<シンポジウム19-1>認知症研究の新しい視点

Learning from History-FTD, ALS and Behaviour

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To speak about history at a meeting dedicated to the "Future of Neurology" might at first seem surprising. However, the relationship between FTD and ALS is an area in which looking to the past might be, at the same time, one of the best ways to move forward. Until relatively recently, Amyotrophic Lateral Sclerosis (ALS) was perceived, particularly in the English-speaking countries, as a purely motor disease, sparing entirely higher mental functions such as cognition, language, personality and behaviour. In my presentation, I will argue that this perception has been influenced by two factors: lack of knowledge of non-English sources and a sharp divide between neurology and psychiatry in the countries dominated by the Anglosaxon tradition.

In contrast, the continental European literature, including countries influenced by the European tradition, such as Japan or Brazil, has produced a wealth of detailed descriptions of cognitive and behavioural changes in ALS patients since the beginning of their scientific study in late 19th Century. However, what is more remarkable than the quantity of these reports is their quality. The picture emerging from the study of the "old masters" points clearly to a specific clinical syndrome: a prodromal phase, dominated by behavioural abnormalities, developing into the full picture of Frontotemporal Dementia (FTD) and followed by a classical ALS, with all its clinical characteristics. An explicit connection between ALS and Pick's Disease (as FTD would have been called at that time) was made as early as in 1932.

Moreover, the clinicians of the first of 20th century have also recognised two peculiar characteristics of the dementia associated with ALS, which go beyond the classical picture of FTD and only now begin to be appreciated by the scientific community: psychotic symptoms, such as delusions and hallucinations and disorders of language, in particular of writing. Thus, our current understanding of ALS is much closer to its early descriptions than it is to those of the last decades.

The relationship between motor, cognitive and behavioural symptoms in ALS reflects a much wider issue of the overlap between movement disorders, dementias and psychiatric syndromes. The close links between neurology and psychiatry in the European tradition are likely to have contributed to en early recognition of cognitive and behavioural symptoms of ALS. Ironically, while the Anglosaxon world is moving enthusiastically towards "neuropsychiatry", Germany is currently considering a break up of the links between neurology and psychiatry.