Focal Limb Dystonia in a Patient With a Cerebellar Mass

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Background: Focal dystonia of acute onset is indicative of a structural lesion in the nervous system. Cerebellar lesions have rarely been associated with dystonia.

Case Description: A 42-year-old woman was admitted to the neurology ward because of fever, confusion, and gait unsteadiness. She was diagnosed as having tuberculous meningitis, and, after a few days of antituberculous treatment, she developed prominent dystonia of the left upper limb. Cranial nuclear magnetic resonance imaging showed an isolated lesion compatible with a tuberculoma in the left cerebellar hemisphere. Both the limb dystonia and the tuberculoma resolved with maintained antituberculous treatment.

Conclusions: In the patient described, the presence of upper-limb dystonia ipsilateral to a focal cerebellar lesion and the resolution of the dystonia and the mass lesion following treatment suggest that the cerebellum or its connections to the thalamus and/or basal ganglia could be involved in the pathophysiology of the dystonia.

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Dystonia is characterized by inappropriately prolonged muscle contractions that forcefully distort the body into abnormal postures. Most cases of dystonia are idiopathic. Acute onset of dystonia, particularly when it begins focally in a limb and remains unilateral, suggests the possibility of a focal central nervous system structural disease. The central nervous system sites that have been most frequently implicated as causing secondary dystonias are the thalamus and the striatopallidal complex. Other sites include, more rarely, the spinal cord, the brainstem, and the cerebral cortex. Lesions that primarily involve the cerebellum or its pathways have not been classically associated with dystonia. We report a case of a patient with left upper-limb dystonia of acute onset associated with a cerebellar tuberculoma.

REPORT OF A CASE

A 42-year-old woman with a history of pulmonary tuberculosis in her husband and 4 children was admitted to the hospital because of signs of apathy, irritability, anorexia, and restlessness at night. Four weeks before admission, she developed headaches, nausea, vomiting, abdominal pain, urinary incontinence, and generalized tonic-clonic seizures. Two weeks later, she displayed daytime sleepiness, fever, and gait instability. On admission, she had a fever and was confused, with a tendency toward sleepiness. Her speech was slow. Funduscopic examination revealed absence of venous pulsations, but the papilla boundaries were well delineated. Pupils were normal and reactive to light. Nystagmus at the right lateral gaze was observed. She had mild right facial and limb paresis. Tendon reflexes were normal and symmetrical. The right plantar response was equivocal and the left was flexor. She had a left cerebellar syndrome with upper-limb intention tremor and with finger-to-nose and heel-knee-shin ataxia. Her gait was unsteady. Findings of sensory examination, including proprioception, were normal.

Results of routine laboratory tests, chest x-rays, and electrocardiogram were normal. A cranial computed tomographic scan showed hydrocephalus. Cerebrospinal fluid examination showed 170 cells/mL, with 95% of them mononuclear; proteins, 300 mg/dL; and glucose; 55 mg/dL (3.0 mmol/L). The result of a Mantoux test was positive. A diagnosis of tuberculous meningitis was made, and antituberculous treatment was started with ethambu-
tol, isoniazid, rifampicin, and pyrazinamide. The result of a Lowenstein-Jensen culture and an enzyme-linked immunosorbent assay for tuberculosis confirmed the diagnosis.

After 2 weeks of antituberculous treatment, the patient had improved but still had mild intermittent confusion. She then developed involuntary movements and postures in the left arm. At rest, she kept a dystonic posture in her left arm with wrist flexion and slight internal rotation in the arm and hand, which tended to become more intense with action. When she was asked to keep her arms outstretched, marked dystonic posture of the left arm developed, with marked internal rotation of the arm and hand and downward wrist flexion accompanied by adduction of the thumb and hyperextension of the fingers. When standing up, she also showed an abnormal limb posture, which was more evident when walking, showing internal rotation and bringing backward the upper left limb with flexion of the wrist and metacarpophalangeal joints and adduction of the thumb. The patient also displayed a postural and intentional tremor of the upper limbs that was more pronounced on the left side. There was a mild right hemiparesis with ataxia. No sensory deficits could be demonstrated. Cranial nuclear magnetic resonance imaging (NMRI) showed hydrocephalus and a 4-cm left cerebellar lesion, predominantly vermian and paravermian, that was hyperintense on T2-weighted sequences and enhanced with gadolinium (Figure). This lesion was compatible with tuberculoma.

Ten days later, dystonia of the upper limb had improved, but gait ataxia persisted for 2 months. After 90 days of treatment, the patient was free of symptoms. A new NMRI of the brain was normal.

**COMMENT**

We report a case of a patient with tuberculous meningitis who, 2 weeks after starting antituberculous therapy, developed prominent upper-limb dystonia of acute onset. Cranial NMRI showed a hyperintense lesion on T2-weighted sequences in the cerebellar hemisphere ipsilateral to the dystonic arm that was enhanced with gadolinium, but no other focal structural lesions were seen. The cerebellar lesion was compatible with a tuberculomas. Both limb dystonia and the cerebellar lesion disappeared with antituberculous treatment. Other neurological symptoms and signs displayed by our patient, such as confusion or hemiparesis, could be related to hydrocephalus or to extracerebellar pathologic conditions not visualized on the cranial NMRI.

Cerebellar disorders are typically not associated with dystonic phenomena, and the cerebellum or its pathways have not been usually considered in the pathophysiology of dystonia. In certain neurodegenerative disorders of the cerebellum, brainstem, and spinal cord, cerebellar ataxia and dystonia can coexist, and focal dystonia of the hand has been described in cases of isolated degenerative ataxia. In these cases of focal dystonia and isolated ataxia, the cerebellar syndrome was more prominent on the side of the dystonic limb, but the authors consider this type of dystonia to be the result of presumed abnormalities of the basal ganglia or their connections. Still, other evidence suggests that the cerebellum or its projections could be involved in dystonic disorders. Human deep cerebellar stimulation can result in abnormal tonic postures of the neck and limbs. Several cases of blepharospasm associated with brainstem lesions had clinical or radiological evidence of cerebel-
lar involvement. A recent case report suggested that symptomatic limb dystonia could be produced by focal brainstem lesions and that cerebellar dysfunction could play a role in its pathophysiology. Another study, conducted with positron emission tomography, emphasized the participation of structures such as the cerebellothalamic pathways in the genesis of dystonic phenomena. In experimental animals, brainstem lesions involving the medial longitudinal fasciculus, medial reticular formation, red nucleus, and brachium conjunctivus have produced dystonia. Finally, it has been suggested that the damage of a single subpopulation of cerebellar neurons could also produce dystonia.

In our patient, the coexistence of a unilateral cerebellar syndrome in the same arm with the focal dystonia and the presence on NMRI of a cerebellar lesion ipsilateral to the dystonic syndrome suggest a cerebellar origin for the abnormal involuntary movement. The concomitant disappearance of the focal dystonia and of the cerebellar mass following treatment also supports this interpretation. Deep small or asymmetrical lesions in or around the basal ganglia secondary to the presence of basilar exudate that were not apparent in the imaging studies could have conditioned the abnormal movements in our patient. However, recent studies in patients with tuberculous meningitis show that abnormal movements are more frequently associated with the presence of demonstrable deep infarcts on imaging studies. The absence of these lesions and the presence of a single cerebellar mass suggest that the dystonia in our patient could be cerebellar in origin.

A cerebellar lesion involving the dentate nucleus or its efferent pathways could cause abnormal dystonic movements and postures ipsilateral to the cerebellar lesion through an abnormal functional input to the contralateral thalamus. In our patient, the left cerebellar lesion, through edema, direct pressure effect, or ischemia, could have compromised the cerebellar input to the contralateral thalamus, leading to contralateral thalamofrontal disinhibition and abnormal central sensorimotor processing, which could have resulted in dystonic movements ipsilateral to the cerebellar lesion.

Secondary focal dystonias have been encountered most often with lesions of the contralateral thalamus and of the lentiform and caudate nuclei, and only rarely in patients with brainstem lesions. The association of arm dystonia in a patient with an ipsilateral cerebellar tuberculoma, described in this report, and the available evidence from the literature support the concept that a cerebellar lesion may cause or significantly contribute to the development of a focal dystonia in some patients.

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REFERENCES