Letter to the editor

Huntington's disease presenting as mixed state episode

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Dear Editor,

A 49-year-old woman presents with depressive symptoms characterized by depressed mood, diminished interest, aggressive behavior and recurrent suicide attempt. Symptoms had begun four years before, involving irritability, impulsive behavior, fatigue at work and persecutory delusions. Two months before, symptoms became more intense and she was diagnosed with bipolar disorder. She was referred to a psychiatric hospital due to the persistence of symptoms after irregular use of haloperidol, risperidone and valproate. There was no previous psychiatric history.

During hospitalization, mixed mood symptoms were observed: dysphoric mood, psychomotor agitation, suicidal ideation and delusions of persecution and ruin. Olanzapine and fluoxetine were started and the patient's mood improved. It was possible to distinguish the persistence of sluggish and impoverished thinking, spatial disorientation, increased response latency and apathy. Neurological examination revealed involuntary choreiform body movements imperceptible to the patient, which had started seven years before, and vocal tics that had appeared six months earlier. The patient had a family history of chorea. Clock drawing test exhibited visual spatial impairment; Mini-Mental State Exam revealed general cognitive decline (10 points; schooling: 8 years).

Laboratory (CBC, VDRL, HIV test, vitamin B12 levels, ionized calcium levels, blood glucose, thyroid, liver and renal function tests) was unremarkable. Computed tomography of the brain showed mild frontal lobe atrophy associated with dilation of the anterior horns of the lateral ventricles and decreased caudate nuclei heads (Figure 1). Genetic testing was performed due to suspected Huntington's disease (HD) and yielded 42 cytosine-adenine-guanine (CAG) repeats. After discharge, patient maintained mood stability and significant reduction of choreiform movements, with the prescription of fluoxetine 40 mg/day and olanzapine 10 mg/day.

HD is a rare inherited neurodegenerative disorder caused by the repeated expansion of a CAG trinucleotide, characterized by progressive motor, cognitive and psychiatric symptoms. Clinical manifestations and disease onset depend on the number of CAG repetitions. Symptoms present when there are more than 36 repetitions. Clinical manifestation is more common between 35 and 45 years, but can occur at any age. The course is chronic, slow and progressive, with an average survival of 10 to 20 years.[1,2]

Psychiatric manifestations are varied and can occur even in the prodromal phase, making diagnosis challenging. Neuropsychiatric symptoms occur in 35% to 73% of cases[2,3]. The most frequent manifestations are mood disorders, especially depressive symptoms, and an increased risk of suicide. Episodes of mania or hypomania may be present in 10% of cases. Studies show that up to 4.8% of HD patients develop bipolar disorder[3-5].

Here, we present the case of a patient diagnosed with HD, presenting as mixed state episode in the context of behavioral manifestations of a neurodegenerative disease. The patient met DSM 5 criteria for manic episode with mixed symptoms, due to dysphoric mood, psychomotor agitation, aggressive behavior, lack of interest and pleasure, delusions of ruin and recurrent thoughts of death. We could not find any reports of mixed state episodes in HD. This report may help the clinician to consider differential diagnoses regarding the onset of psychiatric symptoms associated with neurological manifestations.

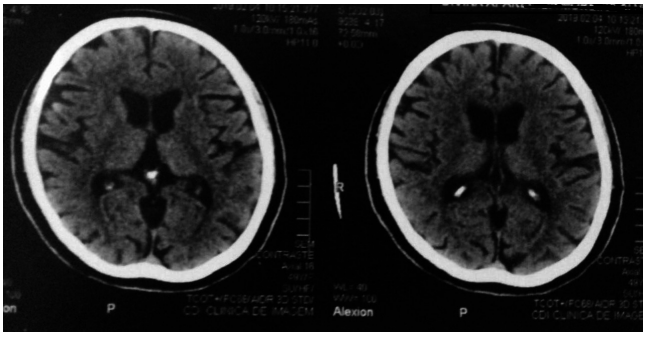

Figure 1. Computed Tomography of the brain showing mild frontal lobe atrophy associated with dilation of the anterior horns of the lateral ventricles and decreased caudate nuclei heads.

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