<シンポジウム19-3>認知症研究の新しい視点

Acquired Savant syndrome in frontotemporal dementia

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The patients with savant syndrome demonstrate outstanding mental capabilities despite coexisting severe mental disabilities. In most cases, savant skills are characterized by its domain-specificity, enhanced memory capability, and excessive focus on low-level perceptual processing. Although, savant skills have been reported especially in patients with Autistic spectrum disorders, mental retardation and left hemisphere damage, whether such skills arise in other neurological diseases has remained to be understood.

Dementia is characterized by a decline of cognitive and behavioral abilities in a previously unimpaired person, beyond what might be expected from normal aging. Recent case series reported that previously learned skills or talents could be conserved in the setting of dementia. In 1996, Miller reported 5 cases of "acquired-savant" patients who improved or newly acquired an artistic savant-like skill in the early stage of frontotemporal dementia (FTD) (Miller BL, 1998). On SPECT or pathology, 4 of 5 patients had the left-temporal variant of FTD. Their artistic products were diverse but share common features with autistic savant; all of them were domain-specific, mainly low-level perceptual abilities and never manifested in the verbal sphere. There are also a few convincing examples of pathology-induced changes of talent due to focal frontal or frontotemporal damage.

Although correct mechanisms underlying acquired savant syndrome remain to be known, several neuropsychological and neuroimaging studies have shed light on the relation ship between enhanced cognitive abilities and brain lesions. Kapur reviewed unexpected occurrence of behavioral improvement following brain injury, and used the term "paradoxical functional facilitation" (Kapur N, 1996). Snyder et al also reproduced savant conditions in healthy normal individuals by using TMS to create a virtual lesion in the left anterior temporal lobe, a site implicated in the savant conditions (Snyder A, 2007).

The degenerative dementias all have their own distinctive anatomy, devastating specific brain areas while leaving other regions unaffected. Focusing on the artistic abilities of these patients may have a positive therapeutic value, by encouraging patients to pursue an activity in which they can improve. Future studies using physiological techniques such as functional magnetic resonance imaging should help us to understand the brain regions that are activated during the artistic process in these patients.

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