

Acquired copper deficiency in 10 patients with and without Wilson disease

F. Ory-magne³, A. Poujois¹, N. Djebrani-oussedik², P. Chaine¹, F. Woimant¹

¹*Department of Neurology, AP-HP, Lariboisière University Hospital, National Reference Centre for Wilson's disease and other rare copper diseases, France*

²*Toxicology Department, National Reference Centre for Wilson's disease and other rare copper diseases, France*

³*Department of Neurology, Toulouse University Hospital, France*

Background: acquired copper deficiency (ACD) is a rare condition usually diagnosed from haematological changes. Aims: characterize the diagnosis features and evolution of patients with ACD. Methods: clinical, biological and MRI data were analysed at diagnosis and during follow-up. Results: Ten patients with ACD were studied: 3 with Wilson disease (WD-CD) and 7 without WD (CD). Mean time to diagnosis was twice longer in WD-CD than in CD patients. Only one patient with WD-CD was diagnosed on pancytopenia, all other patients had a progressive posterior cord syndrome associated with anaemia and lymphopenia. Electrodiagnostic tests diagnosed a lower limbs sensory neuropathy in six patients. Spinal cord MRI was normal in 3/7 CD patients and in all WD-CD patients. Serum copper, exchangeable copper and urinary copper excretion was low. All WD-CD patients had an iron deficiency associated. A decrease copper intake after bariatric or other digestive surgeries and a chronic use of denture adhesive paste containing zinc were the aetiologies of CD patients. All WD-CD patients were on zinc salts. CD patients received copper supplementation while doses of zinc salts were reduced by two third or stopped in WD-CD patients. Evolution was different between the two groups of patients. Haematological disturbances resolved gradually in two months in CD patients while it took 15 months in WD-CD. Neurological symptoms improved in 7/9 patients after a mean follow-up of two years. Conclusions: anaemia and lymphopenia associated or not to a posterior cord syndrome must raise the possibility of an ACD, even in WD patients.