LEVODOPA RESPONSIVE DYSTONIA SECONDARY TO VIRUS ENCEPHALITIS

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Movement disorders secondary to virus encephalitis is an uncommom feature and is mainly observed after the acute or subacute stage of Japanese encephalitis. This condition, described in Asian countries, is characterized by pleomorphic neurologic signs and poor outcome as 30% of patients die and around half of the survivors have severe neurological sequelae[1]. In Brazil, although some extra-piramidal syndromes such as chorea continues to be an endemic post-infectious disease, dystonia has seldom been related to central nervous system infection[2].

In previous paper, our group showed that the neurological sequelae of post infectious meningoencephalitis are extremely pleomorphic, however no case of dystonia was described[3].

CASE

The patient, a 13-year-old girl, was born to a healthy Brazilian family. At the age of 7, she started to have persistent headache, vomiting and high fever followed by epileptic crisis and decreasing consciousness level. CSF examination showed lymphomononuclear pleocytosis (85 cels/mm^3), mild increase protein (60 mg%) and normal glucosis. She had the diagnosis of viral encephalitis and received acyclovir during ten days. After the beginning of treatment she continued in coma and left conjugated eye deviation for more five days with progressive improvement. In the day of hospital discharge, she had right spastic hemiparesis and motor aphasia. After neurological rehabilitation, she persisted with the motor deficit without any language disturbance. At the age of 11, she changed schools and had to walk for long distances. After approximately one month, she began to present pain in the right muscle triceps and involuntary flexion of foot toes at the same side, which disappeared after rest. At the age of 12, the episodes became more frequently, occurring after 200 meters of walking.

In April 2004, she was seen by the movement disorders group of the Division of Neurology and Epidemiology – Federal University of Bahia and her neurological examination revealed mild spastic hemiparesis and right gastrocnemius atrophy. After walking 100 meters in accelerated steps, she had intense pain associated with dystonia and flexion of right toes that persisted for five minutes after interruption of walking. The T2-weighted MRI showed marked, bilateral gliosis of substance nigra with no other encephalic involvement and motor symptoms disappeared after oral administration of 200 mg plain levodopa with carbidopa. So far, the effects have been sustained without any side effects.

DISCUSSION

We report the first case of levodopa responsive dystonia occurring several years after viral encephalitis. Although a number of movement disorders has been associated with virological diseases such as Coxsackie B, measles, polio, herpesvirus, encephalitis lethargica, western equine encephalitis and Japanese encephalitis[4-8], dystonia is an uncommon sequelae of post-viral encephalitis usually, it has been associated with other parkinsonian phenomena[9]. In cases of Japanese encephalitis, the most common of the above diseases, extrapiramidal symptoms were described in 70% of patients[10], but the majority of them had parkinsonian features as the main symptom and even patients with dystonia rigidity, hypokinesia, masked face and tremor were frequent. However, contrary to the cases of Misra and Kalita, which described movement disorders in the acute and subacute stage of JE, in our patient dystonia occurred several years after encephalitis and only under exercise stress resembling that it only appeared after wasting the reserve of nigro-striatal levodopa. However, as it has been sufficiently described, degeneration of dopamine neurons in the substancia nigra is responsible for idiopathic Parkinson's disease, juvenile parkinsonism, hereditary progressive dystonia, and levodopa responsive dystonia, yet the clinical phenotypes in the three disease states are different, which means that they also have different physiopathological process[11]. Regarding our specific case, some particular features, such as the beginning of symptoms after several years of encephalitis, points to a progressive decline of dopamine, which is pronounced after exercise. As it has been stated elsewhere, processes of

DISTONIA SENSÍVEL A LEVEDOPA SECUNDÁRIA A ENCEFALITE VIRAL

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nigrostriatal dysfunction vary, and it is very possible that the clinical manifestations based on the striatal dysfunction maintain continuity and that variable clinical phenotype may develop after several years[12].

In conclusion, our case illustrates the occurrence of post-encephalitic dystonia in an adolescent with bilateral gliosis of substancia nigra, and draws attention to the excellent improvement after levodopa therapy.

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