Multifocal motor neuropathy (MMN) with and without conduction block

Delmont et al. reviewed the clinical features and response to IVIg treatment of 20 patients with MMN vs 13 patients without CB at diagnosis. After a median follow-up of 7 years there were no differences between the two groups in term of age, sex, time from onset to diagnosis, anti-GM1 antibody titers, or CSF data. IVIg benefit was similar in patients without CB (8/13) vs with CB (14/20).

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The editorial by Chaudhry and Swash notes that MMN has a prevalence of 1 to 2/100,000. It was initially recognized as a disorder that could be confused with ALS. If one accepts that CB was adequately excluded in the Delmont et al. cases, the following question is raised: "Is there an axonal form of MMN?" The term multifocal acquired motor axonopathy (MAMA) has been used by other authors to describe such an axonal MMN variant.

A trial of IVIg seems justified in patients with progressive distal asymmetric weakness in a multifocal peripheral nerve distribution with or without CB.

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SSEPs and prognosis in postanoxic coma

Zandbergen et al. found that determination of the N70-response of the somatosensory evoked potentials in addition to N20 may, under certain circumstances, improve the prediction of poor outcome in patients with postanoxic coma.

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The editorial by Thomas P. Bleck notes that the success of cardiopulmonary resuscitation has had the unintended consequence of increasing the number of patients in coma because of hypoxic-ischemic encephalopathy. Prognostication is usually based on physical findings elicited at defined time points after resuscitation. There have been many attempts to improve prognostication with electrophysiologic tests. Studies of the N70 potential in single centers have found the N70 SSEP to have as high a sensitivity as 94% and specificity of 97% for predicting good outcome in patients with preserved N20 potentials. In the Zandbergen et al. multicenter study, local neurophysiologists used equipment already available at each hospital. Here, there was only moderate interobserver agreement on the interpretation of the evoked potentials. In this setting, the addition of the N70 data to the N20 results was not as useful as it appeared to be in earlier investigations.

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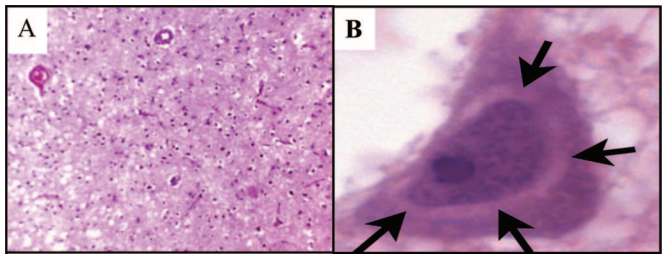
VCP mutations, frontotemporal dementia, Paget disease, and inclusion body myopathy



(A) Diffuse microvacuolization of cortical frontal superficial layers, severely decreased neuronal density, and loss of lamination. (B) Intraneuronal lesions: arrows indicate the perinuclear vacuole.

Guyant-Maréchal et al. found valosin-containing protein gene mutations present in two families in which FTD is the most prominent symptom. The histopathologic study performed in patients harboring the R155C mutation supports the hypothesis that this mutation disrupts normal valosin-containing protein function, leading to diffuse accumulation of ubiquitinated proteins.

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The editorial by Ghetti and Goebel notes that FTD has been found to be linked to at least three chromosomal loci: chromosomes 9, 17, and 3. The condition described by Guyant-Marechal et al. may be considered a member of the growing number of protein aggregate diseases, which includes Alzheimer disease, Parkinson disease, and prion diseases. It is also one of the protein aggregate myopathies, which include the myofibrillar myopathies and the inflammatory and hereditary inclusion body myopathies. Two regions on chromosome 17 have been linked to FTD. Mutations in the MAPT gene, located in chromosome 17q21-22, are associated with FTD. Recently, a dominant form of FTD has been mapped to a pericentromeric region of 12 cM on chromosome 3. Mutations have been found in the charged multivesicular body protein (CHMP2B) gene, which encodes a component of the endosomal sorting complexes required for transport (ESCRTIII complex).

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SAH is followed by temporomesial volume loss: An MRI study

Bendel et al. found that subarachnoid hemorrhage and its treatment may be followed by atrophy in temporomesial structures. Furthermore, the amygdala ipsilateral to the ruptured aneurysm was smaller in patients after surgical than endovascular treatment. A clear correlation was demonstrated between neuropsychological performance and reduced temporomesial volumes.

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