Pure prosodic type of primary progressive apraxia of speech mimicking non-fluent aphasia in patients with corticobasal syndrome

M. Kwon Kwon¹, W. Hyun Shim², Y. Jo², S. Park¹, J.H. Lee¹

¹Department of Neurology, University of Ulsan College of Medicine, Asan Medical Center, South Korea ²Department of Radiology, University of Ulsan College of Medicine, Asan Medical Center, South Korea

Corticobasal syndrome (CBS) shows heterogeneous underlying pathology and clinical features. Recent studies found that speech-language disturbances, mainly non-fluent aphasia, are the commonest features in patients with CBS. However, differential diagnosis of speech-language disorders is complex. Primary progressive apraxia of speech (PPAOS) is rarely reported as a initial symptom of neurodegenerative diseas. Recently, two different subtypes of PPAOS were identified in those patients: predominant phonetic type and prosodic types. We present two cases of patients with pure prosodic type of primary progressive apraxia of speech (PPAOS) mimicking primary progressive non-fluent aphasia (PNFA). A 65-year-old woman (P1) and a 72-year-old man (P2) were referred to our speechlanguage clinic under the possible diagnosis of PNFA. The neurologic examinations revealed no prominent abnormalities except for mild rigidity in their left arms and the insidious onset of speech problems with gradual progression. Their speech was slow and effortful with frequent pauses. They spoke mainly sentences with simple syntactic structures. However, the result of comprehensive aphasia test showed that their receptive language ability was within the normal range. They also wrote fluently a passage consisted of complex sentences. The results of speech analysis demonstrated spastic component of dysarthria with relatively intact articulation in both patients. There was no evidence of phonemic type of apraxia of speech or buccofacial apraxia but mild ideomotor apraxia was noticed in both patients. Their brain MRI was unremarkable. The results of 18F-FDG PET study revealed glucose uptake reduction in their right prefrontal areas. We concluded the patients had possible CBS and pure prosodic type of PPAOS was their initial presentation of CBS. Comprehensive speech-language evaluation and clinic-radiological correlation are essential for differential diagnosis for the patient with CBS. Pure prosodic type of PPAOS can be found as a rare initial presentation of CBS.