Paroxysmal dystonia as first symptom of multiple sclerosis: presenting two cases

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Introduction

Paroxysmal dystonia is a well-known but very uncommon manifestation of multiple sclerosis. It occur when opposing muscle groups contract simultaneously causing twisting or turning around one or multiple joints. The pathophysiological mechanisms are not well established, but one speculation is that of the transversely spreading activation of axons within the demyelinating lesions of the thalamus, midbrain, medulla or spinal cord. We present two cases of patient with multiple sclerosis started with paroxysmal dystonia responsive to steroid treatment in the first case and to Carbamazepine in the second case.

Case report

The first case is a 36 year old woman diagnosed with Neurofibromatosis type 1 in childhood. No family history of movement disorders. At the age of 27, as part of investigations related to the underlying disease, she performed a brain MRI, which showed a focal demyelination but no other investigation were performed at that time. At the age of 34 she developed paresthesias to the right emisoma and right-hand dystonia. The brain and cervical spine MRI showed demyelinating areas both in the brain and in the spinal cord without gadolinium enhancement (Fig.1). Two oligoclonal bands were found in the CSF. She was diagnosed according to the McDonald's criteria with relapsing-remitting MS. The patient recovered with intravenous steroid administration and at the age of 37 she started treatment with glatiramer acetate. Since then she had no relapses or new lesions at MRI scan.

The second case is a 22 year old woman with optic neuritis at the onset in right eye (visual loss and pain); in the following weeks in the outpatient diagnostic path and during the steroid treatment she developed paroxysmal right arm and limb dystonia. At brain MRI scan there were multiple T2 lesion in the white matter of the temporal lobe bilateral, posterior limb of the left internal capsule, in the corpus callosum, in the left medium cerebellar peduncoli; another iperintensity in the right optic nerve with gadolinium enhancement. At cervical MRI scan there was a C3 lesion without gadolinium enhancement (Fig. 2). In the CSF there were five oligoclonal bands. Complete basic and immunological analysis and serological studies including HIV, Borrelia, Brucella, Herpes virus, Epstein-Barr virus and Treponema pallidum were all normal or negative. Visual evoked potentials showed increase in latency in right eye. AQP4 antibodies were negative. CSF viral sample were also negative. She underwent eeg: during the registration she had an episodic right hand dystonia without concomitant epileptic pattern. With the steroid treatment recovered the visual loss, but there was no benefit on paroxysmal dystonia. So Carbamazepine 200 mg twice daily was started with complete control of the symptom. She started also Dimethyl fumarate with no relapse until now.

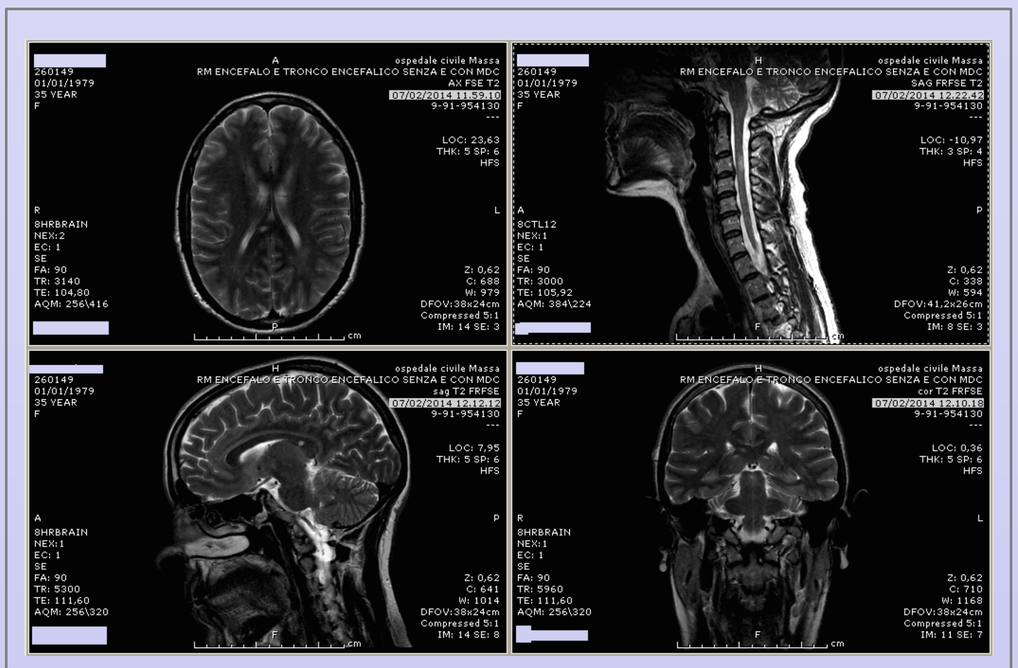

Fig.1

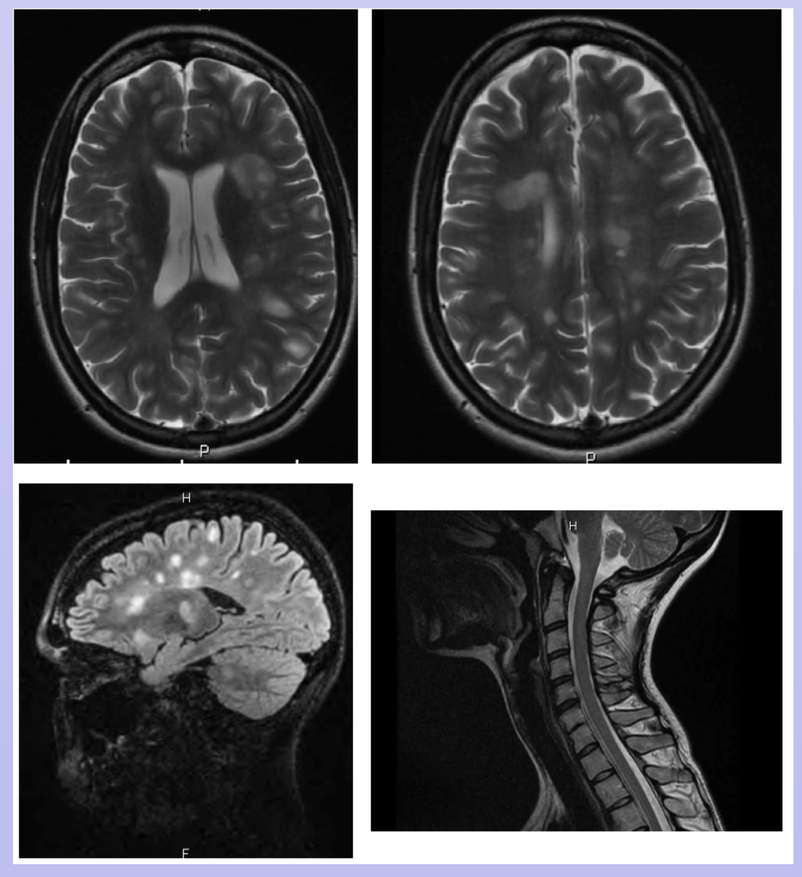

Fig.2

Conclusion

An accurate diagnosis of multiple sclerosis requires the recognition of its atypical clinical manifestations. Paroxysmal dystonia is the most frequently reported type of dystonia in multiple sclerosis, with brief attacks (seconds to a few minutes) that occur several times a day. The proposed pathophysiologic explanation is ephaptic activation of axons secondary to a demyelinating lesion located in the midbrain, posterior limb of the internal capsule, cerebral peduncle, thalamus, subthalamus, and cervical spinal cord, even cases of MS not explainable by anatomical evidence have been reported. There were some typical trigger factors, such as startle reaction, hyperventilation, stress, and fatigue, fact that support the hypothesis of transverse non-synaptic transmission. The symptom may recede without treatment. There are no randomized clinical trials of any treatment of dystonia in multiple sclerosis patients so the strategy should be studied in each case individually. The treatment options include a course of high-dose steroid therapy, carbamazepine, acetazolamide, and valproate.

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