A geographical cluster of progressive supranuclear palsy in northern France

Presented by: Lawrence I. Golbe, MD
Professor of Neurology and Director, Division of Movement Disorders
Rutgers Robert Wood Johnson Medical School, New Brunswick, NJ

An international research team has found the first known geographical cluster of the rare brain disorder progressive supranuclear palsy (PSP) in an area of unusual contamination by industrial metals.

The first such cluster has been identified in a group of suburban industrial towns in northern France centering on Wattrelos and Leers. Since 2005, 98 patients with PSP have been diagnosed by Dominique Caparros-Lefebvre, MD, a geriatric neurologist at the Centre Hospitalier de Wattrelos. This is 11.7 times the number expected based on the area’s population.

Thirteen of the 98 patients have undergone autopsy, which in each case revealed typical PSP, showing that the condition is not a previously unknown PSP mimic. The likelihood of a genetic “founder effect,” where a mutation arises in a recent ancestor of a geographical localized population, is reduced by the fact that none of the 98 patients was known to be related to any other. A toxic cause of the cluster seems most likely because for most of the 20th Century, this area was a center for metals-related industry such as ore extraction, tanning and dyeing. Waste materials from these activities persist in residential areas.

The research group is now starting a study to compare the PSP patients to their healthy neighbors with regard to exposure to metals, other potential toxins and genetic factors. The group has also started experiments to assess the effect of locally identified toxins on brain cells growing in the laboratory. If the cause of this PSP cluster can be found, it will provide an important clue to the cause of PSP elsewhere and of other, more common brain degenerations such as Alzheimer’s and Parkinson’s diseases.