An Unusual Case of Corticobasal Syndrome

Gökçen GÖZÜBATIK-ÇELİK, Ayşegül GÜNDÜZ, Semra OĞUZ, Güneş KIZILTAN, Sibel ERTAN

Istanbul University Cerrahpaşa Faculty of Medicine, Department of Neurology, Istanbul, Turkey

Dear Editor,

The constellation of cortical features and extrapyramidal findings without pathological confirmation is corticobasal syndrome (CBS) (1). Nearly half of the CBS cases have the pathological diagnosis of corticobasal degeneration (CBD). Alien limb and apraxia generally involve the upper extremity at the beginning, and the recognition of apraxia and the diagnosis of CBS are usually delayed. Alien leg or leg apraxia presentations are rarer, increasing the diagnostic challenge.

A 62-year-old right-handed woman was admitted with gait difficulty in 2010. She had initially developed clumsiness of her right leg, specifically difficulty with pushing the dustbin pedal with her right foot in 2003. At this time, she was diagnosed to have sensorimotor demyelinating polyneuropathy. The diagnosis of polyneuropathy led to genetic assessments, which showed a 17p11.2 mutation (HNPP deletion). Her complaints progressed, and she was operated on twice for scoliosis in 2005 and 2010. She had a traffic accident due to inability to use the pedals of the automobile due to the inability to put her foot on the brake on time, despite knowing how to do it. Forgetfulness, literal restriction, and difficulty with writing appeared in the following years. Gradual worsening continued, leading to the inability to walk and stand without assistance, without any response to 375 mg/day levodopa.

On admittance, she had dystonic posture of the right foot, first toe, and right hand; bilateral upper and lower limb ideomotor apraxia predominantly on the right leg; bradykinesia; mild bradykinesia; and impaired discriminative sensation over bilateral palms. She scored 18/30 on the Mini-Mental State Examination.

Cranial magnetic resonance imaging (MRI) showed atrophy of the corpus callosum and frontal parietal cortex, as well as

loss of white matter with widespread asymmetric lesions in white matter without any contrast enhancement. Detailed laboratory cerebrospinal fluid examinations were normal.

The eventual diagnosis was CBS (clinically probable CBD=pCBD) with lower limb apraxia at onset and coexisting hereditary polyneuropathy.

Our case has two major features: 1. presentation of CBS with lower limb apraxia with relatively slow progressive course and 2. coexistence with hereditary sensorimotor demyelinating polyneuropathy.

Lower limb presentation was previously reported in 10 patients out of 36 CBS cases, whereas 20 had upper limb apraxia at the beginning (2). Lalive and colleagues (3) previously reported clinical and radiological findings of a similar patient in whom the eventual diagnosis was CBD, confirmed by pathological analysis. The initial diagnosis in this patient was neuropathy, and the second diagnosis was myelopathy. We may suggest that physicians tend to explain the functional impairment of extremities based on peripheral, cerebellar, or pyramidal involvement but do not carry out a formal apraxia examination.

As we did not follow her from the beginning, 'presentation with apraxia' may seem to be a speculation. Her main complaint from the onset, together with absence of weakness of ankle movements despite the presence of the same symptom as in 2003, absence of any evidence of pyramidal symptoms on examination, or absence of radiological features of myelopathy or radiculopathy, strongly suggests presentation with leg apraxia at onset.

Although there are rare reports of HNPP with central nervous system (CNS) lesions (4), the diagnosis of hereditary polyneuropathy may only be a coincidence. Besides, we think that her lesions in the white matter may not account for her disability

Correspondence Address

Dr. Gökçen Gözübatık-Çelik, İstanbul Üniversitesi Cerrahpaşa Tıp Fakültesi, Nöroloji Anabilim Dalı, İstanbul, Türkiye

Phone: +90 554 937 00 87 E-mail: gokcen3@hotmail.com Received: 15.02.2014 Accepted: 19.02.2014

©Copyright 2014 by Turkish Association of Neuropsychiatry

caused by apraxia, parkinsonism, or dystonia due to their localization and distribution, and cortical/callosal atrophy is more compatible with the diagnosis of probable CBD/CBS.

References

1. Mathew R, Bak TH, Hodges JR. Diagnostic criteria for corticobasal syndrome: a comparative study. *J Neurol Neurosurg Psychiatry* 2012; 83:405-410. [\[CrossRef\]](#)
2. Rinne JO, Lee MS, Thompson PD, Marsden CD. Corticobasal degeneration. A clinical study of 36 cases. *Brain* 1994; 117:1183-1196. [\[CrossRef\]](#)
3. Lalive PH, Personeni O, Slosman DO, Landis T, Burkhard PR. Lower limb corticobasal degeneration. *Eur Neurol* 2000; 44:248-249. [\[CrossRef\]](#)
4. Sanahuja J, Franco E, Rojas-García R, Gallardo E, Combarros O, Begué R, Garnes P, Ila I. Central nervous system involvement in hereditary neuropathy with liability to pressure palsies: description of a large family with this association. *Arch Neurol* 2005; 62:1911-1914. [\[CrossRef\]](#)