

Upper-Limb Dystonia Secondary to a Midbrain Hemorrhage



To the Editor:

Focal dystonia limited to the hand or arm has been described in patients with basal ganglia lesions (1,2). Brain stem lesions have been associated with cervicocranial dystonia in a number of cases (3,4) and, on exceptional occasions, with contralateral hemidystonia (5,6). Unilateral asterixis also has been observed in focal brain lesions, but bilateral asterixis is generally a manifestation of diffuse encephalopathy (7). We report a patient with left upper limb dystonia of acute onset and bilateral asterixis related with a left brain stem hemorrhage.

Case Report

A 46-year-old woman, previously in good health, was admitted to our hospital because she noted sudden dizziness and weakness of the right limbs. When she arrived at the hospital, she was conscious, dysarthric, had mild hypophonia and referred vertical and horizontal diplopia. Her pupils were 2–3 mm in diameter, symmetric, and poorly reactive to light. Spontaneous gaze resulted in a convergent strabismus with an alternant ocular skew deviation. Saccadic and pursuit upward eye movements were absent, and voluntary downward movements were markedly reduced but improved with oculocephalic maneuvers. She had a bilateral pseudo-sixth nerve palsy with convergence spasms. Optokinetic nystagmus was normal. There was a discrete right central facial and right upper limb paresis. Tendon reflexes were brisk and symmetric, but Babinski sign was not present. Touch, pain, and thermal senses were diminished on the right side of the face, the right upper limb, and the right upper area of the trunk. No sensory abnormalities, including proprioceptive and vibration senses, were present on the left limbs. She had a left cerebellar syndrome with finger-nose and heel-knee-shin ataxia, upper limb intention tremor, and gait instability.

A left upper limb abnormal posture was observed at rest, which was more evident when the patient held the arms outstretched. In this position, there was ulnar deviation of the hand, hyperextension of the fingers with abduction of the fourth and fifth fingers, and supination of the arm. This dystonic posture was strikingly aggravated by some voluntary movements. The hand then showed an unwanted posture with abduction and flexion of the metacarpophalangeal joints of the fourth and fifth fingers, accompanied by adduction of the thumb and flexion of the interphalangeal joints of the second and third fingers. Moreover, there was ulnar deviation of the hand and, at times, hyperflexion of the wrist. Bilateral asterixis, more prominent on the right arm, but also present in the proximal and distal muscles, was observed during the first few days.

Routine laboratory tests, including toxicologic screening, chest radiogram, and electrocardiogram were normal. Cranial computed tomography (CT) showed a hem-

orrhagic lesion in the left midbrain and cranial magnetic resonance imaging (MRI) evidenced that this lesion extended from the left pretectal region of the midbrain to the left superior cerebellar peduncle, with ipsilateral red nuclei displacement (Fig. 1). Arteriographic study failed to demonstrate a vascular malformation. Median nerve evoked potentials recorded on the scalp were normal bilaterally. Electromyographic (EMG) recordings showed a characteristic dystonic pattern with cocontraction of agonists and antagonist muscles in the left forearm and sometimes in the left arm, and occasionally irregular periods of electrical silence occurring simultaneously in the flexors and extensors muscles of both forearms.

The patient's clinical evolution was relatively satisfactory. Asterixis diminished dramatically within a few days. A week later, she was able to walk without aid and the cerebellar syndrome improved. Five months later the left dystonia had also improved. Ocular abnormalities were unchanged. MRI showed only a residual deposit of hemosiderin in the midbrain (Fig. 2).

Discussion

Our patient developed upper-limb dystonia of acute onset in association with an ipsilateral brain stem lesion. We have not found any report in the literature indicating such an association. No motor or sensory abnormalities were present in the dystonic limb, and MRI showed a midbrain hemorrhage but no other structural central nervous system lesions. In patients with secondary isolated focal dystonia of the hand and arm, Marsden et al. (2) found that the responsible lesion causing dystonia mainly affected the contralateral thalamus, but caudate or lentiform nucleus were involved in some cases. Reports of secondary limb dystonia occurring in patients with brain stem lesions are exceptional. Leenders et al. (5) published a case of ipsilateral blepharospasm, contralateral hemidystonia, and parkinsonism in a patient with a mesencephalothalamic lesion, and Krauss et al. (6) reported a patient with contralateral hemidystonia and torticollis secondary to a traumatic pontomesencephalic lesion.

Dystonia in patients with lesions in the basal ganglia may be caused by an abnormal input from the thalamus to premotor cortex, including the supplementary motor area, from a lesion either in the thalamus itself or in the striatum (2). An explanation for the pathophysiology of the limb dystonia is difficult in our case, in which no lesions were identified in the basal ganglia and no sensory abnormalities were present in the dystonic limb to explain dystonia on the basis of deafferentation. The left cerebellar syndrome ipsilateral to the dystonia suggests involvement of cerebellar pathways that could have contributed to the development of dystonia in our patient. In previously reported cases of hemidystonia with brain stem lesions, patients had lesions situated at a level that was higher or lower than the lesion in our case; however, the dystonia was contralateral to the lesion. In our case, disruption by the hematoma of the left dentato-rubro-thalamic pathway before the decussation of the brachium conjunctivum at the level of the inferior colliculi could explain why the cerebellar syndrome and dystonia were ipsilateral to the lesion. Classically, lesions involving this

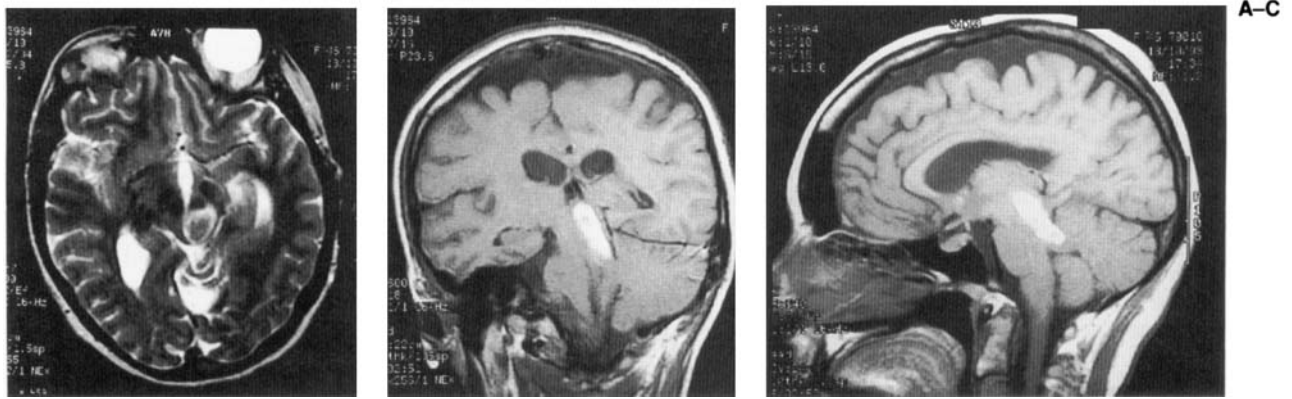


FIG. 1. A: T2-weighted MRI showing a recent hemorrhagic lesion in the left midbrain with mass effect over the ipsilateral red nucleus. B and C: T1-weighted MRIs showing the hematoma in the coronal and sagittal planes, respectively.

fascicle or cerebellum are not associated with dystonia. However, as Hedreen et al. (8) suggested, it is possible that a lesion involving a single population of cerebellar neurons could produce dystonia. Disconnection between the cerebellum and thalamus offers a reasonable explanation for the pathophysiology of the dystonia in our case, but other possibilities may be considered. It has been proposed that limb dystonia in the brain stem (6) or spinal cord lesions (9) may be produced by lesions involving the descending inhibitory supraspinal pathways from reticulospinal tracts. Such a mechanism also could have played a role in our case. Although reticulospinal tracts originate in the central pontine and dorsal and central parts of the medulla (medial reticulospinal tract) and ventrolateral pontine tegmentum (lateral reticulospinal tract) (10), always ventral and caudal to the hemorrhagic area in our patient, we cannot rule out that these nuclei were not receiving an abnormal input from other structures such as the basal ganglia, thalamus, or cerebellum due to the hemorrhage.

In experimental animals, brain stem lesions involving

red nucleus, brachium conjunctivum, portions of the medial reticular formation, and medial longitudinal fasciculus can produce spasmodic torticollis and other facial dyskinesias (4,11). Other investigators (12) have found that glutamic acid decarboxylase activity is elevated in the deep cerebellar nuclei of the rat mutant dystonic, supporting the view that cerebellar dysfunction may be a primary component of the dystonia in these animals. There is some evidence suggesting that the cerebellum or its projections could be involved in human dystonic disorders. Electrical stimulation in the region of the interpositus and dentate nuclei of the cerebellum resulted in abnormal tonic postures, whereas lesions made in the medial dentate region improved athetosis, tremor, or spasticity in patients (13). Several cases of blepharospasm associated with brain stem lesions had clinical or radiological evidence of involvement of the cerebellum or its pathways (3,14); however, these structures have not been considered in the pathophysiology of blepharospasm. Pathological studies in patients presenting with craniocervical dystonia have been contradictory. Some investiga-

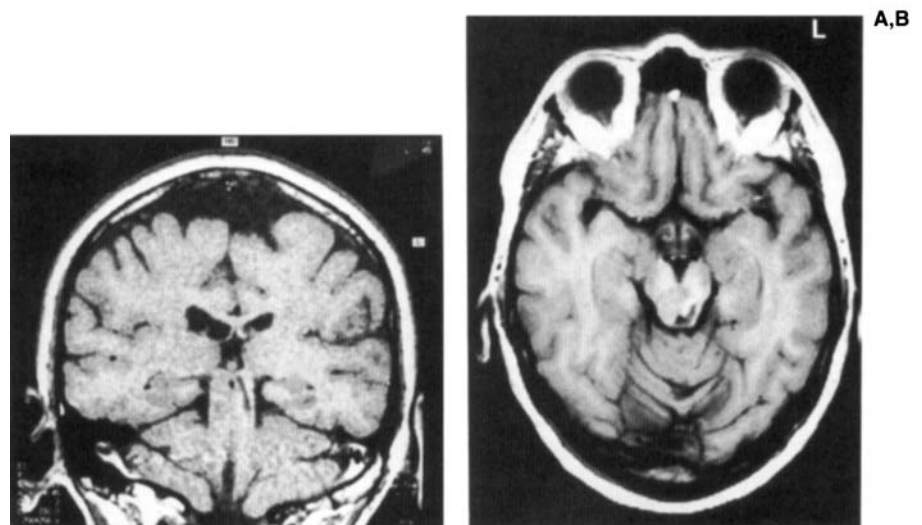


FIG. 2. T1-weighted MRIs in the coronal (A) and axial (B) planes 5 months after stroke showing reabsorption of the hematoma with a residual area of hemosiderin.

tors (15,16) found cell loss in the dentate and other nuclei of the brain stem, but similar findings have not been observed in other cases (17). Finally, the association of cerebellar ataxia and dystonia has been described in neurodegenerative disorders such as dentato-rubro-pallidoluysian atrophy (18), Machado-Joseph disease (19), and olivopontocerebellar atrophy (20), but in these cases the basal ganglia could be involved and dystonia has been considered a minor feature. However, Fletcher et al. (21) reported eight cases of isolated cerebellar ataxia and focal dystonia; three patients had torticollis, and six patients had writer's cramp, but dystonia was also present in the outstretched hand in some of these. The cerebellar syndrome was generally more prominent on the side of the dystonic limb. Although the investigators suggest that basal ganglia involvement is the responsible mechanism of the dystonic disorder, these cases demonstrate focal dystonia associated to degeneration of the cerebellum, brain stem, and spinal cord.

Our patient also had bilateral asterixis. Unilateral asterixis has been reported in one patient with a mesencephalic syndrome, suggesting disturbance in and around the midbrain periaqueductal and pretectal regions (22). Although bilateral asterixis has been associated with metabolic encephalopathy, we have found one case in the literature that describes bilateral asterixis secondary to a structural midbrain lesion (23). The investigators in this study suggested that damage to the mesencephalic reticular formation was the responsible mechanism of the asterixis. Such a mechanism could explain the asterixis in our case because bilateral involvement of the periaqueductal reticular formation due to the hemorrhage could be possible in our patient. Others have described a case of ipsilateral asterixis due to a cerebellar hemorrhage (24) and have concluded that asterixis in their patient may have been due to a reduction of afferent information through the dentato-thalamo-cortical input. Bilateral involvement of the dentato-thalamic pathways is another possible explanation of the bilateral asterixis in our patient because MRI shows that the hemorrhage could be involving either the left dentato-rubro-thalamic fascicle before decussation and perhaps also, partially, the right dentato-rubro-thalamic fascicle above the red nucleus.

We conclude that symptomatic limb dystonia can be produced by a focal brain stem lesion and that cerebellar dysfunction could play a role in its pathophysiology. Furthermore, we believe that the presence of a brain stem lesion may explain bilateral asterixis in the absence of diffuse encephalopathy.

Legends to Videotape

Segment 1. This segment demonstrates the patient's left upper limb dysmetria and intention tremor and left leg heel-knee-shin ataxia.

Segment 2. This segment demonstrates the left arm and hand dystonia (a) when the patient holds her arms outstretched and (b) when she tries to execute some movements with the limb.

Segment 3. Five months after stroke, the patient's dystonia is still present but has improved.

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Walking-Induced Parkinsonism Due to Presumed Idiopathic Normal Pressure Hydrocephalus



To the Editor:

Normal pressure hydrocephalus (NPH) is a clinical syndrome without a known pathological substrate other than hydrocephalus (1). It exists in two forms: idiopathic and secondary (1). The secondary form occurs after inflammatory substances enter the cerebrospinal fluid (CSF), presumably causing obstruction of CSF flow over the convexities of the brain. Thus secondary, or "symptomatic," NPH may occur after subarachnoid hemorrhage, meningitis, or head trauma.

The clinical syndrome of NPH is usually thought of as a triad of dementia, gait failure, and urinary incontinence (2), but it is clear from clinical reports that not all elements of the triad need be present (3). Because there is no pathological abnormality to define the condition, the diagnosis rests on a response to shunting. Unfortunately, the response to ventricular shunting for the idiopathic form of NPH is highly variable, but overall is disappointing (4). Diagnostic tests such as cisternography, neuropsychological testing, withdrawal of large volumes of CSF, and magnetic resonance imaging (MRI) are unreliable due to poor or unknown sensitivity, poor specificity, or both (4,5). In the idiopathic form, the occurrence of gait dysfunction preceding dementia seems to improve the prognosis for a shunt response, and the presence of dementia appears to reduce the benefits of shunting (3,4,6-8), suggesting that idiopathic NPH (INPH), if defined by shunt responsiveness, is primarily a gait disorder. Presumably most demented patients with ventriculomegaly have diagnoses such as Alzheimer's disease or multi-infarct dementia with hydrocephalus ex vacuo, concomitant, unrelated asymptomatic hydrocephalus, or have had the condition so long that plasticity has been lost.

The gait abnormalities of INPH have been described in widely discrepant terms. Sudarsky and Simon (9), who

carefully studied six patients with INPH defined by positive cisternography and beneficial response to CSF removal, basically described the gait as parkinsonism from the waist down. That is, there was shuffling, with reduced stride, increased simultaneous contractions of flexors and extensors of the legs, a flat footed strike, increased time with both feet touching the floor, and a flexed posture but intact arm swing. Adams et al. (10), in the first published report of NPH, described the gait as "difficult to categorize" but unsteady in a way that was not clearly cerebellar in origin. Fisher (2) stated that the majority of the cases were ataxic, with "poor balance, off balance, unsteady, wobbling, drunken." Some patients had spells of weakness. Estanol (11) reported that the gait was apractic because the patients could perform complicated maneuvers with their feet while seated or supported but could not move their feet to walk.

This confusion in describing gait either reflects a wide heterogeneity of presentations of INPH, a large collection of different disorders associated with ventriculomegaly, or a large collection of different disorders in patients who independently had large ventricles unrelated to their neurodegenerative disorder.

The following case describes what I believe is a previously unreported phenomenon, walking-induced parkinsonism, which occurred in an individual with ventriculomegaly and was cured by shunting.

Case Report

The patient was a 68-year-old man referred for evaluation of possible Parkinson's disease. For the 3 years preceding referral he had a slowly progressive gait disorder that he attributed to a feeling of fatigue and weakness in his legs that increased with walking. After resting he stated that he walked normally but only for a few steps, after which the problem began and increased with effort. He also noted a feeling of wanting to run forward when he became tired. He denied any other symptoms of parkinsonism such as slowness, decreased dexterity, tremor, or changes in penmanship, voice, or balance. There were also symptoms of urinary urgency, without incontinence, and nocturia, which had never been evaluated. There was no family history of any neurologic disorder, and the patient had no history of neurologic or psychiatric dysfunction, toxin exposure, or use of dopamine-blocking drugs. He had a history of gout, for which he took colchicine and allopurinol, and angina, for which he took nifedipine.

Evaluations by previous physicians had included a spinal MRI scan showing spondylotic changes in the lumbar spine and two electromyograms; the results of one were normal and the other showed mild polyradiculopathies in both legs. He scored 29 of 30 on a mini-mental status examination 1 year before my evaluation. The neurologic examination was normal except for gait, diminished amplitude on a rapid heel-tapping maneuver bilaterally, slightly decreased rapid alternating movements of the left hand, and absent right ankle jerk. The patient initially walked normally but after a brief period exhibited diminished arm swing on the right and then began to drag his right leg. With prolonged walking, he developed mild flexion of the knees and decreased stride bilaterally. The