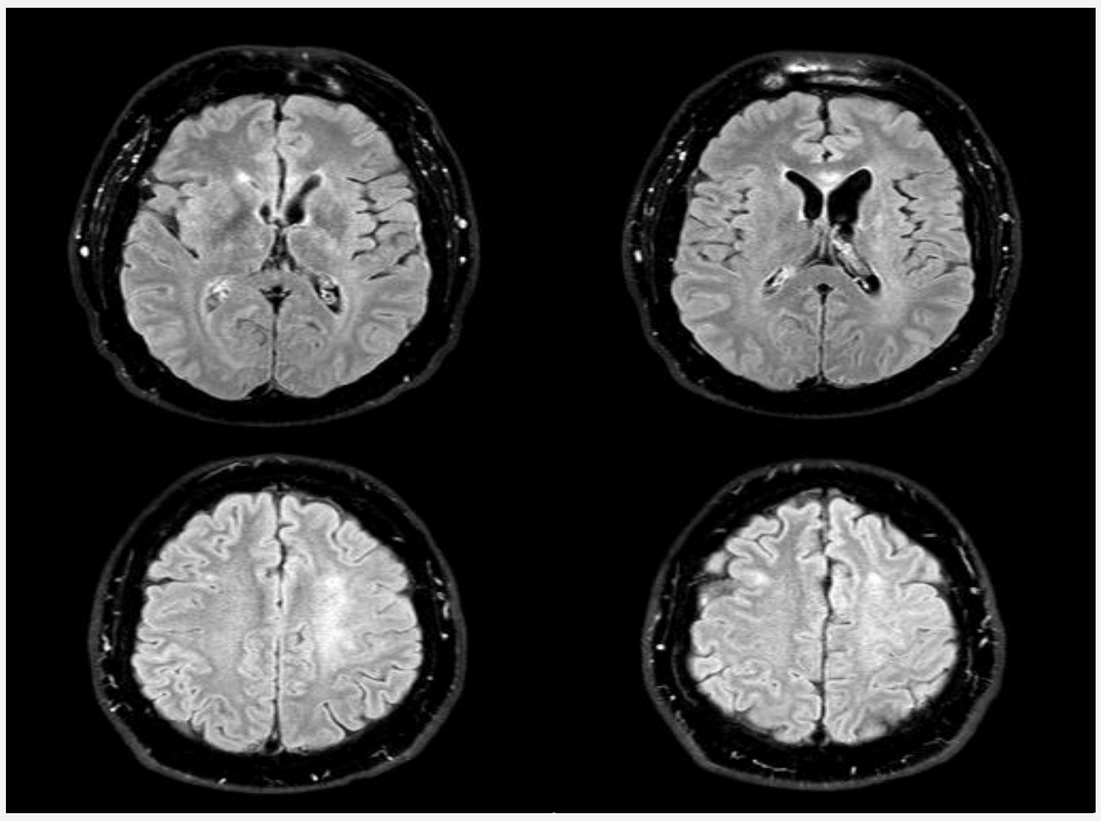

Figure 1. FLAIR-weighted MRI images show hyperintense lesions in the right thalamus and cortico-spinal tract, and in the corpus callosum.

Neuropsychological test	*Raw Score*	*Equivalent Score*
Short –Term Memory		
Visuo-spatial span [a]	6	4
Forward Verbal span [a]	9	4
Backward Verbal span [a]	3	1
Long-Term Memory		
Selective Reminding Test: Long term Storage [b]	22	N
Selective Reminding Test: Consistent Long Term Retrieval [b]	16	N
Selective Reminding Test: Delayed [b]	2	P
Story recall test: immediate recall [a]	5/8	N
Story recall test: Delayed recall [a]	0/8	P
Spatial Recall Test [b]	18	N
Spatial Recall Test- Delayed [b]	2	P
Attentional and Executive Functions		
Frontal Assessment Battery [a]	14	1
Phonological verbal fluency [a]	23	2
Stroop Test: Time [a]	39"	0
Stroop Test: interferend color naming [a]	3	1
Symbol Digit Modalities Test [b]	30	P
PASAT 3" [b]	26	N
PASAT 2" [b]	24	N
Language Abilities		
Word List Generation [b]	17	N
Non Verbal Reasoning Abilities		
Raven's Coloured Progressive Matrices [a]	22/36	N
Psychological Scale		
Frontal Behavioral Inventory-FBI	39	P

Table 1. General Neuropsychological and Psychological Assessment

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