SUPRANUCLEAR BULBAR PALSY (PSEUDOBULBAR PALSY) IN MITRAL STENOSIS

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During the latter half of last century reports of cases referred to as "labio-glosso-pharyngeal palsy" began to appear in the English, French, and German literature. They were a heterogeneous group and included both the form now known as progressive bulbar palsy and other varieties in which bulbar palsy had been produced by bilateral lesions confined to the cerebral hemispheres. Oppenheim and Siemerling (1886) first attempted to differentiate between these groups, which they described as true bulbar paralysis and pseudobulbar paralysis. They added a description of the emotional lability which led to the latter condition being sometimes called "the laughing sickness."

The term "pseudobulbar palsy" has long been recognized as being unsatisfactory. Tilney and Morrison (1912) criticized it as giving no indication of either the site of the lesion or the nature of the disease. The synonym "supranuclear bulbar palsy" is more accurate, but "notwithstanding this," as Tilney and Morrison then commented, "pseudobulbar palsy, although a purely conventional term, has gained a fixed place in the literature." The traditional title dies hard and unfortunately has still not been displaced by the more appropriate and descriptive name of supranuclear bulbar palsy. The occasional combination of nuclear and supranuclear lesions shows how further confusion may arise if the terms "true" and "pseudo" are retained.

It is unnecessary to give an account here of the distinctive clinical features of supranuclear bulbar palsy, as they are fully described elsewhere. Oppenheim's (1911) early account is of interest, and good recent descriptions have been given by Langworthy and Hesser (1940), Alpers (1946), and Wechsler (1947). Kinnier Wilson (1924) has discussed the interesting related problem of pathological laughing and crying.

Supranuclear bulbar palsy most commonly results from arteriosclerosis which has produced bilateral areas of cerebral softening. In many cases, therefore, there is a history of an attack of weakness on one side of the body, followed later by an attack on the other side, the bulbar symptoms developing acutely after the second attack. In its sudden onset, its frequent association with double hemiplegia, its irregular non-progressive course, its lack of lower motor neurone involvement, and in its associated disturbance of emotional control, supranuclear or pseudobulbar palsy is distinct from progressive bulbar palsy (in which local bulbar involvement may be part of motor neurone disease). Supranuclear bulbar palsy may occasionally occur in the course of other pathological conditions (such as disseminated sclerosis, cerebral metastases, or agenesis) in which there are scattered bilateral lesions involving the cortico-bulbar tracts. It is a clinical syndrome and not a disease entity.

Several of the early descriptions of what we now regard as supranuclear bulbar palsy referred to young or middle-aged subjects with bilateral cerebral thromboses, probably of syphilitic origin. Two cases produced by cerebral embolism, however, were described by writers in the last century. Barlow (1877) published the clinical and post-mortem findings in "a case of double hemiplegia with symmetrical lesions." This referred to a boy, aged 10, with congenital heart disease and infective endocarditis. First a left and then a right cerebral embolism occurred, after which he developed the characteristic features of supranuclear bulbar palsy, including both emotional lability and urinary incontinence. Kirchhoff (1881) described the case of a man, aged 24, with mitral stenosis, who apparently developed bulbar paralysis from bilateral cerebral embolism. This resulted in drawling speech, dribbling of saliva, an almost immobile tongue, and bilateral facial weakness.

There seem to have been no further accounts of supranuclear bulbar palsy resulting from bilateral cerebral embolism, and we have been unable to find any modern reports of the condition occurring in rheumatic heart disease. Recently, however, we have seen three cases of mitral stenosis in which well-marked supranuclear bulbar paralysis was apparently produced by successive cerebral emboli. It may therefore be of interest to place the following examples on record.

Case 1

A trained nurse aged 47 was admitted to the Royal Northern Hospital on January 5, 1950. She had had rheumatic fever and chorea in childhood and heart trouble since then. She was known to have been in hospital in recent years with mitral stenosis and auricular fibrillation, and her pulse had remained irregular. She had felt well that afternoon, but on stretching to close a window she suddenly became faint and dizzy. She did not lose consciousness, but developed a headache and vomited. The left side of her face and the left upper and lower limbs seemed numb and she had difficulty in using her left arm.

On examination she was alert and her speech normal. Auricular fibrillation was present, with a pulse rate of 70 a minute and a systolic blood pressure of 120 mm. Hg. A diastolic and a systolic murmur were heard at the mitral area. Optic disks were normal. Detailed examination of the central nervous system was made next morning, when, apart from slight weakness of flexion of the left hip, there were no significant motor, sensory, or reflex abnormalities of the limbs. The only other development was weakness of the internal rectus muscle of each eye. It was thought that her attack was the result of a transient right-sided cerebral embolism, and that the subsequent ocular signs were probably the result of another small embolus involving part of the mid-brain.

During the night of January 8 she had another attack. Her pulse rate fell to 40 a minute and she appeared faint and confused. Next morning she was found to have a right hemiparesis with upper motor neurone type of weakness of face and limbs. A further cerebral embolism, this time involving the left side of the brain, had presumably occurred.

After this episode the whole clinical picture changed and she presented the characteristic appearance of a patient with supranuclear bulbar palsy. Her speech was rapid, spastic, and indistinct, and she had difficulty in swallowing and coughing. The tongue was spastic. She showed gross emotional lability, bursting into either tears or laughter without adequate cause. She became incontinent of faeces and urine. At this stage she had a pronounced jaw-jerk and a bilateral extensor plantar response. There was no sensory impairment. Lumbar puncture on January 11 showed normal cerebrospinal fluid (C.S.F.). The Wassermann reaction and Lange curve were negative.

The power of the right side of the body began to improve within two days and sphincter control was regained in two weeks. After about four weeks she was beginning to get about and walked with the short spastic steps reminiscent of the double hemiplegia of cerebral arteriosclerosis. The defective ocular movements had by this time almost gone. The spastic dysarthria and emotional lability improved gradually. When last seen, at the end of February, she no longer wept or laughed involuntarily, but still smiled and laughed in a slightly spastic fashion and with little retraction of the angles of her mouth. Some weakness of flexion of the hips, especially the right, persisted, and, though her walking had improved, she still took short spastic steps on a rather wide base. Her auricular fibrillation was unaltered. She felt well and was discharged to convalescence.

Case 2

A man aged 32 was referred to the neurological clinic of the Mid-Herts Hospital on October 20, 1949, for speech therapy. He had had rheumatic fever when aged 9, and since then was known to have had mitral stenosis. At the age of 29 he had had an attack of cardiac asthma.

He said that 18 months ago he had suddenly lost consciousness at 6.30 one morning. His wife had noticed jerking movements of his hands at the time. He recovered his senses by about 9 a.m., and then found that the right side of his face and the right upper limb were paralysed and the right lower limb was weak. He was drowsy, aphasic for a few days, and incontinent of urine. The hemiparesis and speech defect improved, and after about two weeks he noticed only clumsiness of the right hand.

Three weeks after this episode he had another attack in the early morning. His pulse was said to have been slow and irregular at the time. He developed sudden weakness of the whole of the left side of the body but did not lose consciousness. He became completely anarthric, his tongue was stiff and immobile, and he could not cough voluntarily. There was dysphagia with nasal regurgitation of liquids and dribbling of saliva. Incontinence of urine was followed by precipitancy and increased frequency. The left hemiparesis and the dysphagia improved during the next few weeks. He became able to speak in a whisper after three months and in a feeble voice after five months.

When seen in October, 1949, he noticed residual weakness on the left side and was subject to occasional involuntary flexion spasms of the left leg. He still had a little difficulty in chewing and swallowing, and said that his tongue felt stiff. Urgency of micturition persisted. His wife said that he had become easily tearful and irritable.

On examination he was alert but slightly emotional. There was no dysphagia, but he spoke in almost a whisper, with his teeth hardly separated. His speech was spastic, indistinct, and tended to accelerate. There was also a tendency to diminuendo repetition of the end of a sentence (palilalia). There was some bilateral weakness of the lower part of the face, although emotional movements were overactive. Eye movements and optic disks were normal. His tongue moved slowly and stiffly. A brisk jaw-jerk was obtained. All his limbs were slightly hypertonic and his movements slow. Power was slightly impaired on the left side and he walked slowly, dragging his left foot. Sensation was normal. The arm reflexes were brisk and equal and finger-jerks were obtained. The knee-jerks and ankle-jerks were also brisk, particularly on the left side. Abdominal reflexes were feeble. The right plantar response was equivocal and the left extensor. His pulse rate was 80 a minute and the rhythm regular. His blood pressure was 120/80 mm. Hg. The heart was slightly enlarged; a loud systolic and a mid-diastolic murmur were heard at the apex, and the pulmonary second sound was accentuated. On subsequent examinations, however, auricular fibrillation was sometimes present.

He was last seen some nine months later, when no further change was evident.

Case 3

A woman aged 20 was admitted semi-conscious to St. Thomas's Hospital on August 9, 1949. She had had chorea when aged 11 years and had begun to be easily breathless at 16. She had been attending the out-patient department since the age of 18 on account of rheumatic heart disease, and had signs of mitral stenosis with normal cardiac rhythm. During the two years that she continued under observation as an out-patient she was troubled by several attacks of nocturnal dyspnoea. She was of poor intelligence, but there were no other abnormal findings and her pulse had remained regular. When seen on the morning of August 9 no change in her physical signs was noted. That afternoon, however, her left hand and arm began to shake. Her husband said that she laughed and then suddenly fell unconscious in her chair. Her left arm appeared limp. She was brought into hospital about one and a half hours later.

On admission she was stuporous but reacted to painful stimuli. Her pulse rate was 70 and the rhythm regular. Eye movements, pupils, and optic disks were normal. Her lower jaw tended to hang open and the limbs were flaccid. The tendon reflexes were brisk and equal, and both plantar responses were extensor. The C.S.F. was entirely normal. The next day she was more conscious. She was dysphasic, but said a few words in a spastic and indistinct fashion. She was liable to outbursts of uncontrollable weeping and laughter and often made loud spastic cries for no good reason. She was incontinent of urine. Slight right facial weakness of upper motor neurone type was evident and there was some defect in conjugate deviation of the eyes to the right. Her palate moved poorly and she had difficulty in swallowing. On trying to drink she would dribble fluid from her mouth and sometimes through her nose. Power in the limbs was diminished, particularly on the left side, and she had to be fed. The tendon reflexes were brisker on the left side than on the right. The abdominal reflexes were absent, and both plantar responses were extensor.

During the next few days the power in her left side improved and she regained control of her bladder. Her speech, which was at first almost unintelligible, improved more slowly. She continued to show pronounced emotional lability and would laugh and cry in a spastic fashion for little or no cause. She got up after about two weeks and was discharged a week later. She then talked and walked fairly well. Both plantar responses remained extensor.

Though much improved when last seen, six months later, she was still easily moved to spastic laughter or tears and spoke with a nasal dysarthria. Her soft palate moved poorly and there was occasional slight dysphagia. Eye movements were normal. There was a residual right facial weakness. The tone of the limbs was normal. The left upper limb was slightly weaker than the right and there was some weakness of flexion of both thighs. She was unable to use her hands normally but she walked without difficulty. The tendon reflexes were brisk, more so on the left side than on the right. The plantar responses were equivocal and attempts to elicit them made her weep. She had full control of her bladder. There had been no further symptoms directly referable to the cardiovascular system.

Discussion

Each of these patients had a previous rheumatic history. All three had mitral stenosis and two of them showed auricular fibrillation.

In Case 1 the history suggested the occurrence of several emboli in the course of three to four days. First the right and then the left side of the brain was involved. With the development of the bilateral cerebral lesions the picture changed to that of a supranuclear bulbar palsy, uncontrolled laughing and crying being a prominent feature. Within two months the emotional symptoms had almost completely disappeared, though some spasticity of speech and gait remained.

Case 2 also had two acute cerebral attacks: the first resulted in a right hemiparesis, and the second, three weeks later, in a left hemiparesis. Supranuclear bulbar palsy developed with the second attack and disturbance of articulation was the most striking feature.

In Case 3 it seems likely that several small cerebral emboli affected both sides of the brain almost simultaneously. Typical supranuclear bulbar palsy immediately resulted and the spastic laughter and tears were very evident. Although emotional control improved considerably it was not complete six months later.

All three cases had marked dysarthria and dysphagia and all were temporarily incontinent of urine. The onset of supranuclear bulbar palsy was in each case abrupt, but all three showed considerable improvement.

In supranuclear bulbar palsy resulting from cerebral arteriosclerosis the vascular degenerative changes are likely to be diffuse and multiple. Bilateral pyramidal-tract signs are usually present and there may be a history of double hemiplegia, though there is often an interval of months or years between the involvement of the two sides. Sometimes the supranuclear bulbar features are accompanied by only a hemiplegia or hemiparesis. More rarely there is no limb paralysis at all, though the plantar responses may be extensor. In addition to the involuntary laughter and crying, progressive dementia is seen in the arteriopathic group and occasional extrapyramidal features have been described. The course of the disease is irregular and may be prolonged.

When supranuclear bulbar palsy is produced by bilateral cerebral embolism, however, there is usually a history suggesting sudden vascular occlusion on one side of the brain, followed quite rapidly by the same process on the opposite side, so that bilateral interference with the cortico-bulbar and cortico-spinal pathways immediately results. The embolic form of supranuclear bulbar palsy may occur in young subjects. Dementia is not a feature of the condition, and, if the emboli are small, rapid resolution of symptoms may occur.

Summary

Supranuclear bulbar palsy (so-called pseudobulbar palsy) most often results from diffuse cerebral arteriosclerosis, with or without a history of double—that is, bilateral—hemiplegia.

Three cases of mitral stenosis are described in which bilateral cerebral emboli resulted in the abrupt development of the characteristic syndrome of supranuclear bulbar palsy with bilateral pyramidal-tract signs.

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Dr. L. Leslie has been made vice-chairman of the Berkshire Executive Council in place of the late Dr. J. McCrea.

MORTALITY IN GERIATRIC SURGERY*

BY

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The life-span has been increasing markedly, as is shown by Chart 1. According to the National Office of Vital Statistics (1950), it is 67.2 years in the United States. Chart 2 indicates that those aged 65 and over constitute 7.6% of the population. The Royal Commission on Population (1949) reports that in 1947, in Great Britain, those who were 60 years and over constituted 15% of the population.

More attention is being focused on problems of ageing. It is our function to apply our art not only to save life but to add years to life and life to years, and to reduce mortality. The surgeon has come to play a most important part in the attainment of these goals. We have had an unusual opportunity for the practice of geriatric surgery at the Goldwater Memorial Hospital, in New York City. This 1,500-bed municipal hospital was opened in 1939 for the care and study of chronic disease. Most of the patients are old and are substandard risks. They are afflicted with chronic and degenerative diseases and with nutritional deficiencies. Nevertheless, we do not hesitate to meet surgical indications if there is a chance to save life or to prevent invalidism.

Only patients over the age of 60 are included in this report. Table I (Carp, 1946) shows comparative operative mortality statistics from a number of clinics. At our clinic, from 1939 to 1944, the mortality rate in 450 major

TABLE I.—Comparative Operative Mortality Statistics (Carp, 1946)

Source	Age Groups	No. of Major Operations			Mortality %		
		M.	F.	Total	M.	F.	Average
Goldwater Memorial Hospital*, 1939-44	61-70	152	50	202	15.78	22.0	18.89
	71-80	148	47	195	24.18	27.6	25.89
	81-90	38	12	50	44.73	25.0	34.86
	91-100		2	2			
	101-110		1	1			
	Total	338	112	450			22.66
Bailey (1934) Parsons and Purks (1942) Clagett (1943) Hay (1940) Evans and Key (1942)	60-84			185			7.6
	60 and over			100			17.0
	60 " "			1,204			9.0
	70 " "			536			16.6
	60 " "			83			7.2
	Grand total			2,558		Grand aver.	13.1

* The emergency operations were 12.88% of the total number.

TABLE II.—Goldwater Memorial Hospital: Major Operations on Substandard Risk Patients Over 60 Years of Age. Post-operative Mortality Rates Within 30 Days

Period	No. of Major Operations		% Emergency of Total	Post-operative Deaths Within 30 Days		% Mortality Rate	
	Total	Emergency		Total	Emergency	Total	Emergency
1945 ..	166	32	19.4	40	23	24.1	71.9
1946 ..	131	32	24.4	25	16	19.1	50.0
1947 ..	162	44	27.2	23	12	14.2	27.3
1948 ..	193	41	21.2	25	11	12.9	26.8
1949 ..	199	56	28.1	34	18	17.1	32.1
1945-9 ..	851	205	24.1	147	80	17.3	39.0
1939-44 ..	450	59	13.0	102	26	22.6	44.0

*Read before the Section of Geriatrics at the Annual Meeting of the British Medical Association, Liverpool, 1950.