

소아재활

게시일시 및 장소 : 10 월 19 일(토) 08:30-12:30 Room G(3F)

질의응답 일시 및 장소 : 10 월 19 일(토) 11:00-11:30 Room G(3F)

## **P 3-102**

### **Absence of Arcuate Fasciculus in a Child with Bilateral Perisylvian Polymicrogyria**

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#### **Introduction**

Congenital bilateral perisylvian polymicrogyria occurs due to bilateral perisylvian cortical malformations, and is characterized by pseudobulbar palsy, feeding disturbances and facial diparesis as well as seizures since early infancy. The speech impairment was more pronounced than what would be expected from the severity of the cortical pseudobulbar palsy. Thus, it has been suggested that the linguistic defect in congenital bilateral perisylvian polymicrogyria patients was caused due to the dysfunction of the perisylvian language networks of the brain. In the current study, we used diffuse tensor tractography (DTT) to identify the dysfunction of the perisylvian language networks. We focused our analysis on the arcuate fasciculus (AF), which belongs to the core perisylvian circuit underlying the speech function.

#### **Case report**

The patient was a 10-year-old female. She visited our institution for evaluation of severe dysarthria. Gestation and pregnancies were unremarkable. She had delayed developmental milestones with severe delay in developing speech. She had difficulty in sucking during the perinatal period. Development of language skills was severely impaired. She had a marked tongue movements deficit, with severe dysarthria and a nasal tone. She also showed oromotor apraxia and pseudobulbar symptoms. Mild mental retardation (IQ 43 and SQ 58.25) was documented on the Korean-Wechsler Intelligence Scale for Children-third edition and Social Maturation Scale. Magnetic resonance images (MRIs) revealed an abnormally thickened frontoparietal perisylvian cortex bilaterally, consistent with bilateral parietal polymicrogyria, Fiber tracking was processed using DTI Studio (Johns Hopkins University, Baltimore, MD, USA) based on fiber assignment using the continuous tracking algorithm which can estimate a single dominant diffusion orientation within the imaging voxel. The termination criteria used for fiber tracking were fractional anisotropy (FA) < 0.25, angle < 70°. the bilateral arcuate fascicles were not seen in the patient

**Discussion**

This would be the first case report, which showed the absence of arcuate fascicle in a patient with congenital bilateral perisylvian polymicrogyria. The arcuate fascicle is a major white matter tract that is one of the primary fiber bundles involved in human language processing. This tract connects the Broca's area in the frontal lobe, a region mainly involved in speech production, with the Wernicke's area in the temporal lobe, a region related to speech comprehension. The absence of AF in our patient might have been caused due to the failure of formation of appropriate connections between the Wernicke's area and the Broca's area. Further mechanistic studies are required to explore the mechanisms for the absence of AF in a patient with congenital bilateral perisylvian polymicrogyria.