

B. Okuda
H. Tachibana
M. Takeda
K. Kawabata
M. Sugita
M. Fukuchi

Focal cortical hypoperfusion in corticobasal degeneration demonstrated by three-dimensional surface display with ^{123}I -IMP: a possible cause of apraxia

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B. Okuda (✉) · H. Tachibana · M. Takeda · K. Kawabata · M. Sugita
Fifth Department of Internal Medicine,
Hyogo College of Medicine, 1-1,
Mukogawacho, Nishinomiya 663, Japan

M. Fukuchi
Department of Nuclear Medicine, Hyogo
College of Medicine, Nishinomiya, Japan

Introduction

Corticobasal degeneration (CBD) is characterised by asymmetrical higher cortical dysfunction and extrapyramidal signs, and the pathological changes are usually asymmetrical [1, 2]. Since a report from Massachusetts General Hospital [3], cases with clinically or pathologically diagnosed CBD have increased in number [1, 2, 4]. Although patients with CBD frequently exhibit limb-kinetic (LKA) and constructional apraxia (CA) [1, 2, 4], the lesions responsible have not been clarified. We report two patients with clinical CBD, in whom three-dimensional (3D) surface display generated from single-photon emission computed tomography (SPECT) demonstrated cortical perfusion defects corresponding to the two types of apraxia.

Materials and methods

The patients were diagnosed as having CBD on the basis of their clinical course, neurological signs, and neuroimaging.

Higher cortical dysfunction in these patients were two types of apraxia: LKA and CA. LKA is defined as a breakdown of previously skillful movements, and is manifest as difficulty with fine

Abstract To clarify cortical lesions responsible for apraxia in corticobasal degeneration (CBD), we reconstructed three-dimensional surface images from single-photon emission computed tomography (SPECT) data with *N*-isopropyl-*p*[I-123]-iodoamphetamine in two patients with CBD. Both had limb-kinetic apraxia (LKA) and one also had constructional apraxia (CA). Both showed asymmetrical cortical hypoperfusion in the perirolandic

area. The patient with CA had unilateral hypoperfusion in the posterior parietal area. Thus, cortical hypoperfusion in the perirolandic area corresponded to LKA, and that in the posterior parietal area to CA.

Key words Corticobasal degeneration · Three-dimensional surface display · Apraxia · Cortical hypoperfusion · Single photon emission computed tomography

finger movements [5]. CA is defined as bimanual difficulty in putting together one-dimensional units so as to form two- or three-dimensional figures or patterns. CA was tested by copying line drawings and stick constructions [6].

To assess cerebral blood flow (CBF), SPECT with *N*-isopropyl-*p*[I-123]-iodoamphetamine (^{123}I -IMP) was conducted 30 min after administration of ^{123}I -IMP. A three-dimensional (3D) surface display was created from the axial SPECT images, using the STARCAM system computer, to show the cortical perfusion defect [7]. The threshold value for the surface boundary was 60 % of the global maximum counts in the SPECT images. We adopted this threshold value, since 11 control subjects without neurological disease (mean age 67.6 ± 10.6 years) rarely showed perfusion defects at 60 % threshold.

Case reports

Case 1

A 71-year-old, left-handed man noticed a progressive movement disorder of the left hand for 2 years. He had neither dementia nor aphasia. He presented with bilateral LKA, especially on the left side, without CA. Strength was preserved but pyramidal signs were positive bilaterally. Muscle tone was increased moderately on the left and mildly on the right. He showed neither cerebellar ataxia nor involuntary movements; eye movements were preserved except for

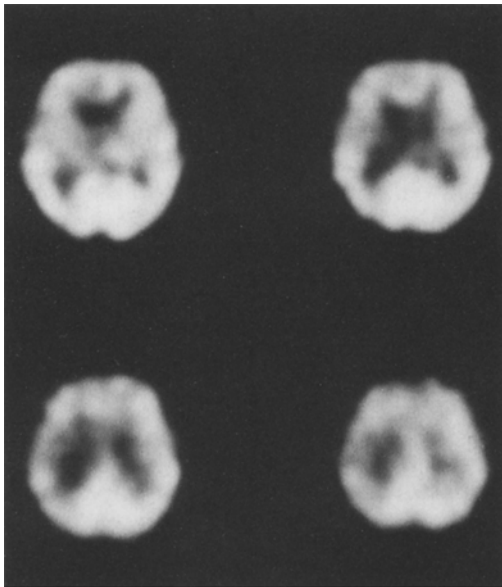


Fig.1 Case 1. SPECT shows hyperperfused areas in the right hemisphere

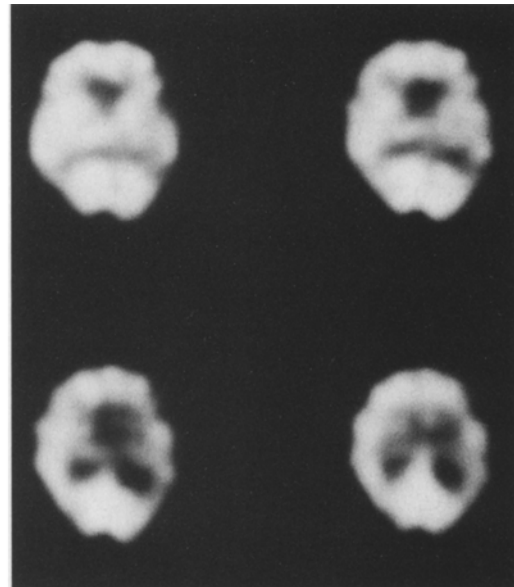


Fig.3 Case 2. SPECT shows hyperperfused areas in the left hemisphere

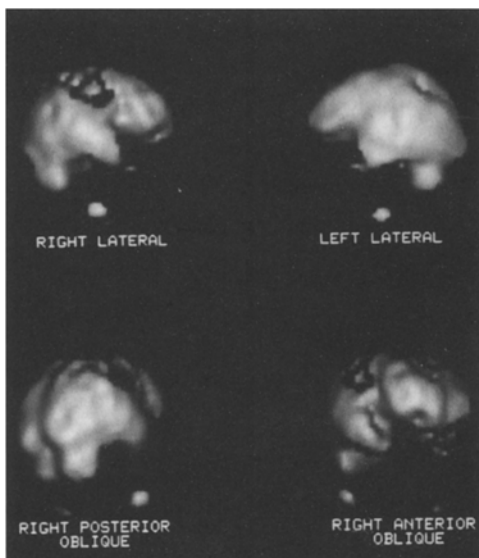


Fig.2 Case 1. 3D surface display shows cortical hypoperfusion in the bilateral perirolandic areas, more marked on the right

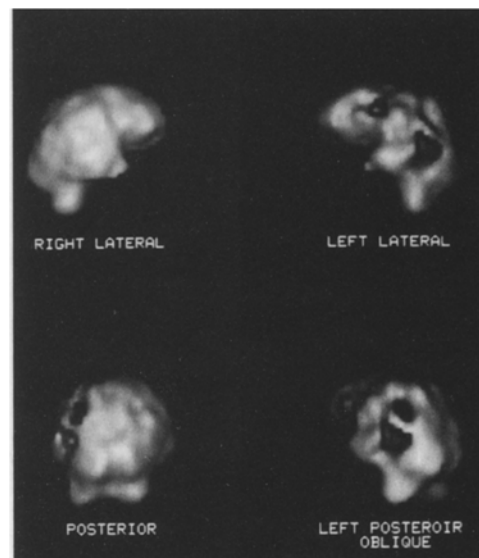


Fig.4 Case 2. 3D surface display shows cortical hypoperfusion in the left perirolandic and posterior parietal areas

impaired convergence. Brain MRI showed no abnormality. SPECT disclosed hypoperfused areas in the right hemisphere, mainly in the frontal lobe and to a lesser extent in the basal ganglia (Fig.1). 3D surface display demonstrated decreased CBF in the perirolandic areas, markedly on the right and mildly on the left (Fig.2).

Case 2

A 65-year-old, right-handed woman suffered from progressive unsteadiness of gait for 3 years. She was moderately demented (Mini-

Mental State Examination score 16/30), but not aphasic. She had marked CA and mild bilateral LKA. Mild bilateral parkinsonism was present. She also showed supranuclear vertical gaze palsy, predominantly on downward gaze. Brain CT showed atrophic changes in the left hemisphere. SPECT disclosed hypoperfused areas in the left frontal and parietal lobes (Fig.3). 3D surface display revealed decreased CBF in the left perirolandic and posterior parietal cortices (Fig.4). Although there was no clinical asymmetry in this case, the distribution of cortical hypoperfusion was consistent with that in previously reported cases [4]. These findings suggested the diagnosis of CBD, but the possibility of as-

sociated progressive supranuclear palsy could not be excluded [1, 8].

Discussion

These cases showed considerably asymmetrical cortical hypoperfusion in the perirolandic or the posterior parietal areas or both. The perirolandic area comprises the precentral and postcentral gyri, and the posterior parietal area the angular gyrus. Perirolandic cortical hypoperfusion can explain contralateral LKA, because LKA has been attributed to a lesion in the sensorimotor cortex [5]. As shown in case 2, a dominant hemisphere lesion can cause bilateral LKA [4]. Other types of limb apraxia such as ideomotor (IMA) or ideational (IA) have been reported [2, 9], but were not found in these patients with CBD. The discrepancy may suggest a certain heterogeneity of CBD, or it may be that motor clumsiness due to LKA can induce apraxic disorders resembling IMA or IA [4, 5]. Since the sensorimotor cortex is predominantly involved in CBD [1, 2], we think that LKA is the most frequent type of limb apraxia in CBD. The lesion responsible for CA is more complicated; CA can occur with bilateral lesions, or right- or left-sided lesions [6], and the intrahemispheric locus of the lesion responsible for CA remains obscure. In general, lesions somewhere in the retrorolandic region tend to be associated with CA [6]. Our patient with CA showed unilateral hypoperfusion in the parietal cortex, particularly in the posterior part, including the angular gyrus. Clinical and experimental studies have

demonstrated that the angular gyrus plays an important role in visuospatial orientation [10, 11]. Thus the posterior parietal cortex, comprising the angular gyrus, appears to be the most likely site for CA in CBD.

Asymmetrical cerebral hypoperfusion or hypometabolism has been described in CBD [4, 12, 13]. Sawle et al. [12] showed that fluorodopa uptake was reduced in the striatum and cortical oxygen metabolism depressed in the superior and posterior temporal, inferior parietal, posterior frontal, and occipital association areas of the cortex. Eidelberg et al. [13] demonstrated that glucose metabolism was decreased in the thalamus, inferior parietal lobule, and hippocampus. In good agreement with neuropathological findings [1–3, 14], these neuroimaging data disclosed the predilection for frontal and parietal cortical involvement in CBD. Our study revealed more demarcated areas of cortical hypoperfusion, in the perirolandic and posterior parietal cortex. The differences in distribution of hypoperfusion or hypometabolism may depend on methodological variations or the duration of the illness.

Although various patterns of regional hypoperfusion or hypometabolism have been reported in CBD, the lesions responsible for higher cortical dysfunction remain to be clarified. 3D surface display appears useful for detecting the sites of lesions responsible for focal cortical deficits. Asymmetrical multifocal hypoperfusion in the perirolandic and posterior parietal cortices may favour the diagnosis of CBD, particularly in the early stage.

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