

Delayed-onset Focal Dystonia After Stroke

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The delayed-onset focal or segmental dystonia is a rare sequelae of cerebrovascular disease. The responsible lesion sites for the dystonia are variable and the pathogenesis is uncertain.

This study reports three patients with delayed-onset focal dystonia as a complication of stroke. The interval between hypoxic insult and onset of dystonia were varied from 1 month to 1 year. Two adults and one child had focal lesions at the contralateral basal ganglia. The interval between the brain damage and dystonia did not appear to be related to the age at the time of hypoxic injury. The site of lesions may serve as an important factor in the pathogenesis.

Key Words: Delayed-onset dystonia, stroke

Dystonia is characterized by sustained muscle contraction, which leads to abnormal posture, twisting and repetitive movements (Fahn, 1988). Since the first description by Hammond in 1871, delayed-onset dystonia is a rare but well known sequelae of cerebrovascular disease (Dooling and Adams, 1975; Traub and Ridley, 1982; Demierre and Rondot, 1983; Russo, 1983; Obeso *et al.* 1984; Chiang and Lu, 1990). It may also develop as a sequelae of many other disorder such as; perinatal anoxia (Burke *et al.* 1980; Marsden *et al.* 1985; Pettigrew and Jancovic, 1985; Saint-Hilaire *et al.* 1991), truama (Andrew *et al.* 1982; Brett and Hoart, 1981; Krauss *et al.* 1992), tumor (Marsden *et al.* 1985; Glatt and Nausieda, 1984; Narbona *et al.* 1984), infection (Dooling and Adams, 1975), vasculitis and SLE (Anegawa *et al.* 1986; Daras *et al.* 1988).

It can occur immediately after brain damage, but more frequently it develops after a variable period (Russo, 1983; Marsden *et al.* 1985; Pettigrew and Jancovic, 1985). Caudate

nucleus, lentiform nucleus, thalamus, internal capsule, or variable combinations of them have been reported as the responsible site for the delayed-onset dystonia (Marsden *et al.* 1985; Pettigrew and Jancovic, 1985).

The pathologic process responsible for the dystonia during the initial recovery period is unknown. Some authors have compared the interval of delayed-onset dystonia in patients of perinatal anoxia and stroke. They emphasized the age of hypoxic injury in deciding the duration of delay in spite of the differences of etiology (Pettigrew and Jancovic, 1985).

We present three patients with delayed-onset focal dystonia after stroke and reviewed the related literatures.

CASE REPORTS

Patient 1

A 57-year-old male patient was referred due to painful paresthesia and dystonic posture of the left hand. Five years ago, he suffered from a sudden onset of left side hemiplegia and sensory loss. Brain CT scan showed an hemorrhage involving the right putamen, thalamus, internal capsule and head of the caudate nucleus (Fig. 1). The left

Received July 10, 1993

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hemiplegia improved enough so that he could walk without assistance several months later. Thereafter his left hand became clumsy. One year since the cerebral hemorrhage, dystonic posture in the left hand consisted of flexion of the metacarpal joints and extension of the interphalangeal joints began to appear. Repetitive myorhythmic dystonic movement of the fingers were noticed. It consisted of slow

spontaneous involuntary twisting, flexion of the metacarpophalangeal joints, fanning of the fingers, and extension of the thumb (Fig. 2) which was accentuated by stress. On examination, mild dysarthria, left facial sensory loss and decreased gag reflex were noted. There was a mild hemiparesis with hyperactive deep tendon reflexes at the left side. There were definite deep and superficial sensory deficits in the left side. A routine hematological test, serum electrolyte, liver function test, and renal function test were normal.



Fig. 1. A brain CT of patient 1 showed hemorrhage involving the right putamen, thalamus, internal capsule and head of the caudate nucleus.

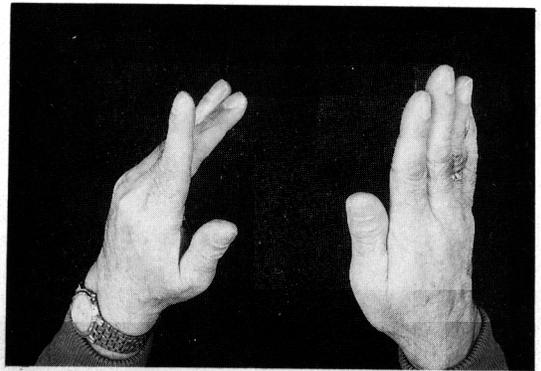


Fig. 2. Patient 1 had a spontaneous involuntary twisting movement of the left hand expressed as flexion of the metacarpophalangeal joints, and separation and extension of the thumb.

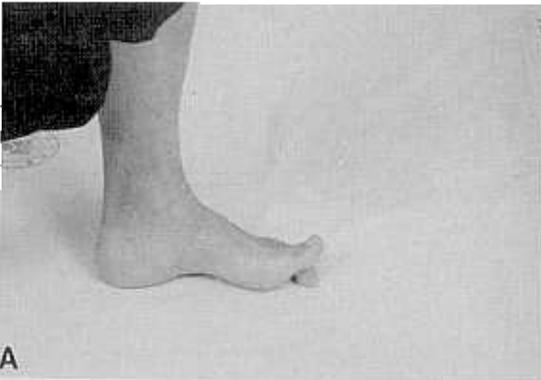


Fig. 3. The dystonic movement of patient 2 was sustained posture of dorsiflexion(A) and was intensified by walking which made him walking discomfort(B).

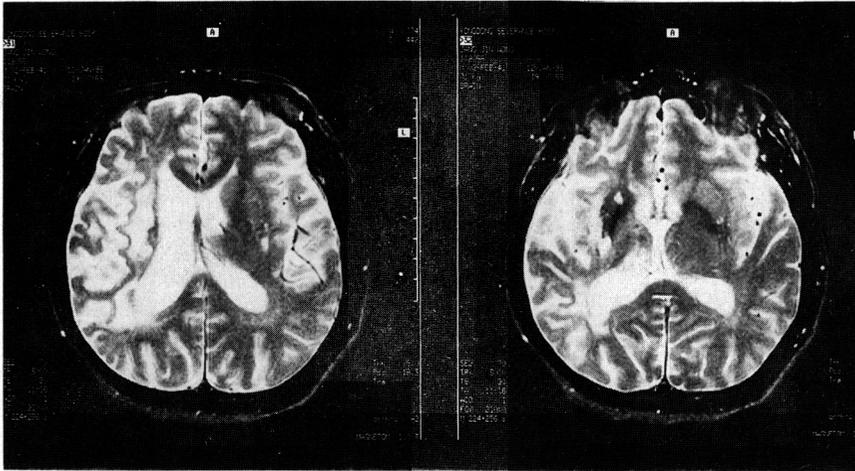


Fig. 4. Brain MRI of patient 2 showed an enlarged right frontal horn with high intensity changes involving the caudate nucleus, lentiform nucleus, and cortex (repetition time, 3000 msec; echo time, 80 msec).

Patient 2

A 52-year old man was referred due to dystonic posture in the left limb. He had a history of hypertension and suffered a sudden left hemiplegia 1 year ago. Examination at the time of admission revealed the left hemiplegia, dysarthria. Computed tomography of the head showed an infarction at the right middle cerebral artery territory. The left hemiparesis improved slowly and he was able to walk without support after several months. Nine months after the stroke he developed dystonic posture of the left great toe. The posture consisted of sustained dorsiflexion, which was aggravated during walking (Fig. 3). On examination, mild dysarthria and left facial weakness were noted. Questionable hemiparesis with hyperactive deep tendon reflexes was noted in the left limbs. Deep and superficial sensory deficits were noted in the left side. A routine hematological test, serum electrolytes, liver function test, and renal function test were normal. Serology for syphilis was negative. A Brain MRI showed an enlarged right frontal horn with high intensity changes involving the caudate nucleus, lentiform nucleus, and cortex (Fig. 4).

Patient 3

A 16-year-old boy was referred for the



Fig. 5. The left foot of patient 3 exhibited an intermittent or continuous spontaneous dystonic spasm of plantar flexion and slight inversion of the toes.

evaluation of the dystonic posture of the left leg. He suffered from sudden weakness of the left leg when he was 7 years old, diagnosed as cerebral infarction at that time. Along with the recovery over the next month, dystonic posture in left foot developed gradually. He had no family history of neurological disease or medical illness. There was no specific findings during pregnancy and delivery. His development has been nor-

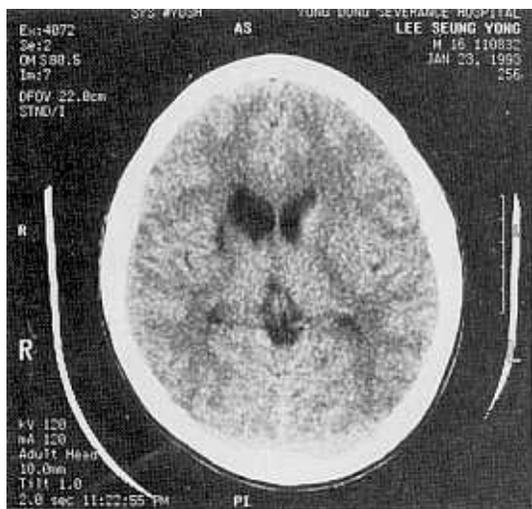


Fig. 6. A brain CT scan of patient 3 revealed an area of low attenuation adjacent to the right internal capsule in the caudate nucleus with an enlarged right frontal horn.

mal. On neurological examination, a very mild left leg monoparesis with hyperactive deep tendon reflex was noted. Deep and superficial sensation were normal. The left foot exhibited dystonic posture and intermittent spontaneous dystonic spasm, which consisted of plantar flexion and slight inversion of the toes (Fig. 5). Left limb muscle tone was increased. These dystonic posture and movements were aggravated by any attempted action of the foot, but it decreased markedly during relaxation and disappeared during sleep. A complete blood count, urinalysis, serum electrolytes, renal function tests, serum ceruloplasmin and copper level were normal. ANA and RF were negative. A brain CT scan revealed an area of low attenuation adjacent to the right internal capsule in the caudate nucleus with an enlarged right frontal horn (Fig. 6). Mild improvement of the dystonic posture was noted after administration of trihexyphenidyl.

DISCUSSION

Delayed-onset dystonia is a rare sequelae of

stroke (Traub and Ridley, 1982; Demierre and Rondot, 1983; Russo, 1983; Obeso *et al.* 1984; Chiang and Lu, 1990). The anatomical basis and pathogenesis of delayed-onset dystonia is uncertain. Mitchell (1974) suggested that the delay in the onset of hemichorea or athetosis following hemiplegia was caused by progressive changes in the original brain lesion. Burke (1980) had hypothesized that the mechanism of delayed-onset dystonia occurring a year or more after the insult may be due to aberrant neuronal sprouting. Pettrigrew and Jankovic (1985) studied 22 patients of delayed-onset dystonia following variable causes. They found the mean interval in the seven patients who had brain lesions below the age of 7 was much longer than the other patients, and postulated that the age of the patient at the time of cerebral injury influenced the latency of dystonia from the acute brain damage. However, the hypothesis of Pettrigrew has limitations. Some patients with immediate onset could not be explained with neuronal regeneration. Therefore we reviewed the literature to find the relationship between the age of the patients at the time of injury and the period of delay in stroke patients. 34 cases of cerebrovascular disorder (Table 1) have been reported. Dystonia following stroke almost always appeared within 1-12 months (mean 6.5 months). There were four patients with contralateral basal ganglia lesions (Table 1, patient No. 6, 14, 31, 32), who had a short delay despite the early age of hypoxic insult (Demierre and Rondot, 1983; Pettrigrew and Jankovic, 1985; Anegawa *et al.* 1986). In our series, one patient with dystonia following stroke in young age had a shorter interval compared to other elderly patients with dystonia following stroke. It became clear that there was no definite relationship between the time at the age of hypoxic insult and the interval of appearance of dystonia following stroke. In contrast, Factor *et al.* (1988) described a patient with long delayed-onset dystonia followed by embolic infarction at the age of 18 months. However, the possibility cannot be excluded that delayed-onset dystonia was caused by perinatal hypoxia due to fetal distress during delivery.

The pathological lesions in the patient of delayed-onset dystonia following stroke have a variety of anatomical lesion sites. Of 34 pa-

Table 1. Clinical characteristics and anatomical lesion in patients reported on the literature with poststroke dystonia (IC: internal capsule; Inf.: infarction)

Reference	No. of patient	Age at insult	Age of onset	Duration of delay	Site of dystonia	Etiology & pathology	Anatomical site
Burke(1980)	1	65	66	1yr	Lt arm	Infarction	Lt MCA territory (Lt MCA occlusion)
	2	54	55	1yr	Lt arm	Infarction	Normal
Grimes(1982)	3	32	32	6mo	Lt hand	Migraine	Rt. putamen, caudate, thalamus
	4	50	50	5mo	Lt arm	Infarction	Rt putamen, thalamus, temporoparietal cortex
	5	60	60	6mo	Lt arm	Infarction	Rt putamen, caudate, anterior IC, thalamus
Demierre (1983)	6	6	6	1mo	Lt hemi.	Infarction	Rt putamen, IC, caudate
	7	17	17	1mo	Lt hemi.	Embolic Inf.	Rt lentiform, caudate, IC
Glatt(1984)	8	68	68	4mo	Lt hemi.	Infarction	Rt thalamus, occipital, temporal cortex
Obesso(1984)	9	22	22	2mo	Lt hemi.	Migraine	Rt thalamus, striatum, IC
Sunohara (1984)	10	61	61	3mo	Lt hemi.	Infarction	Rt thalamus, IC
Burton(1984)	11	47	47	9mo	Lt hemi.	Infarction	Rt putamen, IC
Traub(1982)	12	43	46	3yr	Lt hand	Infarction	Rt lentiform nucleus
Russo(1983)	13	56	56	2mo	Lt U/E	Infarction	Rt lentiform nucleus
Pettigrew (1985)	14	2	2	1mo	Rt hemi.	Infarction	Lt striatum
	15	36	36	1mo	Lt hemi.	Infarction	Rt basal ganglia
	16	61	61	6mo	Lt hemi.	Infarction	Rt internal capsule
	17	46	46	1mo	Lt hand	Embolic Inf.	Rt basal ganglia
	19	72	72	1mo	Lt U/E	Infarction	Rt basal ganglia
	20	22	22	1mo	Rt U/E	Infarction	Normal
	21	22	22	1mo	Lt hemi.	Migraine	Rt internal capsule
	22	52	52	1mo	Rt hemi.	Hemorrhage	Rt IC, basal ganglia
Marsden (1985)	23	68	68	2mo	Rt hemi.	Infarction	Lt thalamus
	24	21	21	2mo	Lt hand	Infarction	Rt thalamus
	25	62	62	6mo	Lt hand	Infarction	Rt thalamus
	26	60	60	9mo	Lt hemi.	Hemorrhage	Rt thalamus
	27	52	52	4mo	Rt hemi.	Infarction	Lt lentiform, caudate, IC
	28	51	51	6mo	Rt foot	Infarction	Lt lentiform, caudate, IC
	29	49	50	1yr	Lt hemi.	Embolic Inf.	Rt lentiform, caudate, IC
	30	48	51	3yr	Lt hemi.	Infarction	Rt lentiform, caudate, IC
Anegawa (1986)	31	5	5	3wk	Rt hemi.	Vasculitis. Inf.	Lt putamen
	32	6	6	2mo	Rt L/E	Vasculitis	Lt putamen
Daras(1987)	33	42	44	2yr	Lt hemi.	SLE, Inf.	Rt putamen
Chiang(1987)	34	74	74	6mo	Rt hemi.	Infarction	Lt thalamus
Present case	1	53	54	1yr	Lt hand	Hemorrhage	Rt putamen, thalamus, caudate, ic.
	2	51	52	9mo	Lt foot	Infarction	Rt lentiform, caudate, cortex
	3	7	7	1mo	Lt foot	Infarction	Rt caudate, IC

tients reported in the literature, 21 (61.7%) had basal ganglia lesion, 13 (38%) internal capsule lesion, 11 (32%) thalamic lesion, 2 (5.8%) cortical lesion and 2 (5.8%) without any lesions. Damage to the neuronal circuit connecting caudate, putamen, globus pallidus and thalamus (Pettigrew and Jancovic, 1985; Fross

et al. 1987) seems to be responsible for the dystonia following stroke. However, it can occur without radiological evidence of striatal lesion (Glatt and Nausieda, 1984). Thalamic degeneration following striatal lesion or cortical lesion have also been reported (Oppenheimer, 1967; Dooling and Adams, 1975; Grimes *et al.*

1982).

In conclusion, the pathogenesis of the delayed-onset dystonia after stroke seems to be the retrograde neuronal degeneration (Powell, 1952; Russel, 1958; Peacock and Combs, 1965) through a damaged pyramidal tract following hypoxic brain damage, and we suspect that the age of hypoxic injury is not an important factor in deciding the interval between the brain damage and dystonia in stroke patients.

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