Cerebral blood flow SPECT scanning in cortico-basal degeneration

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Abstract

BACKGROUND: Idiopathic Parkinson’s disease accounts for ca. 75% of all cases of Parkinsonism. Corticobasal degeneration is a relatively rare example of the so-called “Parkinson-plus” syndrome.

METHODS AND RESULTS: The authors present the case of a 56-year-old woman with rigidity and atypical tremor of upper extremity followed by gait apraxia, dysarthria, bilateral pyramidal signs and myoclonus. There was no improvement after treatment with L-dopa. The disease has progressed, but the patient is still alive. On the basis of clinical data a diagnosis of corticobasal degeneration has been established. Cerebral blood flow SPECT scanning revealed diffuse hypoperfusion of left frontal lobe, antero-inferior part of the left temporal lobe and left basal ganglia. The case illustrates the usefulness of brain SPECT in atypical forms of Parkinson’s disease.

Introduction

75% of all patients with signs of Parkinsonism are patients with idiopathic Parkinson’s disease (1). The remaining cases are “secondary” Parkinson’s syndromes: drug-related, toxic, vascular and so-called “Parkinson-plus” syndromes. “Parkinson-plus” syndromes are characterised by the co-existence of signs of Parkinson’s disease with other neurological signs, no or minimal reaction to the treatment (particularly levodopa) and rapid progressing. “Parkinson-plus” syndromes comprise progressive supranuclear palsy, multiple system atrophy, Alzheimer’s disease with Parkinsonism, amyotrophic lateral sclerosis with Parkinsonism, corticobasal degeneration (CBD), Huntington’s and Wilson’s diseases (2).

Cortico-basal degeneration was described for the first time in 1968 by Rebeiz et al. (3). So far about 70 cases of this disease have been described. It is a progressive disease comprising characteristic features concerning both cortical and basal ganglionic dysfunctions. Manifestations include akinetic-rigid syndrome and apraxia strictly limited at disease onset to one hemibody. Alien limb phenomenon, rigidity, loss of dexterity can mark the beginning of the disease (4). We present a case of a woman with a diagnosis of cortico-basal degeneration, where cerebral blood flow SPECT scanning played a major role in the diagnostic process.

Case description

In 1992 a 51-year-old woman presented for the first time right hand tremor, followed after a few months by lower limbs gait and speech disorders, propulsion, and rigidity. During the next hospital stay apraxia and dysarthria progressed. In 1997 the patient showed total gait apraxia, amimia, anarthria, strong rigidity, bilateral Babinski sign and stimulus sensitive myoclonus. Serum copper, ceruloplasmin and thyroid hormone levels were normal. CT scanning showed moderate parietal cortex atrophy, while MRI PD and T2 scanning showed putamen hyperintensities as well as minor left parietal atrophy. Wechsler’s scale was normal, dysautonomy testing was negative. EEG was unspecific. Treatment with amantadine, levodopa, clonazepam and akineton gave no effect.

Cerebral blood flow SPECT scanning was performed using three-head gammacamera Multispect-3 (Siemens, Erlangen, Germany) 1 hr post i.v. injection of 740 MBq of ⁹⁹mTc-HMPAO (Amsaldo, United Kingdom) using a low-energy, ultra-high resolution collimator.

The data were collected into a 128 x 128 matrix, 4.8 mm per pixel. The raw data were smoothed with a Butterworth filter, cut-off frequency 0.35. Chang attenuation correction was not performed. The images were reoriented in the axial, coronal and sagittal planes.

The data were displayed on a 10-grade colour scale. Focal perfusion abnormalities were read twice by two independent observers. Their depth was assessed utilising an asymmetry index (AI): AI = R – L/ (R + L)/2 x 100 % according to De Cróitofaro (5), where R and L are mean counts/pixel values in the right and left hemisphere, respectively.

Cerebral blood flow SPECT scanning showed a diffuse perfusion deficit of lower part of left frontal lobe (AI=11%), fronto-lower part of the left temporal lobe (AI=16%) and left basal ganglia (AI=16%) (Figure 1).
Discussion

The clinical picture shown above fulfills the criteria of cortico-basal degeneration according to Watts (6). Also the resistance to medication, particularly levodopa, makes this diagnosis the most probable. No exclusion CBG criteria were seen.

The case underlines the usefulness of CBF SPECT scanning in CBD. CT and MRI scanning, although helpful in eliminating the other CNS pathology, are not specific and asymmetrical atrophy may evolve in an advanced stage of the disease. SPECT scanning shows the characteristic blood flow changes: asymmetric, contralateral to clinical signs CBF cortico-subcortical deficit.

The authors are aware of two case reports on CBF deficit in cortico-basal deficit (7, 8). Both reports state contralateral hemisphere blood flow deficits in fronto-parietal cortex (perirolandic area), as in our report. Cortical hypoperfusion in the perirolandic area corresponds to limb-kinetic apraxia, in posterior parietal area to constructional apraxia. This may be secondary to regional cerebral hypometabolism (4).

This case illustrates the difficulties in the differentiation of Parkinson’s syndromes, especially in the initial stage of disease. SPECT cerebral blood flow scanning could be highly helpful in establishing the correct diagnosis.

References


Figure 1. Brain SPECT — thalamic slice.