Constructional Impairment in a Case of Corticobasal Degeneration

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Abstract - Constructional impairment is not often reported in cases of corticobasal degeneration (CBD). We report a case who showed constructional impairment with clinical and neuroimaging features of CBD. The patient, a 64-year-old right-handed woman, presented with an insidious onset and slow progression of myoclonic jerks and dystonic contracture of her left hand within a year prior to this presentation. On examination, there were cortical sensory deficits and ideomotor apraxia. Magnetic resonance imaging showed an asymmetrical fronto-parietal cortical atrophy with more severity in the right parietal region. In the study of single photon emission computer tomography, regions of hypoperfusion were shown in the right parietal lobe as well as in the right basal ganglion. Although she achieved a full score for the Mini-mental State Examination and showed no impairments in simple figure copying, she presented constructional impairment in such psychometry items as Block Design, Object Assembly, and Digital-Symbol(table). In addition, she showed difficulties in global level perception of the global-local analysis and visuopraxic errors in Bender-Gestalt test.

Key Words : Corticobasal degeneration, Global-local analysis, Constructional impairment

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INTRODUCTION

Corticobasal degeneration (CBD) is a rare but clinically distinct syndrome. Awareness about this syndrome gradually grew after it was reported first by Rebeiz et al[1] in 1968 and subsequently, by other series [2,3,4,5]. The syndrome is characterized by an asymmetric extrapyramidal syndrome with reflex myoclonic jerk [6,7,8], severe rigidity, and finally dystonic contracture of the unilateral limb. There are also focal cortical symptoms such as apraxia and cortical sensory impairment[9]. However, less attention has been drawn to cortical functions other than apraxia or cortical sensory deficits.

It has been long recognized from the research of neuropsychology[10,11] that the right hemisphere is more involved in the processing of gestalts or wholes, while the left hemisphere, of parts or details. Hierarchical visuospatial processing in unilateral brain-damaged patients was investigated by analyzing information processing at global and local levels[12,13,14]. Therefore, visuospatial dysfunction could be expected in the case of CBD where parietal lobe involvement is the rule.

We report a patient who suffered from CBD with constructional impairment and who had difficulty in processing global level information as well as other visuopraxic

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functions.

CASE REPORT

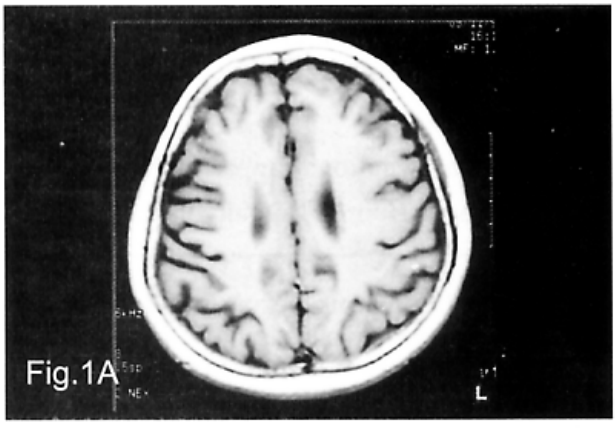
A 64-year-old right-handed Taiwanese housewife started to complain of stiffness and clumsiness of her left upper limb a year and half prior to her admission to our hospital. Initially, she had difficulties in doing up buttons during dressing or tying during needlework. Soon she experienced rhythmic jerk-like tremor of her left hand. Gradually, the stiffness and jerky movement extended to the forearm. Finally, the left hand became painfully contracted.

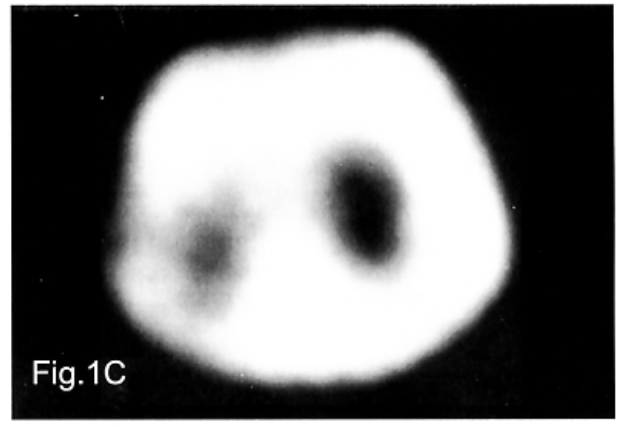
On examination, her left arm adopted a fixed flexion-adduction position with a flexion-contracture hand posture. The left

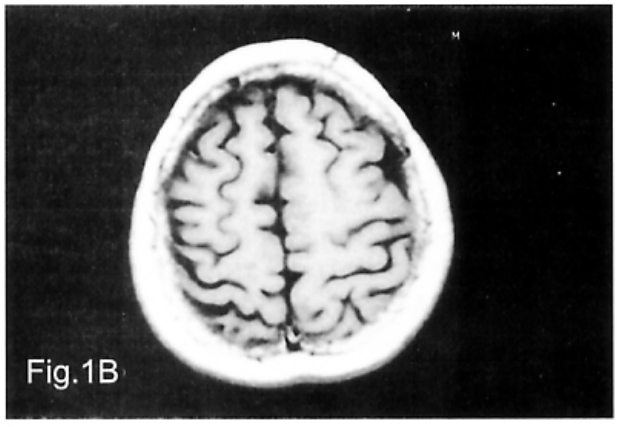
Table 1 Summary of the Neuropsycholgcial Assessment

Test	Result	Remark
MMSE	30/30	
WAIS-R	Verbal IQ 74	Borderline subnormality;
Performance IQ	66	Difficulty in Block Design, Object
Full IQ	70	Assembly, Digital Symbol
Bender-Gestalt Test	Abnormal	Impaired visuopraxic function; poor visual memory
Praxis Test		
Ideomotor	Pantomime (3/10)	Performed on the right upper limb
	Imitation (8/10)	
	Real object (10/10)	
Ideational	Normal	
Perception Test	Tactile agnosia, Agraphesthesia	Poor performance on the left limbs
Global-local Analysis	Perception difficulty at global level	No difficulty in copying and identifying local elements

MMSE: Mini-mental State Examination;
WAIS-R: Wechesler Adult Intelligence Scale - Revised.







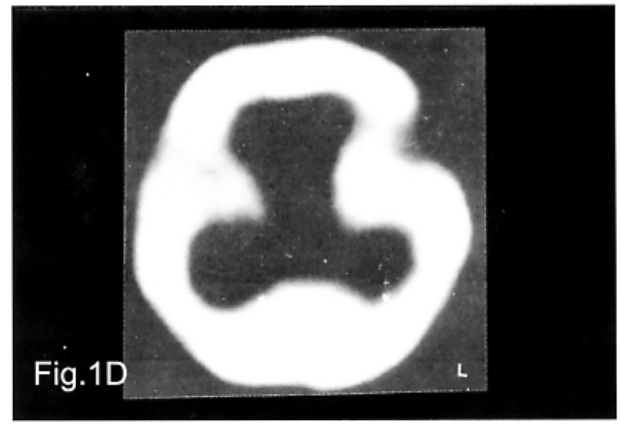


Fig. 1. MRI showing asymmetrical cortical atrophy, mainly in the right parietal and frontal lobes (A, B). HMPAO-SPECT showing marked hypoperfusion in the right parietal lobe (C) and right basal ganglion and mild in the bifrontal areas (D).

index finger assumed an extension position while other fingers of the same hand remained tightly flexed. The left wrist flexors were mildly hypertrophic. The left upper limb showed severe rigidity and intermittent quasi-rhythmic myoclonic jerks. The jerky tremor aggravated when the patient was initiating an action, sustaining a posture, or giving her attention to it. The jerks stopped when she fell asleep. There was also mild rigidity in her left lower limb and right limbs. On walking, she showed some axial dystonia with her left shoulder extended and externally rotated. Her left foot intermittently showed mild dystonic inversion. Her gait was slow and mildly shuffling but postural reflex was still good. No limitation was observed in vertical gaze. There were no slow saccades. The tendon reflexes were symmetric and brisk, and bilateral plantar responses were flexor.

Magnetic resonance imaging showed asymmetrical frontoparietal cortical atrophy (Fig. 1A, 1B) with more severity in the right parietal lobe contralateral to the more affected limb. Regional cerebral blood flow study using single photon emission tomography, with tracer ^{99}Technetium hexamethyl-propylenamine showed significantly reduced uptake in the right parietal region (Fig. 1C) as well as in the right basal ganglion (Fig. 1D). Mildly decreased uptake in the bifrontal regions was also noted.

Surface electromyography (EMG) recorded at the wrist flexor and extensor of the left upper limb showed highly synchronized discharges in clusters of 2-4 short bursts (at a duration of 20-60 ms). Each train had a duration of 200-500 ms and recurred at a rate of 2-4 Hz with co-contractions of the agonist and antagonist.The long latency response,C-response, evoked by stimulating the left median nerve at wrist induced a large amplitude (2-2.5 mV) response recorded at the left abductor pollicis brevis with a latency of 38.5 ms but not on the right side. Electroencephalography showed intermittent diffuse slow waves at 3-7 Hz, 30-60 μV with slight emphasis in the right hemisphere.

On a structured praxis test[9], the use of object to command, such as scissors and chopsticks, was impaired. The performance of

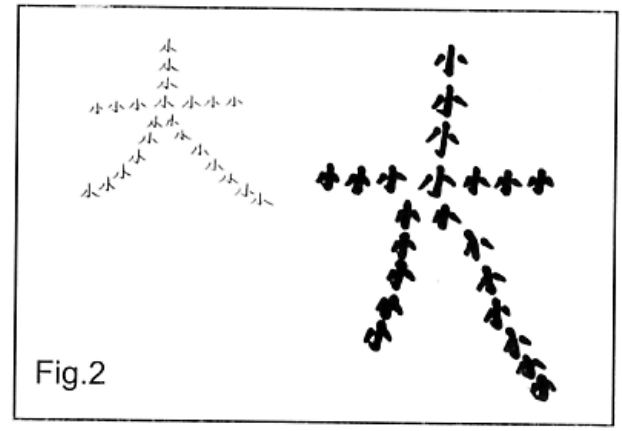


Fig. 2 Target character for the patient to identify and to copy (upper left) and the character copied by the patient (right). The patient could copy the target characters well but could not identify the global stimulus. The small character that make up the local stimuli is "小" which means "small" in Chinese and the large character that make up the global stimulus is "大", which means "large" in Chinese.

the ideomator praxis was poor with imagined objects (3/10), better with imitations (8/10), and normal with handling of the real objects (10/10). Since the left hand was not able to perform any command because of the flexion contracture, we performed the praxis test on her right hand. No ideational apraxia or alien hand was observed.

She had cortical sensory deficits, such as tactile agnosia and agraphesthesia on the left hand and agraphesthesia in the left foot. She did not experience aphasia or have difficulty with the random numeral cancellation task.

Her score at the Mini-mental State Examination was full (30/30) during the clonazepam free period (versus a score of 26 under clonazepam 1.0 mg twice a day). The psychometry, Wechesler Adult Intelligence Scale - Revised (WAIS-R), showed a full scale IQ of 70, a verbal IQ of 76, and a performance IQ of 66. The verbal IQ was about or slightly below the average referring to her age and her 2-year school educational background.The performance IQ was significantly lower than that of her age-matched control. It was inadequate in the items of Block Design, Object Assembly, and Digital-Symbol. She did not exhibit difficulty in drawing a clock or copying a flower or simple geometry designs.

With the Bender-Gestalt test, she had impairment in visuopraxic function, e.g., inverted curvature in the figure that she copied. Poor visual memory (1/6) was also found in the same test. In the global-local analysis, hierarchical stimuli were presented as geometry form, numeral and Chinese character for identification and copying. She showed impairment in the processing of global level information. For example, she was able to copy the local elements well and the global form fairly (Fig. 2), and identify the local targets readily. She, however, ignored the global target unless cues were given from the examiner by outlining the global form. This phenomenon prevailed in all characters (5/5), most geometry forms (4/5), and numerals (4/5).

During the hospitalization, L-dopa with carbidopa, and trihexyphenidyl were tried with no effect. On the other hand, diazepam and clonazepam showed significant effect in reducing the stiffness and myoclonic jerks with the adverse effects of sleepiness and mental slowness.

DISCUSSION

Differential diagnosis must be made between CBD and other akinetic rigid syndromes[4] with asymmetric involvement, such as Parkinson's disease and Steele-Richardson-Olszewski syndrome. However, the poor clinical response to L-dopa [2,3,4], early manifestations of apraxia, cortical sensory impairment, unilateral myoclonic jerks and dystonia all help to ease this diagnostic task. Furthermore, the typical hand posture is also a good hint pointing to the diagnosis of CBD. The ancillary laboratory examinations offer further supporting evidence. For example, the characteristic findings in surface EMG [7] elucidate the coexistence of dystonia and myoclonic jerks. The C-reflex validates the existence of a reflex myoclonus. The neuroimages confirm the structural, perfusional[15] or metabolic defects[16,17] within the cortical and basal ganglionic regions.

Constructional impairment is rarely reported in CBD cases. In the cases of Gibb et al[2], evidence of the visuospatial impairment manifested as difficulty in copying simple geometric designs and poor performance on the Block Design test. Our patient, however, did well with copying simple geometry designs and with simple object drawing. She showed constructional impairment on the psychometric test of WAIS-R. Some visuo-constructive disability, such as picture arrangement, has also been reported in the study of apraxia in Leiguarda et al [9]. In our case, similar dysfunction can also be found on the Bender-Gestalt test. In addition, the Bender test disclosed her poor visual memory. The complex visuographic-copying task appears to be associated with parietal lesion, especially with the right parietal lobe. The visuopractic task, such as picture arrangement, is also sensitive to frontal dysfunction [18].

In addition to the preceding constructional impairments, the patient showed difficulty in the global-level information processing. The global-local analysis provides a hierachical stimulus consisting of a large global form composed of a series of smaller local forms or elements. The task is capable of examining the effects of directed attention and interference in the processing of global and local information [11,12]. Patients with right inferior parietal lobe lesion were shown to have global perception impairment while those with left parietal lobe lesion, local perception deficits[12]. Therefore, the global perception difficulty in our case is in agreement with the right parietal lobe lesion proposed by such model.

Ideomotor apraxia is the most frequent type of apraxia in CBD[9]. Our case showed significant ideomotor apraxia but not ideational apraxia. The "motor engram" from learning a motor skill is thought to be stored in the inferior parietal lobe[19]. Either damage to the storage area or disruption of production pathway of the praxis system may result in ideomotor apraxia. On the other hand, ideational apraxia may be due to the failure of the "conceptual" system, which is often associated with general intellectual dysfunction, such as Alzheimer's dementia. Usually the praxic disturbance of CBD can be examined only in the less affected limb because there is always severe rigidity or even

dystonic contracture in the more affected limb.

We conclude that CBD is an asymmetrical extrapyramidal disorder with focal cortical atrophy. Clinical findings as well as laboratory evidence indicated lesions at the basal ganglion and focal cortical areas emphasizing parietal and frontal lobes. Therefore, the importance of recognizing various focal cortical symptoms in CBD cases could never be overemphasized. Constructional impairment, cortical sensory dysfunction, and ideomotor apraxia should be carefully looked for in all cases of this unilateral akinetic rigid syndrome.

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