Case of Treatable Autoimmunity Initially Diagnosed as Dementia

BY KERRI WACHTER  Senior Writer

BUDAPEST, HUNGARY — Autoimmune stratal dysfunction may be the underlying cause of dementia-like presentations, in rare cases. This was the case for a 48-year-old woman, who presented with a 1-year history of progressive difficulties with attention and memory, said Gabriel C. Léger, M.D., speaking at the 4th International Congress on Autoimmunity.

Her husband complained that there had been a profound change in her personality. In particular, he found her to have become uninhibited and exhaustingly hypersexual. She had progressive difficulties functioning at home and at work—finally losing her job. Her clinical history included an episode of self-limiting, ballistic-like movements of the right side of her body during her early 20s. Exhaustive testing turned up no cause.

She had no history of rheumatic fever or childhood chorea. However, at the age of 17 years her mother had Sydenham’s chorea, which lasted about 1 year.

Cognitive testing also revealed attention and memory (acquisition) deficits. In addition, the patient displayed frontal network dysfunction, which psychologic testing confirmed.

Her physical exam was unremarkable, except for some mild psychomotor activity of the right hand and right side of the face. The usual battery of tests was unrevealing, as were infectious and autoimmune serologies, said Dr. Léger, a neurologist at the University of Montreal.

The clinicians initially diagnosed frontotemporal dementia, but “the presence of a very mild focal examination bothered us just a little bit,” said Dr. Léger. An FDG-PET scan revealed a dramatic increase in metabolism in the left striatum—70% more metabolic activity than in the right striatum.

Hypermetabolic lesions have traditionally been associated with diseases of autoimmunity, Dr. Léger noted. They treated her with a 3-day course of pulsed methylprednisolone sodium succinate, followed by a 2-week prednisone taper.

Within weeks, she noted a resolution of her attention deficits. FDG-PET imaging demonstrated a fairly substantial resolution of the asymmetry of the striatum.

Based on the suspicion that autoimmunity against the basal ganglia was involved, the researchers and post-therapy plasma were sent to the laboratory for analysis.

High titters of antibodies to the striatum—the antibodies found in Sydenham’s chorea—were found in the pretreatment sample, reduced titters were found in the post-treatment sample.

Unlike this case, previous studies involving hypermetabolic lesions have also involved previously diagnosed disorders, such as Sydenham’s chorea, Dr. Léger said.

Neuropsychiatric Medicine 61

March 2007 • www.eclinicalpsychiatrynews.com

In two studies, ischemic stroke incidence was 60% higher in those consuming over 65 g of fat daily.

BY MITCHEL L. ZOLER  Philadelphia Bureau

NEW ORLEANS — For the first time, physicians have evidence independently linking high dietary levels of fat and sodium to an increased risk of ischemic stroke, based on findings from an epidemiologic study in New York.

Experts who heard these findings at the 38th International Stroke Conference hailed them as a key step for changing pub health guidelines in the United States. Until now, “there has not been evidence of a strong association” between dietary fat and sodium, commented Lawrence Brass, M.D., professor of neurology, epidemiology, and public health at Yale University, New Haven.

“These new data will allow us to make a stronger statement,” added Dr. Brass, a member of the American Stroke Association’s committee that writes guidelines for the primary prevention of stroke.

“I can’t speak for the whole committee, but this is the type of information that may prompt us to [look again at] the guidelines,” he said.

A link between fat and sodium stroke “may sound like common sense, but guidelines are based on the available scientific data. Without the data we can’t include them,” said Ralph Sacco, M.D., professor of neurology and epidemiology at Columbia University in New York and senior investigator for the Northern Manhattan Study.

Both studies used data collected in the Northern Manhattan Study, which enrolled 3,298 people who lived in northern Manhattan in 1993. The subjects were thoroughly assessed at baseline, and 3,183 answered a food frequency questionnaire that was used to calculate their average, daily intake of fat and sodium. At entry, this group had an average age of 70 years; 63% were women, 21% were white, 24% were African American, and 32% were Hispanic. They were followed for an average of 5.5 years, during which 142 had an ischemic stroke.

The analysis that examined the link between stroke incidence and dietary intake as a continuous variable and as a dichotomous variable, with 65 g/day as the dividing line between a low- and high-fat diet. Current guidelines, the researchers said, “are based on a strong mechanistic link between dietary fat and stroke. High-fat diets usually lead to increased serum levels of total cholesterol and LDL cholesterol, he said.

“People who eat more than 4 or 4.5 g of fat per day have a slightly increased risk of stroke, compared with those with the lowest intake level, but the difference was not statistically significant. But the analysis controlled for differences in blood pressure, it’s possible that some other, unknown biologic mechanism explains the link between dietary fat and stroke risk. But blood pressure may still play a role, Dr. Sacco said at the conference, sponsored by the American Stroke Association.

“We controlled for differences in blood pressure, but there might have still been smaller differences in blood pressure that we couldn’t detect. We can’t exclude a blood pressure effect,” he said.

The ‘very mild focal examination bothered us just a little bit’ and led to an FDG-PET scan and the discovery of hypermetabolic lesions in the left striatum.

Deep Brain Stimulation May Control Severe Seizures

SAN FRANCISCO — Deep brain stimulation of the anterior thalamic nuclei shows promise in the management of epileptic seizures, judging from the findings of a small pilot study, Robert Fisher, M.D., said at the annual meeting of the Congress of Neurological Surgeons.

If further research results confirm the procedure’s initial promise, deep brain stimulation could become one of a group of targeted therapies, he said.

That group of therapies includes radiation and local drug perfusion, which control severe epileptic seizures, said Dr. Fisher, who is professor of neurology at Stanford (Calif.) University.

The study population consisted of seven men and seven women, 19-44 years old, with partial and apparently generalized tonic-clonic seizures originating in the temporofrontal lobes and from multifocal regions.

Deep brain stimulation efficacy was expressed in terms of responder rate, defined as the percentage of patients who experienced a decline of 50% or more in the number of seizures after deep brain stimulation.

By that criterion, the responder rate was 57% as long as 12 months post procedure, he reported.

Improvement was especially dramatic in four of five patients whose seizures were severe enough to make them fall down.

There was no control group in the study. However, these findings compare favorably with data that come from controlled clinical trials that involved gabapentin and lamotrigine in which responder rates were only 15%-20% among patients who were taking the drugs, Dr. Fisher pointed out.

One concern about deep brain stimulation is that it may inhibit neurologic activity at seizure foci but stimulate activity in surrounding areas of the brain.

“We will have to watch for that as we move forward,” he said.

A controlled, multicenter trial is now underway.

—Norra Macready

Atherosclerosis may be the mechanistic link since high-fat diets usually lead to increased serum levels of total cholesterol and LDL cholesterol.

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