

ACUTE PSEUDOBULBAR MUTISM DUE TO DISCRETE BILATERAL CAPSULAR INFARCTION IN THE TERRITORY OF THE ANTERIOR CHOROIDAL ARTERY

by CATHY HELGASON¹, ANDREW WILBUR², ANNE WEISS³,
KATHLEEN J. REDMOND⁴ and NANCY A. KINGSBURY⁵

(From the ^{1,3}Department of Neurology, ²Department of Radiology and ^{4,5}Department of Otolaryngology—Head and Neck Surgery, University of Illinois Medical Center at Chicago, Chicago, Illinois, USA)

SUMMARY

Pseudobulbar mutism is rarely attributed to bilateral discrete posterior limb internal capsule-medial globus pallidus infarction. Few cases of bilateral anterior choroidal (AchA) artery territory infarction have been reported. We present 8 patients with ischaemic stroke in this location and vascular distribution who have a characterizable syndrome. All had the abrupt onset of inability to speak, swallow or phonate, accompanied by varying degrees of facial diplegia, hemiparesis, hemisensory loss, lethargy, neglect and change in affect. The appearance of clinical signs depends upon the presence of a new infarct contralateral to an older lesion in mirror position. The pathogenesis and progression of neurological deficit appears to be intimately related to hypertension. The role of intrinsic intracranial vascular pathology related to diabetes mellitus, embolism of cardiac origin and atherosclerosis is currently undefined. The prognosis for recovery is poor. Half of our patients died within a year of onset of symptoms. Capsular pseudobulbar mutism is recognized by the abrupt appearance of neurological deficit consistent with internal capsular pathology and is confirmed by CT scan or MRI.

INTRODUCTION

Acute mutism with pseudobulbar palsy due to discrete bilateral internal capsule infarction is rare. Reports bearing this clinical-anatomical correlation have emphasized interruption of the corticobulbar tract at the capsular genu as the locus of pathology. We now highlight the pertinent features of 8 cases of acute pseudobulbar paralysis with a severe degree of speech loss due to ischaemic stroke. Infarction in these patients was localized by CT scanning and MRI to the posterior two-thirds of the posterior limb of the internal capsule and adjacent medial globus pallidus, in the territory supplied by the anterior choroidal artery (AchA).

Correspondence to: Dr Cathy M. Helgason, Department of Neurology, University of Illinois Medical Center at Chicago, 912 S. Wood Street, Chicago, Illinois 60612, USA.

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CASE REPORTS

Case 1

This 61-yr-old right-handed woman had a 10 yr history of hypertension and 45 yrs of heavy smoking. For 6 months before admission, she had had intermittent bowel and bladder urgency. One month before, she had developed weakness in her right arm and leg with slurred speech. These symptoms improved but she was left with burning pain in the leg. In addition, she complained of intermittent blurring of vision of both eyes. On admission, the blood pressure was 190/100 mmHg; the heart rate was regular, 70/min. She was alert and orientated. Arteriovenous nipping was prominent on fundoscopy. The pupils and eye movements were normal. She had a depressed corneal reflex and facial perception of pin-prick on the right. The right half of the soft palate did not elevate on phonation. The right arm and leg were moderately weak and ataxic. She used a cane in order to walk. Pin-prick and touch sensation on the right was decreased. The tendon reflexes were bilaterally brisk, more so on the right, and there were bilateral extensor plantar responses.

Two days after admission the patient complained of new left-sided weakness and increased slurring of speech, the weakness progressing that day to hemiplegia. The next day urinary retention was present and over the following weeks intermittent catheterization was required. She regained strength in the left arm, but the left leg remained paralysed. Intermittently during this time, despite medication, her blood pressure was raised, at times up to 210/120 mmHg. Suddenly, 6 weeks after admission, the patient was found lethargic with a blood pressure of 240/120 mmHg. On examination she was cooperative. There was a left head and eye gaze preference as well as right-sided auditory neglect. She was unable to move her eyes past the midline to the right. The oculocephalic reflex was intact. The pupils were normal, the right corneal reflex absent and there was severe facial diplegia, eye closure equally weak to mouth movement. Her gag reflex was absent, the palate hung low in the back of the throat, and she was unable to swallow, phonate or protrude her tongue. Tongue movement was slowed and limited in range. She could not approximate her lips. The right-sided hemiparesis had now become complete. Bilateral extensor plantar responses, left leg paralysis, hyperreflexia and sensory findings were unchanged. A CT scan showed periventricular lucencies and a discrete right-sided lucency in the middle third of the posterior limb of the internal capsule. Repeat CT scan and MRI 1 week later showed evolution of the right lucency and a new lucent area in mirror position on the left. Both lucencies now extended superiorly into the caudate nucleus immediately lateral to the lateral ventricle and adjacent white matter (fig. 1). Other laboratory results are summarized in Tables 1-3.

A gastrostomy was performed because of persistent dysphagia. Two months after admission, the left leg remained paralysed and the other limbs were mildly paretic. Her pseudobulbar paralysis remained severe. Her speech had slightly improved but was limited to severely distorted single words. Her voice was breathy, low pitched, monotonous and hypernasal. Although intermittently lethargic, she could now communicate with the help of an alphabet pointing board and by nodding 'yes' and 'no'. She was discharged to a nursing home and 1 year later was still alive.

Case 2

This 68-yr-old right-handed man developed sudden numbness of the left leg one evening. During the night, his wife noted that he had unintelligible speech and marked weakness of the right face and leg. The following morning he could not stand. He had a 7 yr history of untreated hypertension. He had had several strokes, causing repeated weakness of the right limbs, minimal slurring of speech and mild dysphagia, bowel and bladder incontinence and depression.

On admission, blood pressure was 160/100 mmHg, the pulse was 96/min and regular. He was alert but mute. Pupillary reactions and eye movements were normal. There was right-sided face weakness, affecting the upper and lower face, the gag reflex was depressed and the tongue deviated to the right. There was spastic paralysis of the right arm. There was moderate weakness of the right leg, mild left arm and leg weakness, bilateral hyperreflexia, greater on the right, and a right extensor plantar response. In the following days the patient was intermittently lethargic. He developed a left extensor

TABLE 1. CLINICAL COURSE

Case	Age (yrs), sex	Onset	Outcome	CT	Angiography and carotid Doppler sonography	Possible mechanism
1	61F	Stepwise onset with sudden deterioration and transient BP elevation	Gastrostomy; nursing home; severely impaired	PVL, bilat. middle $\frac{1}{2}$ posterior limb capsule extending superiorly	Unknown	Small vessel disease related to hypertension or diabetes
2	68M	Stepwise onset with smooth deterioration	Gastrostomy; death within 1 yr of onset	R posterior $\frac{1}{2}$ posterior limb internal capsule, L entire posterior limb	Doppler: normal common carotid, suboptimal study for distal vessels, calcified carotids on x-ray	Small vessel disease related to hypertension
3	67F	Stepwise onset, then sudden deterioration with abrupt BP drop	Gastrostomy; death within 1 mo. of onset	Bilat. middle $\frac{1}{2}$ posterior limb internal capsule and adjacent globus pallidus extending superiorly	Doppler: technically limited. Angiogram: narrowing of petrosal carotid and attenuation of AchA on left	Small vessel disease related to hypertension or diabetes; intracranial atherosclerosis
4	53F	Stuttering onset with waxing and waning symptoms then smooth deterioration	Home with moderate-severe impairment	PVL, R middle $\frac{1}{2}$ posterior limb, L middle $\frac{1}{2}$ posterior limb extending superiorly	Normal Doppler	Small vessel disease related to hypertension, diabetes; vasculitis; embolism of possible cardiac origin; possible atherosclerosis
5	71F	Stepwise onset with abrupt deterioration	Gastrostomy; death within 1 mo. of onset	Bilat. middle $\frac{1}{2}$ posterior limb internal capsule extending superiorly	Unknown	Small vessel disease related to hypertension; radiation-induced vascular changes
6	62M	Sudden onset and stepwise deterioration with transient BP elevation	Gastrostomy; rehab. unit, near locked-in at 1 mo.	R post limb internal capsule, and lateral geniculate, L posterior $\frac{1}{2}$ posterior limb extending superiorly	Minimal atheroma bilaterally by Doppler	Small vessel disease related to hypertension or diabetes
7	46F	Sudden onset with no deterioration	Home with moderate impairment	PVL R posterior $\frac{1}{2}$ posterior limb internal capsule and adjacent globus pallidus, L mirror lesion	Normal Doppler	Small vessel disease related to hypertension
8	79M	Stepwise onset	Death within 4 mo. of onset	Bilat. posterior $\frac{1}{2}$ posterior limb internal capsule and adjacent globus pallidus extending superiorly	Unknown	Small vessel disease related to hypertension, diabetes; syphilis

PVL = periventricular lucencies; AchA = anterior choroidal artery.

TABLE 2. FURTHER CLINICAL DATA

<i>Case</i>	<i>Cardiac disease</i>	<i>Previous stroke</i>	<i>Hypertension</i>	<i>Diabetes</i>	<i>Admission BP</i>	<i>Fasting blood glucose (mmol/l)</i>	<i>Smoking</i>	<i>Other</i>
1	Ischaemic heart disease	—	10 yrs	—	190/100	3.9	45 yrs, heavy	
2	Hypertensive	+	7 yrs	—	160/100	5.0		HbAS
3	Ischaemic heart disease	+	20 yrs	20 yrs	220/120	26.9		
4	Ischaemic cardiomyopathy	+	7 yrs	1 yr	220/106	10.8	Heavy	HbAC+ rheumatoid factor
5	Chronic AF; ischaemic heart disease	—	+	—	160/110	5.1	Heavy	
6	Not known	+	+	+	200/100	12.8		
7	1° AV block	+	Many yrs	—	160/98	6.4		Elevated triglycerides
8	Ischaemic cardiomyopathy	+	20 yrs	20 yrs	180/90	22.6		+ RPR and FTA-abs.

RPR = serum syphilitic reagin.

TABLE 3. MAXIMAL SYMPTOMS AND SIGNS
Case

	1	2	3	4	5	6	7	8
Lethargy	+	+	+	-	+	+	-	-
Pseudobulbar affect	-	-	+	+	-	-	+	+
Depression	-	+	-	+	+	+	-	-
Head or eye gaze preference	L	-	R	-	L	L	-	R
Visual field defect	-	?	?	-	?	L homonymous hemianopia	?	?
Severe paralysis tongue, oropharynx	+	+	+	+	+	+	+	+
Mute	+	+	-	+	+	+	-	-
Hemiplegia	R then L	R arm	R arm	L leg	L	R	R, L leg	R face, L body
Hemisensory loss	R	?	R	Bilat.	L	R	?	?
Quadriparesis	+	+	+	+	-	-	-	+
Urinary/bowel incontinence or retention	+	+	-	+	-	-	-	+
Extensor plantar responses	Bilat.	Bilat.	Bilat.	Bilat.	Bilat.	Bilat.	Bilat.	Bilat.
Other	R auditory neglect		R tactile extinction	Cheyne-Stokes	Denial of deficit, L hemi-inattention, constructional apraxia			Confusion

+ = present, - = absent, ? = unknown.

plantar response and left facial weakness, and was only able to grunt in a strained voice. He was now unable to protrude his tongue or to pucker his lips. The gag reflex was absent and the palate did not move on phonation. He was unable to swallow and frequently coughed and choked on his saliva. A gastrostomy was performed.

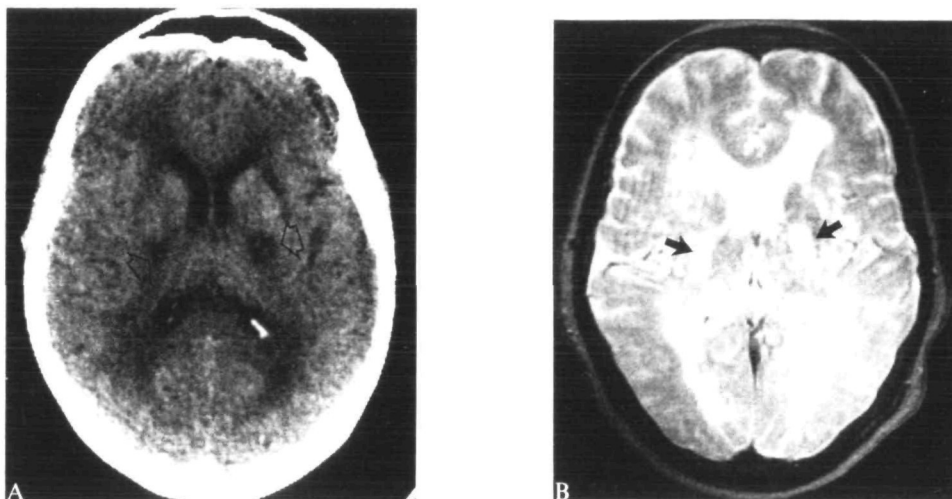


FIG. 1. Sequential bilateral development of infarction in the posterior limb of the internal capsule extending superiorly as shown in CT scan (A) and MRI (B) in *Case 1*.

A CT scan showed a new lucency in the middle third of the posterior limb of the right internal capsule and the entire posterior limb on the left. An MRI scan showed similar findings with the right-sided lucency in the posterior two-thirds of the posterior limb of the capsule (fig. 2). Tables 1–3 summarize the laboratory results.

One month after admission the patient remained bedridden and could communicate only with his left hand by pointing to a picture board with variable success. He was discharged to a rehabilitation centre 1½ months after admission. At 1 yr follow-up he was reported to be dead after aspiration pneumonia.

Case 3

This obese 67-yr-old, right-handed woman had a 20 yr history of hypertension and diabetes mellitus. Three years before admission she had undergone an extracranial-intracranial bypass operation for a right hemisphere stroke believed to be related to stenosis of the petrous portion of the right internal carotid artery as defined by angiography. Her stroke had left her with a mild left hemiparesis. She now had an abrupt onset of slurred speech one evening. The following morning she awoke with increased dysarthria. Later that day, she became weak on the right and was unable to get off the toilet. On admission her blood pressure was 220/120 mmHg. This was reduced rapidly to 140/85 mmHg. Concomitantly her neurological state deteriorated; she was now lethargic and hemiplegic. The following day, still intermittently lethargic, she was able to cooperate on examination. Fits of uncontrollable laughter and a right motor neglect were present. Right tactile but no auditory or visual extinction was noted. The pupils were normal. She was unable to gaze fully conjugately to the right. The oculocephalic reflex was normal. A severe facial diplegia, mouth more than eyes, was present, worse on the right. She was markedly dysarthric and unable to swallow. She coughed on her saliva. Lip

approximation was weak, and the range of movement, strength of protrusion of the tongue were severely limited. Lingual movements were slowed. Her soft palate drooped bilaterally and did not elevate on phonation. The gag reflex was absent on the right. Articulation was severely impaired and speech was limited to one syllable utterances. The voice was strained and hypernasal in quality. The right arm was paralysed, the leg weak and sensation on the right was depressed for pin-prick and touch, position sense being spared. The tendon reflexes were brisk and both plantar responses were extensor.

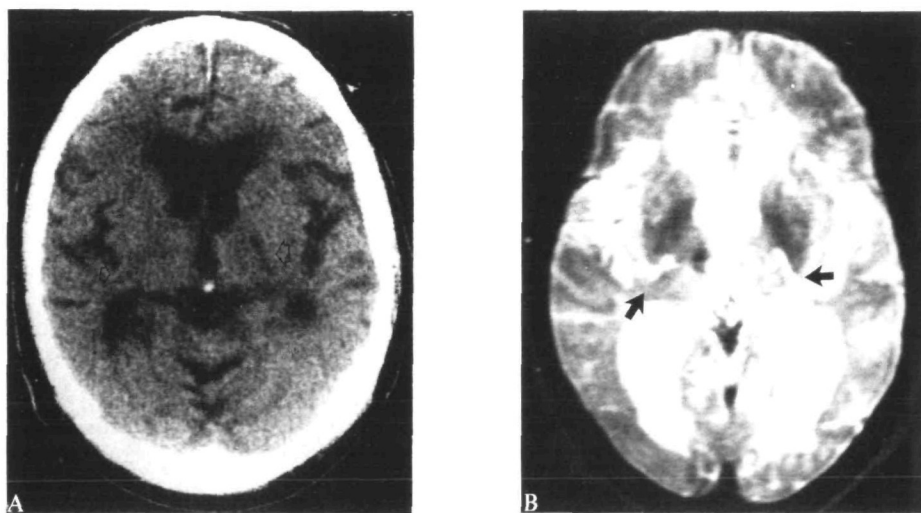


FIG. 2. CT scan (A) and MRI (B) in *Case 2* showing infarction in the posterior limbs of the internal capsules.

A CT scan showed bilateral lucencies in the posterior limb of the internal capsule in its middle third and the adjacent globus pallidus on the left with extension superiorly into the caudate nucleus and adjacent white matter immediately lateral to the lateral ventricle. Retrospective review of a CT scan performed in 1982 showed that the right capsular lesion was then present. It was now better defined in longitudinal extent. Tables 1–3 give other laboratory results.

A gastrostomy was performed and the patient's subsequent hospital course was marked by the new development of inferior myocardial ischaemia on ECG and ultimately by sudden respiratory followed by cardiac arrest and death 1 month after the onset of her symptoms.

Case 4

This 53-yr-old right-handed woman had a 7 yr history of hypertension and a 1 yr history of diabetes mellitus and congestive heart failure. One year before she had had a stroke causing left-sided weakness from which she had largely recovered. For 2 weeks, she had had variable slurring of speech accompanied by a hoarse voice. Subsequently her speech progressed to unintelligible mumbling. She had also noted increased weakness of the left leg and a tendency to fall to the right. She was noncompliant with medications, a heavy smoker and had had intermittent urinary incontinence since the first stroke.

Blood pressure on admission was 220/106 mmHg, the pulse regular at 88/min. Cheyne-Stokes respiration was present. A systolic ejection murmur was present at the left sternal border. The patient was markedly dysarthric, had mild weakness of the left leg, bilateral hyperreflexia and extensor plantar

responses. Over the following few days she showed increased dysarthria, dysphonia and hypernasality. She was unable to protrude her tongue. Lip and tongue movements were slowed and palatal elevation was absent on the right. She was dysphagic. A moderate facial diplegia was present, worse on the left. There was severe weakness of the left leg, increased weakness of the left arm, and bilateral hypalgesia of the face and left leg for pin-prick. Two weeks later, there was moderate improvement in her speech and swallowing and she was discharged to a rehabilitation centre with moderate dysarthria, resolved dysphagia, a right hemiparesis, and weakness of the left leg. One year later she was walking with the aid of a frame and, although otherwise unchanged, had uncontrollable fits of crying.

A CT scan showed a focal lucency in the middle third of the posterior limb of the internal capsule near its junction with the posterior third on the right. On the left, a focal lucency was seen in the middle third of the posterior limb of the capsule extending superiorly into the caudate nucleus and white matter adjacent and lateral to the lateral ventricle. A separate lucency was seen in the anterior limb of the capsule but the genu was spared. Periventricular lucencies were present. Other laboratory findings are summarized in Tables 1-3.

Case 5

This 71-yr-old woman had had bilateral radical neck dissection and radiation for oral cancer 8 yrs before. She was a heavy smoker and had a history of hypertension as well as of chronic atrial fibrillation. She suddenly developed slurred speech and mild weakness of the left face, arm and leg. On admission, her heart rate was irregular at 100 beats/min; blood pressure was 160/110 mmHg. She was intermittently lethargic and had a blunted affect. She denied any new deficit until specifically asked. She bisected a line to the right of the midpoint and had mild difficulty copying a design. There was a right gaze preference and incomplete conjugate gaze to the left. Her right pupil was smaller than the left, but this was of long standing. There was marked left facial weakness, mouth more than eye, as well as reduced forehead wrinkling on that side. Her mouth hung open at rest and she was unable to close it. Her dysarthria was severe and her voice low-pitched and hypernasal. The gag reflex was depressed, more so on the left, and palatal elevation on phonation was absent. Tongue movements were slow and restricted in range. A mild left hemiparesis was present, face and arm more than leg, worse proximally. There was tactile extinction for touch on the left as well as reduced appreciation of temperature and pin-prick which had a dysaesthetic quality. Position sense was intact. The tendon reflexes were brisk bilaterally and both plantar responses were extensor. Two days later the patient deteriorated abruptly. Although more alert, there was no initiation of speech and she had a marked facial diplegia, involving eye closure, forehead wrinkling and mouth movement. Dysphagia was severe. Her blood pressure was 210/128 mmHg.

A CT scan showed bilateral internal capsule lucencies, located in the middle third of the posterior limb and extending superiorly into the caudate nucleus and adjacent white matter immediately lateral to the lateral ventricle. Tables 1-3 give other laboratory findings.

A gastrostomy was performed. There was no clinical improvement and 4 weeks after admission, the patient died in a nursing home of sudden cardiorespiratory arrest.

Case 6

This 62-yr-old right-handed man 4 yrs before admission had undergone craniotomy for removal of a pituitary adenoma. Since then, he had received thyroid and steroid replacement medication and had developed diabetes. One year before admission he had had an episode of transient dysarthria. On the day of admission, while kneeling and praying aloud, his speech suddenly became slurred and he had momentary bilateral arm and leg weakness accompanied by tingling. On admission, blood pressure was 200/100 mmHg and the pulse was 96/min. An old left homonymous hemianopia was noted. The pupils reacted sluggishly to light; this had also been noted previously. There was left head and eye gaze preference. His eyes moved slowly and inconsistently but conjugately to the right. The oculocephalic reflex was intact. A depressed right corneal reflex was noted. He had facial diplegia, the right eye and mouth muscles being severely affected, the left moderately so. Forehead wrinkling was

diminished on the right. He could pucker his lips but at rest they gaped open. Jaw strength was normal but palatal elevation on phonation was weak. He had severely limited protrusion of the tongue and tongue movements were weak and slow. The gag reflex was absent, and the patient coughed and choked on his saliva. Involuntary smiling was preserved. His voice was strained and hypernasal. A moderate right hemiparesis was present, the arm and leg being affected equally. Diminished pin-prick appreciation on the right, bilateral hyperreflexia, left more than right and a right extensor plantar response were also found. Over the next 24 h the patient had a transient blood pressure rise of 240/120 mmHg and his condition deteriorated. He became totally hemiplegic and mute, able only occasionally to grunt. Lip movement was severely restricted and he was unable to move the tongue. He was lethargic and had Cheyne-Stokes respiration. Nonetheless he remained responsive and cooperative to command. Sensory deficit to deep pain and pin-prick was now severe on the right. A CT scan on the day after admission showed a lacunar infarct in the posterior third of the posterior limb of the right internal capsule. MRI 4 days after onset showed a new lesion in the posterior limb of the left internal capsule in its posterior two-thirds and the adjacent globus pallidus, with superior extension into the adjacent caudate nucleus and white matter immediately lateral to it. Repeat CT showed evolution of the right-sided lesion which was now better defined.

A gastrostomy was performed. Minimal improvement of facial and tongue movement occurred and he was moved 3 weeks after the onset of his stroke to a rehabilitation unit. Hemiplegic and mute, he was able to communicate only by weakly nodding his head and using his left hand to point.

Case 7

This 46-yr-old woman had a history of four previous strokes, with a residual moderate right hemiparesis and mild dysarthria. She is hypertensive and has had seizures for many years but has been noncompliant with medication. She suddenly developed increased slurring of speech, inability to swallow, increased right-sided and new left-sided weakness, and inability to walk. Blood pressure on admission was 160/98 mmHg, the pulse regular, 76/min. Her left radial pulse was weaker than the right. Uncontrollable fits of laughter occurred, but she denied any emotional content for these outbursts. Her left pupil was unreactive to light. Her jaw jerk was increased and there was left facial weakness of moderate severity affecting eye closure and mouth movement. The palate drooped at rest and did not elevate fully on phonation. The gag reflex was absent on the left. Lip and tongue movements were slow, weak and awkward and her voice was strained, harsh and hypernasal. Her speech was explosive and severely distorted. The right arm and leg were mildly but equally weak and there was a left foot drop. Both plantar responses were extensor.

A CT scan showed a discrete lucency in the upper portion of the posterior third of the posterior limb of the right internal capsule near the junction with the middle third and adjacent medial globus pallidus. On the left there was a well defined lucency in the mirror position in the posterior limb of the internal capsule. Periventricular lucencies were noted.

The patient was discharged to a rehabilitation centre where she improved her gait and swallowing ability. She did not require a gastrostomy. Six months later she was unchanged.

Case 8

This 79-yr-old man had a 20 yr history of hypertension, diabetes mellitus and more recent chronic renal insufficiency as well as peripheral vascular disease necessitating a right leg amputation. He was noncompliant with medications. Eight years before he had had a stroke which left him with a moderate right spastic hemiparesis. One day before admission he suddenly became unable to move his left arm or leg and seemed confused. Two months previously, he had had intermittent bladder and stool incontinence and 3 days before admission had complained of chest pain, became unable to speak clearly or swallow, and regurgitated his food. Blood pressure on admission was 189/90 mmHg; the pulse was regular, 82/min. He was obese and had fits of uncontrollable crying for which he denied any emotional content. There was motor impersistence for eye closure and no recognition of the new left hemiparesis. His pupils reacted sluggishly to light. There was a right gaze preference, but conjugate eye

movements were full. A severe right facial weakness affecting voluntary forehead wrinkling, eye closure and smiling was present. On the left, moderate similar deficits were present. He had decreased palatal elevation on phonation and his voice was hoarse and hypernasal. Food pocketed in both cheeks and swallowing was delayed. There was a right spastic hemiplegia, tone was increased in the left arm and leg. The left arm was moderately paretic the leg mildly so. On sensory testing, his replies were inconsistent for pin-prick and touch, but position sense was intact. The tendon reflexes were brisk, especially on the right, and the left plantar response was extensor.

CT scanning revealed bilateral lucencies in the posterior two-thirds of the posterior limbs of the internal capsules, each extending into the adjacent medial globus pallidus and superiorly into the caudate nucleus and white matter immediately lateral to it, especially on the left. His blood FTA test was positive.

While in the hospital, the patient developed chest pain of acute onset lasting for 20 min, but without new ECG findings. Over the week following admission, his pseudobulbar symptoms improved slightly as did his left-sided weakness and he was able to return home. Three months later he had a fatal cardiorespiratory arrest at home.

DISCUSSION

Mutism of sudden onset with pseudobulbar palsy has rarely been attributed to circumscribed bilateral capsular infarction. In our 8 cases, sequential bilateral capsular destruction with interruption of efferent pathways from the motor speech cortex to the bulbar apparatus produces mutism with paralysis of face, oropharynx and tongue.

Marie (1906) described acute anarthria without aphasia due to unilateral lentiform nucleus haemorrhage. He postulated an 'anarthric' area of brain, the destruction of which would cause the loss of speech. This area lies between transverse lines drawn from the anterior and posterior limits of the insula to the lateral ventricle and medial border of the hemisphere and includes these structures. More particularly he considered that the 'zone lenticulaire', white matter between the insula and lentiform nucleus and including the external portion of the latter, to be the area responsible for anarthria in his case. His patient had a caudato-putaminal-capsular haemorrhage at autopsy. The presence of associated pseudobulbar symptoms in his patient was not mentioned. Our patients with bilateral capsular-pallidal infarcts have in addition to anarthria, bulbar, hemimotor and hemisensory signs. As seen by CT scan and MRI their strokes lie posteriorly and medially in Marie's 'lesion d'anarthrie', in the territory of the anterior choroidal artery (AchA).

Unilateral infarction of the AchA territory has been discussed recently by Decroix *et al.* (1986), Helgason *et al.* (1986) and Bogousslavsky *et al.* (1986). None of these patients showed pseudobulbar features and only one was mute. However, dysarthria was a common finding and found in 2 out of 5 cases of Helgason *et al.*, 6 out of 16 of Decroix *et al.* and all those of Graff-Radford *et al.* (1985). We have seen one unreported case of unilateral AchA territory infarction accompanied by severe bulbar dysfunction, right hemiplegia and hemisensory loss. He had severe dysarthria, right-sided facial paralysis, tongue deviation to the right, absent palatal movement and total inability to swallow. Unlike the bilateral cases, he made a rapid

recovery. The patient needed a gastrostomy tube but over 8 weeks regained total ability to speak and swallow. A CT scan revealed a lucency in the posterior limb of the left internal capsule extending into the adjoining caudate nucleus adjacent to the lateral ventricle in the territory of the AchA.

Case 16 of Decroix *et al.* (1986) with unilateral AchA territory infarction presented with an initial loss of speech (mutism), a moderate right hemiparesis with faciobrachial predominance, and ataxia of the right arm. A new lucency in the capsular posterior limb on the left was found, but an old contralateral anterior limb lucency was present. The clinical significance of the latter is unknown. There was no mention of other bulbar dysfunction. This case represents the only reported case of unilateral AchA infarction with mutism.

Unilateral pseudobulbar paralysis has been described in unilateral capsular infarcts. Reports have emphasized the genu as the site of pathology. Iwata (1984) reviewed a case of unilateral palatal paralysis which he believed was caused by unilateral interruption of the corticofugal motor pathway at the level of the corona radiata and adjacent capsular genu. A small left lenticulocapsular haematoma which presented as right faciobrachial paresis, dysarthria and right lingual paresis was described by Bogousslavsky and Regli (1984) as an incomplete 'opercular' syndrome. Again the capsular extent of the lesion was limited to the genu and anterior part of the posterior limb. Langworthy and Hesser (1940) reviewed unilateral injury of corticobulbar fibres. This may cause transient loss of function of bulbar muscles supplied by cranial nerves V and VII as well as gaze preference. Minimal palatal elevation with deviation of the uvula to the unaffected side, pharyngeal and laryngeal symptoms can occur and the tongue may be more severely as well as permanently affected as in our unreported patient with unilateral AchA territory infarction.

It would seem justifiable to conclude that unilateral capsular infarcts can on rare occasions present with acute mutism and pseudobulbar signs. Mutism when due to infarction at capsular level depends on the presence of bilateral disease, and may often be traced to infarction in the territory of vascular supply of the AchA, in a capsular-pallidal distribution.

Few accounts of mutism due to bilateral internal capsular infarction exist. Fisher (1979) examined a man (his Case 5) who had infarcts which were of lacunar size, of different ages, the older being localized pathologically to the midportion of the posterior limb of the internal capsule on the right, the more recent to the capsular genu on the left. This patient had had a previous left hemiparesis which had begun acutely 3 months before and from which he had almost entirely recovered. The more recent stroke began suddenly with right-sided weakness, aphonia and total paralysis of the lips and tongue. The patient had had hypertension of long standing. At necropsy stenosis of the lumen of branches of the AchA was found bilaterally. In places arterial wall pathology appeared to be atheromatous in nature, at others lipohyalinotic. Leys *et al.* (1985) reported a case of sudden paralysis of facio-labio-glosso-velo-pharyngeal distribution without limb weakness due to bilaterally

symmetric genu-corona radiata junction infarction. The lesion was identified by CT scanning and attributed to watershed ischaemia of the territory supplied by the external lenticulostriate branches of the middle cerebral artery and end branches of the posterolateral choroidal artery. Leys *et al.* postulated undocumented hypotension to be responsible. A single case of acute pseudobulbar palsy ascribed to bilateral capsular infarction lying in AchA territory of vascularization was reported by Buge *et al.* (1979). The patient suddenly developed paralysis of lingual, pharyngeal and laryngeal function accompanied by severe dysarthria, facial diplegia, worse on the right, limited vertical movement of the right eye, and bilaterally brisk tendon reflexes. At necropsy, bilateral symmetric posterior limb capsular-medial globus pallidus infarcts were delineated. The old infarct on the right had no clinical correlate from the history. The aetiology was judged to be thromboembolic, with emboli arising from ipsilateral proximal internal carotid artery atheroma. The patient was a heavy smoker, had an atrial septal defect, and was hypertensive and diabetic. Thus embolism of cardiac origin or intrinsic small vessel disease possibly played a pathogenic role. Chia (1984) had a patient with tetraplegia, mutism and total paralysis of the face, jaw, tongue and palate, and for gag and coughing. The patient was young and had no history of diabetes, hypertension or heart disease. The precipitating event was haemorrhagic shock. CT scanning showed lucencies localized to the posterior limbs of both internal capsules and the caudal parts of the genu in the end zone of supply of branches of the posterior communicating arteries. The patient of Mohr (1986) had an abrupt onset of anarthria, severe dysphonia and dysphagia with mild right arm weakness. Infarcts of sequential appearance as defined by CT scanning were responsible. The addition of a new lacune in the genu of the left internal capsule to a previously identified infarct in the posterior limb on the right yielded the acute symptoms.

The infarctions in our cases were localized to the posterior limb of the internal capsule and medial globus pallidus (*see* fig. 3). They resemble the case of Buge *et al.* (1979) anatomically as well as clinically. In 6 patients the lesions extend superiorly to involve an area immediately lateral and adjacent to the lateral ventricle. This site most probably represents the caudate nucleus and white matter immediately lateral to it, the most inferior portion of the corona radiata. The lucencies noted in our patients on the higher CT cuts represent the extension of an AchA infarct, particularly when seen to occur in direct continuity with those noted in the CT cuts below (H. Damasio, personal communication). Damasio has justly indicated that the AchA supplies the tail of the caudate nucleus. When infarcted in its superior and anterior portion, a lucency would appear on CT scanning immediately adjacent to the lateral ventricle. Damasio's CT template (Damasio, 1983, 1987) and the definition of cerebral arterial supply obtained by the injection of 87 brains (Beever, 1907) show AchA territory images very similar to the location of stroke in our patients.

In all our cases except one (Case 2), the posterior two-thirds of the internal capsule was involved. Importantly, the genu of the internal capsule was never

involved. Other authors have emphasized corticobulbar interruption at the genu as causative for pseudobulbar symptoms and mutism. The genu was infarcted in the patients of Fisher (1979), Leys *et al.* (1985), Chia (1984) and Mohr (1986). This was not so in the case of Buge *et al.* (1979) who, as our patients, had lesions of the middle third of the posterior limb of the internal capsule and medial globus pallidus. Our infarcts are documented by CT scanning and correspond to the CT localization of the AchA territory of vascular supply made by Damasio (1983), Takahashi *et al.* (1980) and Sterbini *et al.* (1987) (see fig. 3).

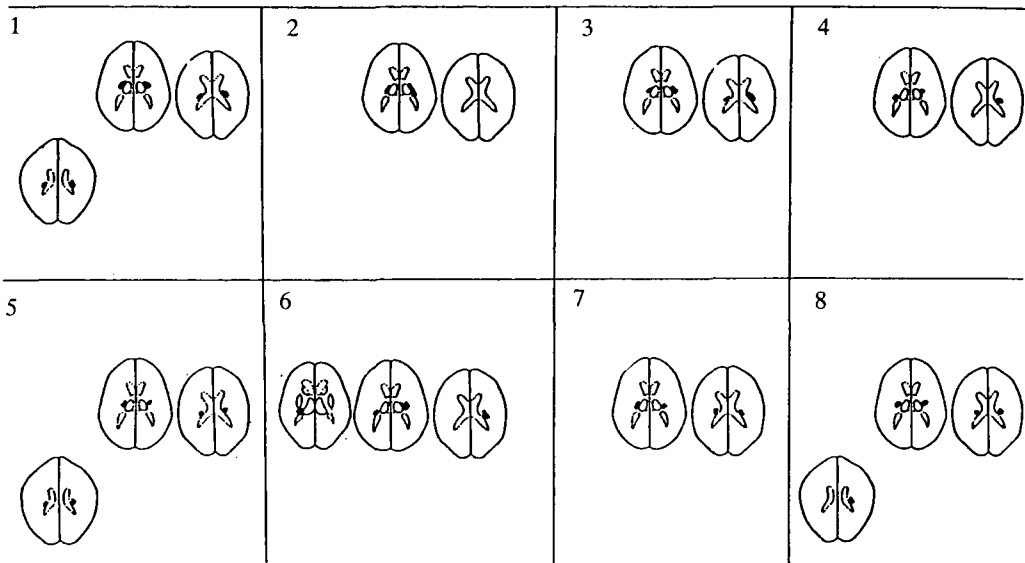


FIG. 3. Diagrammatic representation of location of infarcts in Cases 1-8.

Although it is likely that interruption of the corticobulbar tracts caused pseudobulbar mutism in our patients, it is possible that other structures crossing the posterior limb of the internal capsule were affected (Marie and Guillain, 1902). Pallidofugal fibres originating in the medial globus pallidus and travelling through and near the posterior limb to end in the ventrolateral and ventroanterior thalamus include the ansa and fasciculus lenticularis. Bilateral thalamotomy has been complicated by dysphonia, dysarthria and dysphagia following selective lesions of the Vim, Vce, Vci and Cem nuclei (Andrew *et al.*, 1983). Word blocking has occurred after selective bilateral destruction of the ventrolateral thalamic nuclei (Matsumoto *et al.*, 1984).

Although no definite necropsy or arteriographic proof of AchA occlusion exists, we believe that clinically our cases represent the first reported series of the syndrome of bilateral anterior choroidal artery territory infarction. They present features of the few previously reported single cases of bilateral AchA infarction as well as a

combination of elements of the unilateral AchA syndrome. Abbie (1933), Buge *et al.* (1979) and Fisher (1979) described the only reported cases of bilateral infarction in the AchA territory. These cases, when considered with ours, delineate a spectrum of clinical signs of bilateral AchA territory infarction. Abbie's case had no pseudobulbar symptoms or mutism as were present in the cases of Buge *et al.* and Fisher, but had instead bilateral superior quadrantanopia of acute onset. Abbie's patient had the complete triad of AchA territory infarction, namely a homonymous visual field defect, in this case a superior quadrantanopia, hemiparesis and hemisensory loss on the right, together with a superior quadrantanopia on the left. Our cases resemble those of Buge *et al.* and Fisher in the presence of pseudobulbar mutism, but lack the visual field abnormalities of Abbie's patient. Case 6 had a left homonymous hemianopia which could have been due to infarction of the geniculocalcarine tract at its origin in the AchA territory on the right. It was considered, however, that it had occurred after removal of his pituitary adenoma several years before. Visual field testing was attempted only in 2 other of our patients (Cases 1, 2), but without success because of their inability to communicate. Deficits of this nature may, therefore, have been overlooked. All our cases, as did those of Abbie, Buge *et al.* and Fisher, had hemiparetic and hemisensory symptoms explained by interruption of the corticospinal and sensory radiations in the posterior limb of the internal capsule. Abbie's patient, who was a young woman with syphilis, 2 months before the sudden onset of bilateral homonymous superior quadrantanopia, had developed a hemiparesis, and a hemisensory deficit for all modalities with hyperreflexia and an extensor of plantar response on the right. Fisher's case did not have hemisensory signs or visual field defects, nor did the case reported by Buge *et al.* Abbie's case at necropsy showed necrosis of both lateral geniculate bodies with degeneration of the cerebral peduncles. The postmortem description of the brain at the time of Abbie's report was incomplete and information on capsular or globus pallidus pathology is lacking.

Mirror AchA infarctions are acquired sequentially. The time interval between the appearance of bilateral lesions in reported cases has been short, being 2–3 months in Fisher's and Abbie's patients, although it was undefined in the case of Buge *et al.* The bilaterality in our patients was acquired in time ranges of hours (Case 6), days to weeks (Case 1), months to under 10 yrs (Cases 2, 3, 4, 7, 8) and undefined in Case 5. The rapid acquisition of bilateral capsular lesions may reflect an advanced stage of arterial wall pathology in the AchAs, or the presence of a source for repeated cardiac emboli or untreated syphilis (Case 8).

The time from onset of symptoms to maximal neurological deficit varied. One of our cases had a true capsular warning syndrome (Case 1). This has been noted before in a case of AchA territory infarction (Donnan and Bladin, 1987). In 3 cases, there was a waxing and waning of symptoms for several days before medical attention was sought (Cases 4, 7, 8). Other patients were mute within minutes to hours of the onset (Cases 1, 2, 6), while some became increasingly pseudobulbar in a stepwise manner and then mute within days of the onset of bulbar dysfunction

(Cases 3, 5). The patients of Abbie, Fisher and Buge *et al.* had deficits which were sudden and maximal within minutes of onset.

The outcome in all of our cases was poor. Three out of the 8 patients were dead within 3 months (Cases 3, 5, 8) and 1 (Case 2) within a year of the onset of pseudobulbar mutism. Of those who remain alive, all are severely impaired. Death was precipitated in at least 1 case by a respiratory arrest (Case 3) and aspiration pneumonia in another (Case 2). Details of the other 2 deaths are not available. Two patients (Cases 2, 8) had evidence of myocardial ischaemia and all had the potential to aspirate.

The pathogenesis of ischaemic stroke in our cases is similar to that offered by Abbie, Fisher and Buge *et al.* Hypertensive small vessel disease must have been present in our patients as all had a history of elevated blood pressure. In most cases this was untreated. The patients of Fisher and Buge *et al.* were hypertensive, and the case reported by Buge *et al.* had diabetes, as did half of our cases. Abbie's patient had syphilis and at postmortem a syphilitic endarteritis of both anterior choroidal arteries was found. One of our cases (Case 8) had syphilis but refused lumbar puncture, thus making a diagnosis of meningovascular syphilis unverifiable. The case of Buge *et al.* had a patent foramen ovale. Five of our patients had evidence of cardiac disease. Case 5 had chronic atrial fibrillation and the others ischaemic myocardial disease. Fisher found atheroma of the AchA in his patient at postmortem. Of our cases at least one patient (Case 3) had angiographically documented intracranial atheroma of the internal carotid artery proximal to the AchA. The patient of Buge *et al.* and our Cases 2 and 6, had bilateral extracranial internal carotid artery atheroma. Where deaths occurred in our series, autopsy was denied, so that detailed pathological confirmation of vascular disease was not possible.

All our patients had elevated blood pressure on admission (Table 2). A sudden rapid rise in blood pressure was associated with the onset and worsening of stroke in at least 3 of our patients. Recent evidence (Dutka *et al.*, 1987) supports the concept of limitation of reperfusion after ischaemia because of the potentiation of endothelial damage through the interaction between cellular elements in the blood and the coagulation and complement systems during transient elevation of blood pressure in the immediate poststroke period. One patient, however, worsened as her blood pressure fell precipitously on a background of chronic hypertension and probable altered cerebral autoregulation.

Certain clinical features shown by our patients bear mention. The majority had a period of lethargy with or without Cheyne-Stokes respiration. Bilateral occlusion of the AchA in surgery for parkinsonism has been followed by lethargy (Cooper, 1956). Four patients had uncontrollable fits of laughter or crying devoid of emotional content. Others had marked depression or blunted affect. Hypothalamic and pallidal efferent fibres mediating emotional impulses to the bulbar cranial nerve nuclei do not travel in the internal capsule and are spared in our patients (Brodal, 1981). The role played by caudate destruction with regard to changes in affect has

yet to be defined. Abnormality of ocular movement with head and eye gaze preference has been reported in unilateral AchA stroke (Helgason *et al.*, 1986) and in several of our present cases. It is reminiscent of deep putaminal infarcts or haemorrhages and large hemisphere strokes. Head and eye gaze preference in our patients appeared to be related to neglect. This and other dysfunctions of visuospatial ability or language are well recognized in unilateral AchA syndromes (Cambier *et al.*, 1983; Decroix *et al.*, 1986; Helgason *et al.*, 1986). Urinary and bladder incontinence was noted in almost all our cases, but no patient was severely demented. Alajouanine and Thurel (1933*b*) have discussed the independence of emotional and urinary incontinence due to bilateral upper motor neuron interruption from dementia.

Conclusions

In this paper, we have presented 8 instances of pseudobulbar mutism of abrupt onset due to discrete lacunar or slightly larger infarcts localized by CT and MRI to the posterior limb of the internal capsule, the medial globus pallidus and the adjacent caudate nucleus and white matter in the area of supply of the AchA. We do not have proof that the AchA was occluded. Our basis for implicating its involvement rests on our CT-MRI definition of vascular supply (Beevor, 1907; Damasio, 1983, 1987, personal communication). Although there was no necropsy confirmation of the pathogenesis of the strokes in our patients, the possibility exists that emboli of cardiac as well as atheromatous arterial origin, and small vessel disease related to hypertension, diabetes or intracranial atheroma, were present in our cases. Poorly controlled hypertension and acute transient elevation of blood pressure were common to all our cases. Both therefore appear to play an intimate role in the pathogenesis of stroke in our patients.

Bilateral AchA territory infarction reflects an advanced stage of cerebrovascular disease and carries a poor prognosis. It can be recognized by clinical signs and confirmed by CT scanning and MRI. Although acute pseudobulbar symptoms with various degrees of mutism have been described in unilateral and bilateral cortico-opercular (Tournier, 1898; Foix *et al.*, 1926; Alajouanine and Thurel, 1933*a*; Alajouanine *et al.*, 1959; Boudin *et al.*, 1960), lenticular (Marie, 1906) or brainstem (Karp and Hurtig, 1974) location, it is the presence of additional elements of the unilateral AchA syndrome and the lack of definite brainstem signs, in combination with a previous history of stroke localizing to the internal capsule, which characterize the nature of the lesion. Assiduous investigation and recognition of underlying vascular or cardiac disease and treatment at the stage of unilateral capsular infarction may prevent the occurrence of contralateral capsular ischaemia. Careful control of blood pressure in the acute stages of ischaemia may prevent postischaemic deterioration which often leaves the patient virtually in a locked-in and moribund state.

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