

Progressive dysphasic dementia with bucco-facial apraxia: a case report

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A patient with progressive dementia, prominent non-fluent aphasia and signs of frontal lobe involvement, was evaluated by neuropsychological testing, magnetic resonance imaging (MRI) and high resolution single photon emission tomography (SPET). The presence of severe bucco-facial apraxia, associated with spared imitation of limb movements, correlated well with a marked reduction of cerebral perfusion in the left fronto-temporal cortex. This case emphasizes the usefulness of SPET as a valuable alternative to PET for the diagnosis of conditions, such as progressive neuropsychological syndromes, where a coupled reduction of metabolism and blood flow can be expected.

Keywords: Apraxia – Dementia – Single photon emission tomography

INTRODUCTION

We report the case of a patient with a progressive dementia, characterized by severe deterioration of language and neuropsychological signs of frontal lobe involvement, who had a severe bucco-facial apraxia and a flawless performance on tests of limb praxis. The clinical picture suggested relatively selective pathological involvement in the fronto-temporal areas of the left hemisphere. Regional cerebral perfusion was assessed with high resolution single photon emission tomography (SPET). The imaging results were in agreement with the clinical findings, and comparable to the positron emission tomography (PET) findings reported by Tyrrell *et al.* (1991) in three patients with a similar clinical presentation.

CASE REPORT

The patient was a 46-year-old right-handed male factory worker, with an unremarkable past medical history. There was no family history of dementia or Down's syndrome. Two years before admission he had begun to have some difficulties in understanding speech, initially attributed to loss of hearing; after some months the patient's speech started to deteriorate, becoming slurred, effortful and monotonous, with frequent pauses for word searching. A parallel impairment of written language was also evident, and his handwriting had become indecipherable and there

was a progressive reading impairment. Moreover, he was no longer able to read music (he was an amateur harmonium player). The only abnormality in the patient's behaviour was the cheerful lack of concern for his language difficulties. He had to leave work, but could still go out alone and take part in social activities.

The general neurological examination was unremarkable: in particular, muscle tone was normal and no involuntary movements were observed; frontal "release" signs were not evident.

Neuropsychological examination

The patient had a severe impairment of language. During the examination he was pleasant, cooperative and showed an appropriate communicative intent. His spontaneous speech was limited to single content words, usually in his regional dialect, with a prevalence of "multipurpose" morphemes: the production was effortful, with phonetic distortions. Visual naming was studied with a 90-items test, including 30 animals, 30 fruit and vegetables and 30 man-made objects. The performance was overall very poor, and was characterized by the perseverative production of a prototypical specimen of the appropriate category (as "apple" for fruit, "dog" for animals). Single-word (picture-word matching with three distractors) and oral command comprehension was severely impaired. Repetition was good, with some articulatory defects, for

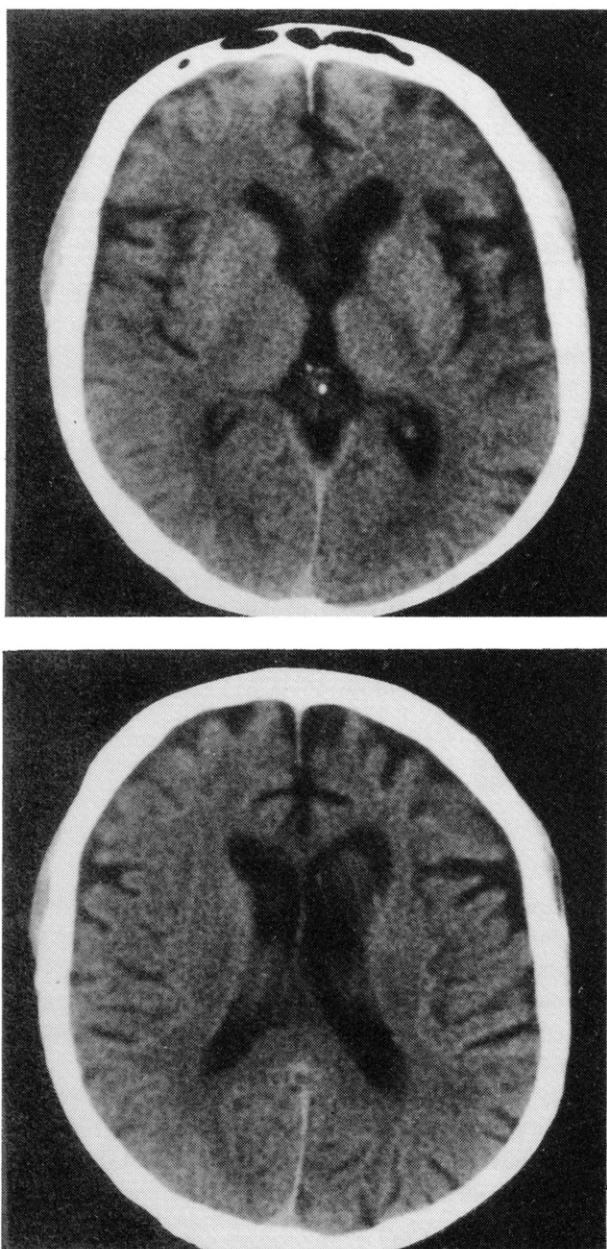


FIG. 1. CT scans: cortical atrophy and ventricular dilatation, more marked on the left side.

words, grammatical morphemes and non-words. Reading aloud and reading comprehension were severely impaired; writing was characterized by sparse neologistic productions.

Testing of apraxia with the imitation of 12 oro-facial movements (De Renzi *et al.*, 1967) evoked eight errors (with inappropriate vocalizations, incomplete movement and perseverations). On the other hand, his performance on an extensive test of imitative limb praxis was excellent. The test includes both meaningful and meaningless arm gestures ($n = 17$), single and repetitive hand movements ($n = 11$ for each hand) and object utilization ($n = 18$). A quan-

titative scoring system, as well as a detailed qualitative evaluation of error types is available. The patient's score was 122, well above the cut-off value of 100 (from 20 normal controls). The severity of the language disorder prevented the evaluation of verbal memory. Corsi span was 2. The copy of Rey's figure was severely distorted; recall after 10 min was nil. The patient failed to score on Raven's coloured PM, due to lack of comprehension of test requirements: he pointed randomly to the response choices, with perseverations.

Ancillary examinations

The EEG was normal. CT and MRI showed diffuse cortical and ventricular atrophy, with a bilateral fronto-temporal involvement, more prominent on the left (Figs 1 and 2). Perfusion imaging was obtained using a brain-dedicated single photon emission tomography (SPET) system (ASPECT, Digital Scintigraphic Incorporation) (Genna and Smith, 1988), with a spatial resolution of approximately 8.3 mm FWHM in plane, after intravenous injection of approximately 280 MBq (22 mCi) of (^{99m}Tc)HM-PAO. In 30 min acquisition time 6 million total counts were collected. Cross-sectional images were reconstructed and corrected for attenuation. Image analysis was performed both by visual inspection and by semi-quantitative analysis: circular regions of interest (ROIs) with a diameter equal to 1.5 FWHM of the system (11 mm) were positioned on the cortical ribbon, making reference to a published atlas (Damasio and Damasio, 1990). Ratios between the mean counts of each ROI and the mean count value of all supratentorial ROIs of both hemispheres were computed (Table I). A marked relative reduction of perfusion (40% decrease from mean value) was found in medial frontal areas bilaterally, in the left lateral and orbital frontal lobe and in the left temporal pole. Perfusion was relatively spared in sensorimotor, temporo-

TABLE I. Perfusion ratios (regional count value/mean supratentorial bihemispheric count value)

Region	R	L
Frontal lateral	0.97	0.70
Frontal orbital	0.89	0.74
Frontal rolandic	1.02	0.85
Frontal mesial	0.73	0.56
Temporal pole	0.91	0.60
Temporal superior	1.10	0.91
Temporal middle	1.13	1.01
Temporal inferior	1.08	0.93
Parietal	1.21	1.19
Occipital lateral	1.12	1.00
Occipital calcarine	1.35	1.33
Thalamus	1.14	1.13
Basal ganglia	1.39	1.28

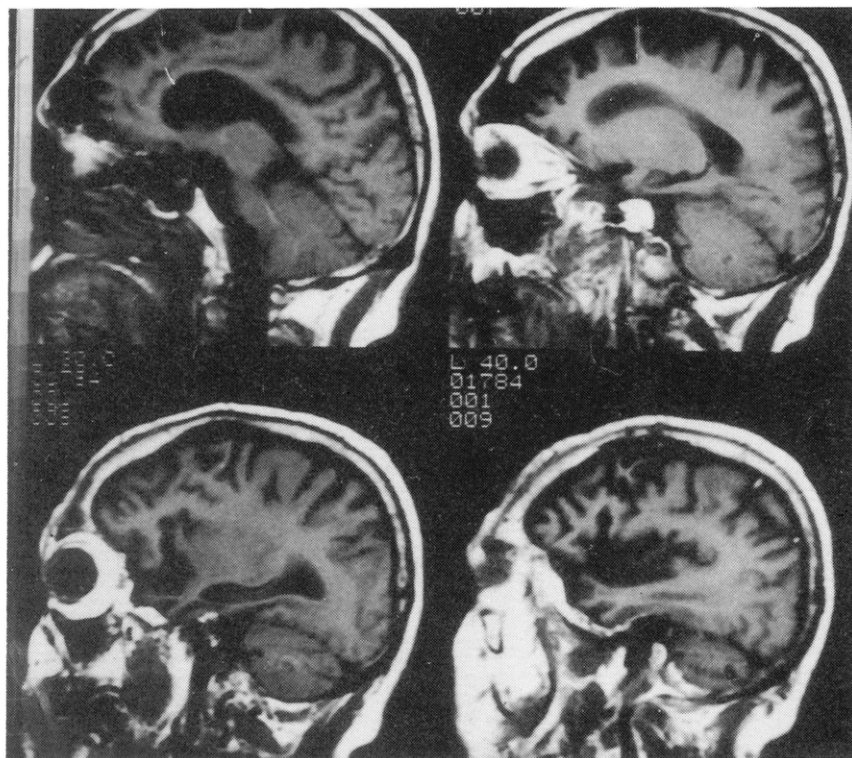


FIG. 2. MRI: sagittal cuts, confirming the severe atrophy.

parietal and occipital areas and in subcortical nuclei. There was a mild crossed (right) cerebellar diaschisis (Fig. 3).

Over the ensuing months after the present investigation, the patient deteriorated with the onset of severe behavioural disturbances, characterized by agitation and violent outbursts, requiring psychiatric treatment and finally hospital admission. Total lack of cooperation prevented further neuropsychological reassessment.

DISCUSSION

This patient presented with a progressive dementia, characterized by severe language impairment (non-fluent aphasia) and by a bucco-facial apraxia; on the other hand, imitation of limb movements was excellent. At the moment of our evaluation there was neuropsychological evidence of memory and visuo-spatial impairment, although the patient was still able to go out alone and take part in social activities. Perseveration, which was prominent in most aspects of behaviour, was not observed during the testing of imitation of limb gestures. SPET indicated a severe left fronto-temporal hypoperfusion: the functional findings were in agreement with the prevalent left fronto-temporal atrophy shown by MRI.

These findings are in close correspondence with the clinico-anatomical correlations observed in patients with

focal cerebral lesions. Apraxia, the acquired inability to reproduce movements and gestures on verbal command or imitation, can involve limb and/or bucco-facial movements: a double dissociation between oral and limb praxis is found in patients with localized left hemisphere lesions (De Renzi *et al.*, 1967). Differential lesion location can account for this finding: bucco-facial apraxia is most commonly found after damage to the frontal and central operculum and the anterior insula of the left hemisphere (Tognola and Vignolo, 1980), while limb apraxia is particularly frequent and severe after parietal lesions (Faglioni and Basso, 1985). While ideomotor apraxia is frequently present in Alzheimer's disease, especially in the intermediate and advanced stages, limb transitive movements appear to be more vulnerable than bucco-facial gestures (Della Sala *et al.*, 1987), suggesting that apraxia in Alzheimer's disease is probably the result of temporo-parietal involvement by the pathological process.

Tyrrell *et al.*, (1991) have recently described three patients with a similar clinical picture, characterized by progressive speech impairment associated with bucco-facial dyspraxia, who subsequently progressed to a more widespread cognitive impairment. A study of cerebral oxygen metabolism with PET in these patients revealed a marked bifrontal reduction.

The underlying histopathology in the present case is a

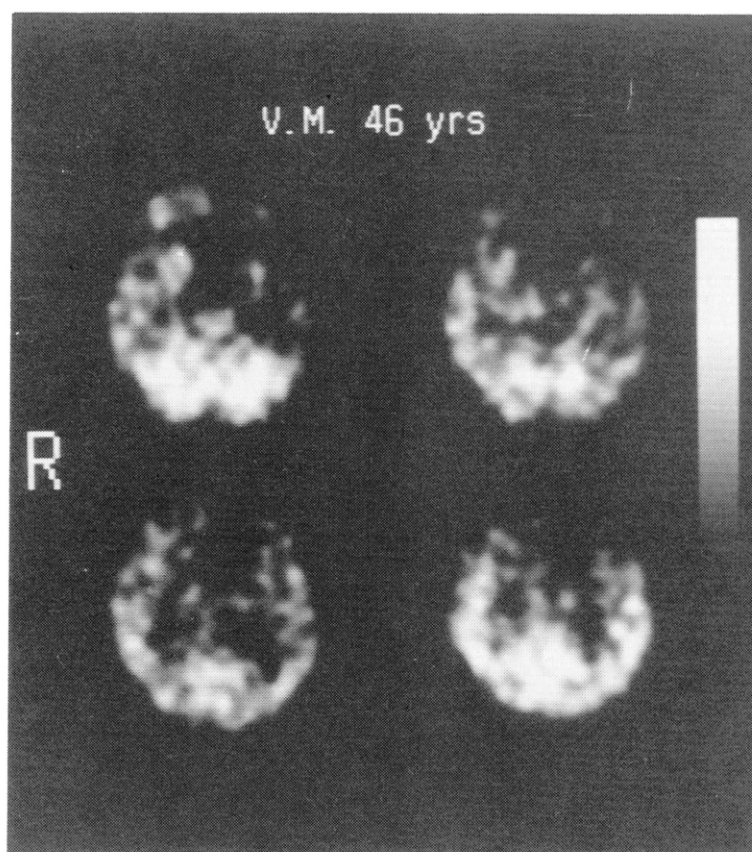


FIG. 3. SPET axial images: severe reduction of tracer uptake in the fronto-temporal regions, in particular on the left side.

matter of speculation. Several different degenerative processes, such as Alzheimer's disease, Pick's disease, Creutzfeldt-Jacob disease and cortical basal degeneration, have been associated with a syndrome of slowly progressive aphasia (Wechsler *et al.*, 1982; Pogcar and Williams, 1984; Mandell *et al.*, 1989; Lippa *et al.*, 1991). Progressive speech impairment with orofacial apraxia has been reported in the early clinical course of a case of cortical basal degeneration (Lang, 1992). On the other hand, Pick's disease was the most frequent association of this syndrome in Tyrrell's series (Rossor and Tyrrell, 1992). Prominent involvement of the frontal lobes, both on neuropathological and neuroimaging grounds, is typical of Pick's disease (Graff-Radford *et al.*, 1990) and of frontal lobe dementia (Gustafson, 1987; Miller *et al.*, 1991). In our patient, the presence of fronto-temporal involvement on SPET and the rapidly progressive clinical picture, with severe behavioural disorders, are compatible with a diagnosis of Pick's disease.

Functional imaging methods are the most suitable tool to identify the patterns of cerebral involvement *in vivo* and to follow their evolution in time. Several reports have established the role of positron emission tomography in

progressive neuropsychological syndromes (Tyrrell *et al.*, 1990, 1991). The present case, together with other recent studies (Miller *et al.*, 1991; McDaniel *et al.*, 1991), indicates that the assessment of regional perfusion with single photon emission tomography can be a useful diagnostic alternative in these conditions, where the reduction of metabolism is coupled to decreased blood flow. The lack of absolute quantification does not prevent the identification of diagnostic patterns of cerebral involvement, if the spatial resolution of the SPET system is adequate for the size of the cerebral structures under investigation.

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