The arcuate fasciculus is the neural tract connecting the two major speech centers (Broca's and Wernicke's areas) and plays a critical role in language function (Casati and Mesulam, 2008). Injury of this tract can lead to several types of language impairment, including conduction aphasia, Broca's aphasia, anomic aphasia, and apraxia of speech (Anderson et al., 1999; Bartha and Benke, 2003; Casati and Mesulam, 2008; Bernal and Ardila, 2009; Kim et al., 2011).

Motor neuron diseases (MNDs) are a group of neurological disorders characterized by progressive degeneration of upper motor neurons originating in the motor and premotor cortex, and lower motor neurons originating in the spinal cord and brain stem. MNDs include primary lateral sclerosis (selectively affecting upper motor neurons), progressive muscular atrophy (exclusively causing damage to lower motor neurons), amyotrophic lateral sclerosis (involving both upper motor neurons and lower motor neurons), and progressive bulbar palsy (primarily involving motor neurons in the brain stem) (Adams et al., 1997). As a result of damage to motor neurons, patients with MNDs present with muscle weakness, atrophy, and fasciculation throughout the body (Adams et al., 1997).

MNDs are known to predominantly affect motor function. However, recent studies have reported accompanying extra-motor manifestations, including aphasia, in patients with MNDs (Caselli et al., 1993, 2004; Tsuichiya et al., 2000; Bak et al., 2001; da Rocha et al., 2007), although the prevalence of the co-occurrence of motor dysfunction and extra-motor symptoms has not been clarified. The pathogenic mechanisms of aphasia in patients with MNDs have been reported to be the result of atrophy or hypo-perfusion in the fronto-temporal lobe, which has been demonstrated using conventional brain CT/MRI, radionuclide imaging, and post-mortem brain autopsies (Tsuichiya et al., 2000; Bak et al., 2001; Catani et al., 2004; da Rocha et al., 2007). However, these methods are limited in that they cannot demonstrate lesions of neural tracts. By contrast, the recently developed diffusion tensor tractography (DTT), which is derived from diffusion tensor imaging (DTI), has enabled three-dimensional visualization and detailed estimation of neural tracts (Jang et al., 2013a, b). Several studies have demonstrated the degeneration of neural tracts using DTI, including the corticospinal tract, corpus calloso fibers, and uncinate fasciculus in patients with MNDs (Sage et al., 2007; Hong et al., 2008; Ben Bashat et al., 2011; Sarro et al., 2011; van der Graaff et al., 2011; Zhang et al., 2011). However, little is known about injury of the arcuate fasciculus.

The classic MND is characterized by progressive loss of limb, bulbar, and respiratory muscle function caused by selective degeneration of upper motor neurons and lower motor neurons, sparing the rest of the nervous system (Brooks, 1994). Accordingly, most previous DTI studies on patients with MNDs have focused on the corticospinal tract, demonstrating the correlation of injury of the corticospinal tract with deterioration of motor function (Sage et al., 2007; Hong et al., 2008; Zhang et al., 2011). Regarding aphasia in patients with MNDs, since the study reported by Caselli et al. in 1993, several studies have reported non-fluent aphasia in patients with MNDs (Caselli et al., 1993, 2004; Tsuichiya et al., 2000; Bak et al., 2001; Catani et al., 2004; da Rocha et al., 2007). In those previous studies, atrophy and hypo-perfusion in the left fronto-temporal lobe were consistently observed on conventional CT/MRI and radionuclide imaging, respectively (Tsuichiya et al., 2000; Bak et al., 2001; Catani et al., 2004; da Rocha et al., 2007). In addition, in postmortem examination, the most common findings were atrophy and neuronal loss in the left fronto-temporal lobe (Caselli et al., 1993, 2004; Bak et al., 2001; da Rocha et al., 2007). However, so far, no study evaluating the arcuate fasciculus in MND patients with aphasia using DTI has been reported. Therefore, this is the first study using DTT to demonstrate injury of the left arcuate fasciculus in an MND patient.

In the current study, using the configuration and parameters of DTT, we attempted to demonstrate damage of the arcuate fasciculus in a patient with progressive bulbar palsy who presented with Broca's aphasia. A 74-year-old right-handed man visited the rehabilitation department of a university hospital for evaluation of language disturbance and dysphasia. He had no family history of neurological disease. Five years ago, the patient had begun to notice slurred speech, which had shown slow deterioration with time. One year ago, he had begun to show dysphagia (coughing during eating or drinking, difficulty in chewing and controlling food in the mouth, and drooling of saliva). On neurological examination, he was alert and oriented. He exhibited decreased gag reflex and soft palatal movements, and the jaw jerk reflex and tongue fasciculation were observed. In addition, he had mild quadriparesis (4/5 in the right upper and lower extremities and 4/5 in the left upper and lower extremities) and a hemi-hypoesthesia in the right hand. The deep tendon reflex was significantly increased, particularly in the left upper and lower extremities. He showed a full score of 30 points on the Mini-Mental Status Examination for evaluation of cognitive function (Han et al., 2008). Conventional brain MRI and electromyography/nerve conduction studies revealed no abnormal findings (Figure 1A). The patient showed dysarthria (articulatory errors at sentence level: 30.1%) on the language function test, and Broca's aphasia (aphasia quotient: 74%ile, fluency: 61%ile, comprehension: 86%ile, repetition: 71%ile, naming: 86%ile) on the Korean-Western Aphasia Battery (K-WAB) (Kim and Na, 2004). As a result, based on the patient's history and neurological examination, he was diagnosed as progressive bulbar palsy. This study was approved by our hospital review board.

DTI data were acquired at 5 years after symptom onset using a 6-channel head coil on a 1.5 T. Sixty contiguous slices (reconstruction matrix = 192 × 192; field of view = 240 × 240 mm²; echo time = 76 ms, b = 1,000 mm²/s, number of excitations = 1, thickness = 2.5 mm) were acquired for each of the 32 noncollinear diffusion-sensitizing gradients. DTT-Studio software was used for reconstruction of the arcuate fasciculus. Based on the method of Nucifora et al. (2005), we placed the region of interest (ROI) for tracking of the arcuate fasciculus (the seed ROI in the posterior parietal area of the superior longitudinal fascicle and the target ROI in the posterior temporal lobe). The seed ROI was shown in green and the target ROI was shown in blue on DTT-based color-coded maps. Terminological criteria for tracking were fractional anisotropy < 0.2 and angle < 60° (Jang and Lee, 2014). On DTT images, the arcuate fasciculus in the right hemisphere of the patient originated from Wernicke's area, passed through the known arcuate fasciculus pathway, and then entered into Broca's area (Figure 1B). Also, we recruited an age- and sex-matched normal subject (72 years old, male) with no history of neurological or psychiatric diseases. The DTT of the normal subject showed well-preserved bilateral arcuate fasciculus, passing through its known pathway.

In the current study, using the configuration and parameters of DTT, we found discontinuation of the left arcuate fasciculus (in the dominant hemisphere) in a patient with progressive bulbar palsy between Wernicke's and Broca's areas, although no remarkable lesions were observed on conventional magnetic resonance images. The injury of the left arcuate fasciculus in this patient appeared to coincide with Broca's aphasia. We recommend evaluation of the arcuate fasciculus using DTT in patients with MNDs who present with aphasia. However, this study has several limitations. First, the diagnosis of progressive bulbar palsy is usually conducted by physical examination and history taking and often confused with other types of MND (such as ALS, primary lateral sclerosis, and progressive muscular atrophy), thus accurate diagnosis of progressive bulbar palsy is limited. Second, we could not verify the absence...
of other diseases (small size infarction or traumatic axonal injury) which can cause the aphasia or arcuate fasciculus lesions. Third, we obtained DTI data using 1.5 T MRI machine; by contrast, higher tesla MRI machine can show better resolution. Fourth, this is a single case study. Therefore, further studies addressing these limitations are needed.

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Last, our study is limited by the fact that the patient had a unique lesion. Further studies are needed to address this limitation.

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