should be similar. Although a consensus has yet to be found for the management of this unpredictable, relatively new entity, GTR, thorough systemic investigation and long-term follow-up are mandatory.

Appendix A. Supplementary material

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.jocn.2012.02.031.

References

1. Carneiro SS, Scheithauer BW, Nascimento AG, et al. Solitary fibrous tumor of the meninges: a lesion distinct from fibrous meningioma-A clinicopathologic and immunohistochemical study. *Am J Clin Pathol* 1996;**106**:217–24.
2. Wagner E. Das tuberkelähnliche lymphadenom (Der cytogene oder reticulirte Tuberkel). *Arch Heilk (Leipzig)* 1870;**11**:497–9.
3. Klemperer P, Rabin CB. Primary neoplasms of the pleura. *Arch Pathol* 1931;**11**:385–412.
4. Jallo GI, Roonprapunt C, Konthbauer K, et al. Spinal solitary fibrous tumor: a series of four patients: case report. *Neurosurgery* 2005;**57**:E195.
5. Metellus P, Bouvier C, Guyotat J, et al. Solitary fibrous tumors of the central nervous system: clinicopathological and therapeutic considerations of 18 cases. *Neurosurgery* 2007;**60**:715–22.
6. Munoz E, Prat A, Adamo B, et al. A rare case of malignant solitary fibrous tumor of the spinal cord. *Spine* 2008;**33**:E397–399.
7. Louis DN, Ohgaki H, Wiestler OD, et al. The 2007 WHO classification of tumors of the central nervous system. *Acta Neuropathol* 2007;**114**:97–109.
8. Gengler C, Guillou L. Solitary fibrous tumor and haemangiopericytoma: evolution of a concept. *Histopathology* 2006;**48**:63–7.
9. Cole CD, Schmidt MH. Hemangiopericytomas of the spine: case report and review of the literature. *Rare Tumors* 2009;**1**:e43.

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Is progressive upper-body apraxia a corticobasal syndrome?

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ABSTRACT

Corticobasal degeneration (CBD) is characterized by various clinical manifestations including corticobasal syndrome, progressive supranuclear palsy-like syndrome and frontotemporal dementia. Focal cortical atrophy syndrome as the initial manifestation rarely occurs in CBD. Here, we present a 62-year-old man and a 70-year-old man who were admitted due to clumsiness in the arms. On initial neurological examination, they showed asymmetric limb apraxia without parkinsonism or global cognitive dysfunction. Brain MRI showed focal atrophy in the frontal and prefrontal cortices, and brain positron emission tomography scan revealed decreased metabolism in these same brain locations. Although these patients developed parkinsonism and dystonia within several years, the neurological signs were limited to the arms for a long period. "Progressive upper-body apraxia" may be a rare clinical manifestation of CBD which shows a benign clinical outcome. The patients described may enhance our understanding of the clinical heterogeneity of this disease.

1. Introduction

Corticobasal degeneration (CBD) is a neuropathologic disease in which neurological signs progress rapidly in most patients, leading to neurological disability shortly after the initial diagnosis.[1,2] One prominent clinical feature of CBD is corticobasal syndrome (CBS), which consists of prominent asymmetric parkinsonism combined with cortical dysfunction. However, clinically diagnosed CBS has only a 24% positive predictive value for pathologically confirmed CBD.[3] CBD is associated with various clinical manifestations;[2,3] for example, some patients with CBD present initially with an isolated focal cortical atrophy syndrome,[2,4–7] which consists of isolated cortical dysfunction without other neurological deficits, although parkinsonism develops as the disease progresses. Isolated focal cortical atrophy syndrome has been associated with a wide spectrum of clinical outcomes that range from grave to favourable. Here we describe two patients who presented initially with isolated "upper-body apraxia" and whose neurological signs were limited to the upper extremities for a long period.

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2. Case report

2.1. Patient 1

A 62-year-old man was admitted due to clumsiness in both hands that started in his left hand 2 years prior and gradually progressed to his right hand over the subsequent 6 months. On neurological examination, he showed difficulty imitating meaningless gestures with both hands, with the defect being more severe on his left side. He showed errors in producing both transitive and intransitive gestures. However, his general cognitive function was not impaired (Table 1). His facial expressions were slightly decreased, and his range of arm movement was reduced on the left side while walking. There was no bradykinesia or rigidity in his legs. Gait and postural stability were not impaired. His vertical eye movement was normal. Brain MRI showed atrophy in his central and inferior frontal gyri, which was more pronounced on the right side (Fig. 1A). Brain positron emission tomography (PET) revealed decreased metabolism in these same regions (Fig. 1B). Three years after diagnosis, the patient was unable to move both arms because of marked parkinsonism and apraxia, despite high doses of levodopa. However, he was still able to walk without assistance, and parkinsonism was not apparent in his lower limbs.

Table 1
Neuropsychological test in two patients with progressive upper-body apraxia

Test components		Patient 1		Patient 2	
		Score	Percentile score	Score	Percentile score
K-MMSE		25	18.4	28	66.3
CDR		0		0	
Praxis					
Intransitive gestures					
Waving good-bye		Correct		Incorrect	
Saluting		Incorrect		Incorrect	
Beckoning		Correct		Correct	
Holding the nose		Correct		Correct	
Hailing		Correct		Correct	
Transitive gestures					
Cutting Kimbab		Incorrect		Incorrect	
Nailing with a hammer		Incorrect		Correct	
Handling a screwdriver		Correct		Correct	
Turning a key		Correct		Correct	
Combing hair		Correct		Correct	
Mimicking 5 meaningless gestures		Incorrect in 5 gestures		Incorrect in 4 gestures	
Buccofacial praxis					
Sniffing		Correct		Correct	
Whistling		Correct		Correct	
Sucking		Correct		Correct	
Wink		Correct		Correct	
Blowing		Correct		Correct	
Memory					
Delayed recall		6	42.1	5	27.6
Recognition		10/1	44.0	10/1	69.9
Language					
Fluency		Fluent		Fluent	
Comprehension		Normal		Normal	
Repetition		Normal		Normal	
Naming (K-BNT)		44	51.6	39	42.1
Reading		Normal		Normal	
Writing		Normal		Normal	
Calculation		Normal		Normal	
Frontal/executive function					
Motor impersistence		Normal		Normal	
Verbal fluency	Animal	17	59.5	5	0.5
	Supermarket	18	55.2	5	3.6
Korea-Color Word Stroop Test (Time-per-item score)		0.64	73.6	0.86	53.2
Visuospatial function (Rey Complex Figure Test)		32	51.6	20	0.2

CDR = Clinical Dementia Rating, K-BNT = Korean form of Boston Naming Test, K-MMSE = Korean version of the Minimental Status Examination.

2.2. Patient 2

A 70-year-old man was admitted due to clumsiness in the right arm and mild dysarthria that had developed 2 years prior. He had been taking antihypertensive and antidiabetic medications during the previous 3 years. Neurological examination showed that he had difficulties imitating meaningless hand gestures on his right side. He was not able to execute intransitive and transitive tasks precisely. Neuropsychological tests showed mild dysfunction in frontal-executive tasks and visuospatial tasks (Table 1). The level of parkinsonism was not remarkable and vertical eye movement was normal. Brain MRI showed atrophy in the left frontal and prefrontal cortices (Fig. 1C), and brain PET revealed decreased metabolism in these same brain locations (Fig. 1D). The clumsiness in his right arm gradually progressed, involving his other arm over the next 2 years. Follow-up neurological examination showed a dystonic posture in his right arm and overt parkinsonism in his left arm. However, he was able to walk without assistance because parkinsonism did not involve his lower limbs.

3. Discussion

Both patients described here showed clumsiness of the upper extremities as the initial presentation of the disease. They had more difficulty in imitating meaningless hand gestures than meaningful performances, although they showed some errors in producing both transitive and intransitive gestures upon verbal command. These findings are consistent with the common patterns of apraxia in CBS.[8] The brain imaging findings suggest that limb apraxia in these patients began as cortical atrophy in the frontal area. Overt parkinsonism appeared in patient 1, and parkinsonism with dystonia became evident in patient 2 on follow-up neurological examination. These findings support a diagnosis of CBS.

The concept of CBD has been changing since the unique clinical manifestations of this disease were originally described.[2] The classic signs of CBD include prominent asymmetrical clumsiness of the arms due to a combination of cortical and basal ganglia dysfunction. Recent clinico-pathological studies have shown that pathologically diagnosed CBD can include various clinical manifestations, including CBS, progressive supranuclear palsy (PSP)-like syndrome, primary non-fluent aphasia, and frontotemporal dementia.[2,3] Patients with clinically diagnosed CBS, conversely, may have different pathological disease entities, including CBD, PSP, frontotemporal lobar degeneration and Alzheimer's disease. Therefore, it may be more appropriate to call these conditions "tauopathies", a molecular–pathologic term. Although we were not able to confirm the exact pathologic diagnosis in these patients, the clinical manifestation and neuroimaging findings in both suggest a diagnosis of CBD.

Recently, several patients presenting with unusual clinical manifestations of CBD have been described. These clinical