Progressive aphasia with right-sided extrapyramidal signs: another manifestation of localised cerebral atrophy

Sir: Slowly progressive aphasia without generalised dementia has been recognised as a degenerative condition with non-Alzheimer pathology and localised histological changes have been demonstrated in the dominant perisylvian regions. We report a unique case in which progressive aphasia is combined with right-sided tremor and rigidity. Computed tomography and single photon emission tomography indicated a striking, predominantly left hemisphere disorder.

A right handed 64 year old man presented with an 18 month history of insidiously progressive deterioration in his language together with tremor of the right hand. He had been previously well and there was no history of vascular disease or head injury. His mother, who died aged 83, was said to have had Parkinson’s disease and became “confused and wandering” late in life. General physical examination was normal. Visual fields were intact but eye movements were abnormal with reduced voluntary upgaze and impaired convergence. He had an expressionless face and slight right-sided facial asymmetry. Tone was minimally increased on the right and there was a resting tremor of the right hand. Reflexes were symmetrically brisk and plantar responses flexor. Sensory testing, dexterity and gait were normal.

Neuropsychological assessment revealed a selective language disorder. His spontaneous speech was non-fluent, stuttering and effortful with word finding difficulty and the occasional intrusion of literal and verbal paraphasias. Comprehension and repetition were mildly impaired. He could read aloud and understand single words, but misread complete phrases and sentences. His writing, although conveying the sense adequately, contained spelling errors and was telegraphic in style. In contrast to his impaired linguistic ability, praxis, visual perception, spatial localisation and navigational skills and non-verbal memory were preserved. His personality was unchanged and his social conduct was appropriate.

Computed tomography revealed widening of the left sylvian fissure and prominence of the left lateral ventricle. Otherwise, routine laboratory and neurological investigations including electroencephalography were normal. Carotid angiography demonstrated a tonsillar loop of the right internal carotid artery but was otherwise normal.

Two years later he had deteriorated both physically and in his powers of communication. Neurological examination revealed emotional lability, increase in tremor of his right hand, moderate right-sided hypertonia and bradykinesia. He dragged his right leg when walking. His spontaneous speech was limited to “yes”, “no” and infrequent stereotyped phrases. Occasional neologisms occurred. Comprehension had deteriorated although he could follow elementary commands. He was severely anomic. He could no longer sign his name. He had difficulty communicating by gesture or pantomime. Perceptual and spatial abilities remained well preserved. Memory could not be formally tested, although his wife believed that he could recall day to day events.

Single photon emission tomography using the tracer Tc-HMPAO demonstrated a striking reduction in uptake in the left frontal, temporal and parietal regions and in the left subcortex, whilst right hemisphere uptake was normal (fig (a)).

One year later a mild right-sided pyramidal weakness had developed in addition to the extrapyramidal signs. Tendon reflexes on the right were brisk and the right plantar response equivocal. He had a right-sided grasp reflex, with pout and suckling reflexes. Speech was limited to an arbitary production of “yes” and “no”, and comprised hesitation to following midline commands. Perception and spatial functions again appeared preserved in that he negotiated his environment without becoming lost, and recognised objects, oriented and used them appropriately. Performance was normal on a forced choice object recognition test suggesting some preservation of visual memory. His personality remained unchanged and he continued to demonstrate initiative in daily activities. A repeated electroencephalogram remained normal whilst computed tomography demonstrated increased atrophy and ventricular dilatation on the left, with the addition of mild right-sided involutions and changes (fig (b)).

There are strong grounds for assuming that this patient has a degenerative condition due to localised cerebral atrophy. There has been a slow, progressive development of neurological signs in the complete absence of stroke-like episodes and without risk factors for vascular disease. Cerebral angiography has excluded large vessel disease and the possibility that emboli from the heart or a small vessel disease would produce such a striking, predominant involvement of the left cerebral hemisphere is unlikely. Computed tomography showed no low density lesions to suggest cerebral infarction. The electroencephalogram was normal whereas in vascular dementia focal slowing is usual.
It is unlikely that this patient suffers from Alzheimer’s disease. The presence of progressive and selective language disorder in the absence of other cognitive deficits is distinct from the characteristic pattern of Alzheimer dementia, in which linguistic impairment is combined with profound anemia and visuo-spatial impairment. Moreover, the effortful, paraplastic speech is dissimilar to the fluent speech normally associated with Alzheimer’s disease. The normal electroencephalogram and strikingly asymmetric computed tomogram contrast with the significant slowing of wave forms on electroencephalography and either normal or generalised atrophic scan appearances reported in Alzheimer’s disease. Reduced uptake in the temporoparietal regions in Alzheimer’s disease has consistently been demonstrated by positron emission tomography and single photon emission tomatography. Whilst the left hemisphere may be predominantly affected in patients with disproportionate language disorder, abnormalities are not exclusively unilateral. In the present case, abnormalities appeared to involve virtually the entire left hemisphere, with the exception of the occipital areas, whilst right hemisphere uptake was normal.

This patient closely resembles cases of slowly progressive aphasia without generalised dementia, one of whom also developed a right hemiparesis. Left temporal and parietal hypometabolism found by positron emission tomography is consistent with the left hemisphere abnormality demonstrated in the present case. Although an association has previously been reported between progressive aphasia and extrapyramidal features, unlike the present case, this was in the context of more widespread cognitive changes, indicating bilateral affection. Occasionally extrapyramidal disorder has occurred in combination with fronto-temporal atrophy, but most commonly with additional clinical features of motor neuron disease.

Common to forms of localised cerebral atrophy are the pathological findings of cell loss, gliosis and spongy change, and a minority of these cases have additional neuronal swellings and argyrophilic inclusions. There appears to be a spectrum of topographical distribution of pathological change. In most cases both frontal and temporal lobes are involved but purely frontal, purely temporal and predominantly subcortical presentations have been described. In other cases with clinical features of motor neuron disease there has been additional involvement of the corticospinal tracts and anterior horn cells. Cases of progressive aphasia without dementia have demonstrated pathological change in the dominant perisylvian region. The case described in this letter is an example of a predominantly left hemisphere degeneration presumably involving primarily the perisylvian regions and subcortex.

It is suggested that the clinical picture in our patient of progressive aphasia and progressive right-sided extrapyramidal signs represents one of a spectrum of possible presentations of localised cerebral atrophy all of which may share a similar pathological basis.

**References**

Severe aggravation of blepharospasm in Fisher's syndrome

Sir: Essential blepharospasm has been considered a cranial dystonia caused by a biochemical imbalance of the extrapyramidal system with no relevant peripheral nervous system contribution. 

The syndrome is characterised by external ophthalmoplegia, ataxia and areflexia and represents a limited form of acute idiopathic polyneuropathy (Guillain-Barré) syndrome. Both processes coincided in a 67 year old male. At 20 years of age he began to suffer occasional involuntary lid closure, more prominent in the right eye, but without disability. These spasms had increased slightly in recent years, often triggered by bright light. After an episode of acute febrile rhinopharyngitis 15 days earlier, over a week he developed severe progressive ataxia, complete external and internal ophthalmoplegia, the eyes remaining in neutral position, ptotic but without diplopia, and general areflexia. Consciousness was normal. CSF showed 0 cells, glucose 0-68 g/l and protein 0-63 g/l. In the next few days, transitory breathing and swallowing difficulties developed, as well as mild weakness of the facial musculature. In this situation of complete ophthal paralysis, the patient made constant gesticulation due to frequent, occasionally sustained, bilateral blepharospasm attacks. This picture regressed to the previous situation after the ophthalmoplegia resolved some months later.

The severe aggravation of facial spasms in our case was striking, well in excess of what could have been expected from the emotional stress of hospitalisation or appearance of new symptoms. A coincidental relation to an improbable midbrain lesion is purely speculative. A lesion located in the midbrain tegmentum was discovered in only one case of Fisher's syndrome and had not been confirmed in other necropsy cases. On the other hand, in only one case of blepharospasm was a well-localised upper brain stem lesion found. In our patient, the futility of efforts to counter ocular paralysis and palpebral weakness (m. elevator palpebrarum) may have accentuated the actions of antagonist muscles (m. orbicularis oculi). The excess of frustrated central excitation and lack of reciprocal inhibition is considered the EMG pattern of dystonia and could explain our case.

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References

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