A clinicopathologic study of FTLN, CBD, and PSP

Josephs et al. studied 127 cases of autopsy-confirmed frontotemporal lobar degeneration (FTLD), corticobasal degeneration (CBD), and progressive supranuclear palsy (PSP). They found that the prediction of tau-positive pathology from a CSD or PSP-like presentation was good, while the FTD-MND syndrome predicted motor neuron degeneration (MND). PSP and CBD accounted for most cases classified as progressive non-fluent aphasia. Frontal variant FTD was unpredictable in terms of its biochemistry.

see page 41

MRI in two variants of autopsy-confirmed FTLN

Whitwell et al. studied two variants of tau-negative frontotemporal lobar degeneration and demonstrated a localized pattern of frontal lobe atrophy in cases with motor neuron degeneration, distinct from those without.

see page 102

The editorial by Paul G. Ince and John C. Morris discusses these two articles from the Mayo Clinic which address clinical issues in the diagnosis of disorders in the spectrum of frontotemporal lobar degenerations (FTLD) from the perspective of the underlying pathologies. Both articles take as their starting point autopsy-confirmed cases re-examined with state-of-the-art immunohistochemistry. Two broad categories emerged: tau pathology—Pick's disease, CBD, PSP, multi-system tauopathy, and frontotemporal dementia and parkinsonism linked to chromosome 17; and without tau pathology—FTLD with ubiquitin-only inclusions (FTLD-U), FTLD with motor neuron degeneration (FTLD-MND), and neurofilament inclusion body disease, postulating a theoretical protein as the basis for the ubiquitinated inclusion in amyotrophic lateral sclerosis. They hypothesized that there is a family of clinical syndromes caused by "Gehrigopathy" that exhibit similar phenotypic diversity to those associated with tauopathy and with α -synucleinopathy.

see page 8

Cognitive function in essential tremor

Benito-León et al. evaluated cognition in a population-based sample of essential tremor (ET) cases and controls. Cases performed more poorly on formal neuropsychological testing. Mild cognitive deficits are a feature of mild, largely untreated ET cases.

see page 69

Autologous peripheral blood stem cell transplantation (auto-PBSCT) for POEMS syndrome

POEMS syndrome is associated with overproduction of vascular endothelial growth factor (VEGF) by plasmacytoma. Kuwabara et al. treated four patients with high-dose chemotherapy and peripheral blood stem cell transplantation. Within 6 months, there was clinical improvement with rapid normalization of serum VEGF levels.

see page 105

The editorial by Dyck et al. notes that the POEMS syndrome is an important example of the remote effects of a plasma cell proliferative disorder on neural and non-neural tissues not due to direct invasion by malignant cells. The remote neurologic syndromes sensory neuropathy and cerebellar degeneration are frequently associated with small cell carcinoma of the lung or the myasthenic syndrome of Lambert and Eaton; an antibody-targeted attack on neural cells is the likely mechanism. POEMS syndrome is different: Plasma cells produce a specific cytokine (VEGF) whose target is endothelial cells, which results in a proliferative effect on vessels and functional alterations. It is not known whether all manifestations of POEMS and the shortened life expectancy of this disease result from increased VEGF level or whether life expectancy is altered by control of VEGF levels.

see page 10

Alternating hemiplegia of childhood and topiramate

Di Rosa et al. treated a 12-year-old girl with alternating hemiplegia of childhood with topiramate up to 4 mg/kg/day. Hemiplegic attacks decreased from daily to monthly.

see page 146

Sleep disordered breathing in Chiari malformation

PSG data before and after decompression surgery.

Gagnadoux et al. performed polysomnography in 16 patients with Chiari malformation. They found a high frequency of sleep disordered breathing and noted improvement of central apnea after decompressive surgery.

see page 136

Plasma β -amyloid and white matter disease

Gurol et al. report an independent association between plasma β -amyloid-40 and volume of white matter hyperintensities on MRIs of subjects with Alzheimer disease (AD) or mild cognitive impairment vs 42 with cerebral amyloid angiopathy. The data support β -amyloid as a possible biomarker or risk factor for cerebral microvascular disease.

see page 23

The editorial by Jin-Moo Lee and Hugh S. Markus notes that whether the increased plasma β -amyloid-40 is the cause or the effect of white matter lesions is unclear. However, there is strong evidence of a link between cerebral amyloid angiopathy and white matter diseases and a likely contribution of vascular dysfunction to cognitive decline in AD.

see page 6

NEUROLOGY INVITES "CLINICAL TRIALS RECRUITING" ANNOUNCEMENTS

Neurology now publishes a "Clinical Trials Recruiting" print and online section. This section is prominently displayed on the cover, in the Table of Contents, and on the *Neurology* Web site home page.

Neurology has the widest readership of any neurological journal, and *Neurology* online receives heavy traffic from the public. This online section has open access and is featured on the *Neurology* home page.

Line ads can be placed at no charge and display ads [full or 1/2 page] can also be placed for a fee. The ad must be IRB approved before submission. Contact ctrecruiting@urmc.rochester.edu or click on "Clinical Trials Recruiting" on the home page [www.neurology.org] for more information.

Neurology®

January 10 Highlights

Neurology 2006;66;2-3

DOI 10.1212/01.wnl.0000196862.36013.22

This information is current as of January 9, 2006

Updated Information & Services

including high resolution figures, can be found at:
<http://n.neurology.org/content/66/1/2.full>

Permissions & Licensing

Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/about/about_the_journal#permissions

Reprints

Information about ordering reprints can be found online:
<http://n.neurology.org/subscribers/advertise>

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright . All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.