

Acute copper-deficiency myelopathy in a patient with celiac disease.

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Introduction: Acquired copper deficiency represent a rare cause of progressive myelopathy presenting with sensory ataxia and spastic gait. Commonly, MRI shows increased signal on T2-weighted images involving the dorsal column in the cervico-thoracic cord. Despite frequently associated with a history of gastric surgery, malabsorption syndromes and excessive zinc consumption, cause for copper deficiency remains often unclear. The prevalence of copper deficiency in patients with celiac disease is unknown, representing indeed an uncommon complication of celiac sprue. We report a case of patient who developed subacute myelopathy associated with copper deficiency due to undiagnosed and asymptomatic celiac disease.

Case report: A 62 years old man was admitted with a three weeks history of progressive gait imbalance, tingling and numbness in his feet and ascending lower limbs weakness. Neurological examination showed diffuse sensory ataxia mainly in lower extremities, with the patient unable to stand upright or walk, absent vibration and position sense, distal lower limbs weakness but increased tendon reflexes and bilateral Babinski's sign. Spine MRI revealed T2-weighted and STIR (Short Time Inversion Recovery) hyperintensity involving cervical and dorsal posterior columns of spinal cord (Fig. 1). Neurophysiological studies showed reduction of MEP amplitude, increased central conduction time (CCT) coupled with impaired central sensory conduction in lower limb SSEP (Fig. 2). Normocytic anaemia, very mild vitamin B12 deficiency (112 pg/ml) were present together with undetectable serum copper levels, low serum ceruloplasmin (5 mg/dl) and normal serum zinc (1016 ug/L). In order to search for causes of hypocupremia, upper gastrointestinal endoscopy was performed revealing duodenal villous atrophy consistent with a malabsorption pattern. Antibodies screening for coeliac disease was positive for serum anti-transglutaminase and anti-gliadin antibodies. A gluten-free diet was started associated with intravenous copper repletion prolonged for 7 days which led to progressive clinical improvement and normalization of serum copper levels (474 ug/L).



Fig 1: Spine MRI revealed T2-weighted and STIR hyperintensity involving cervical and dorsal posterior columns of spinal cord without subtentorial abnormalities in Brain MRI.

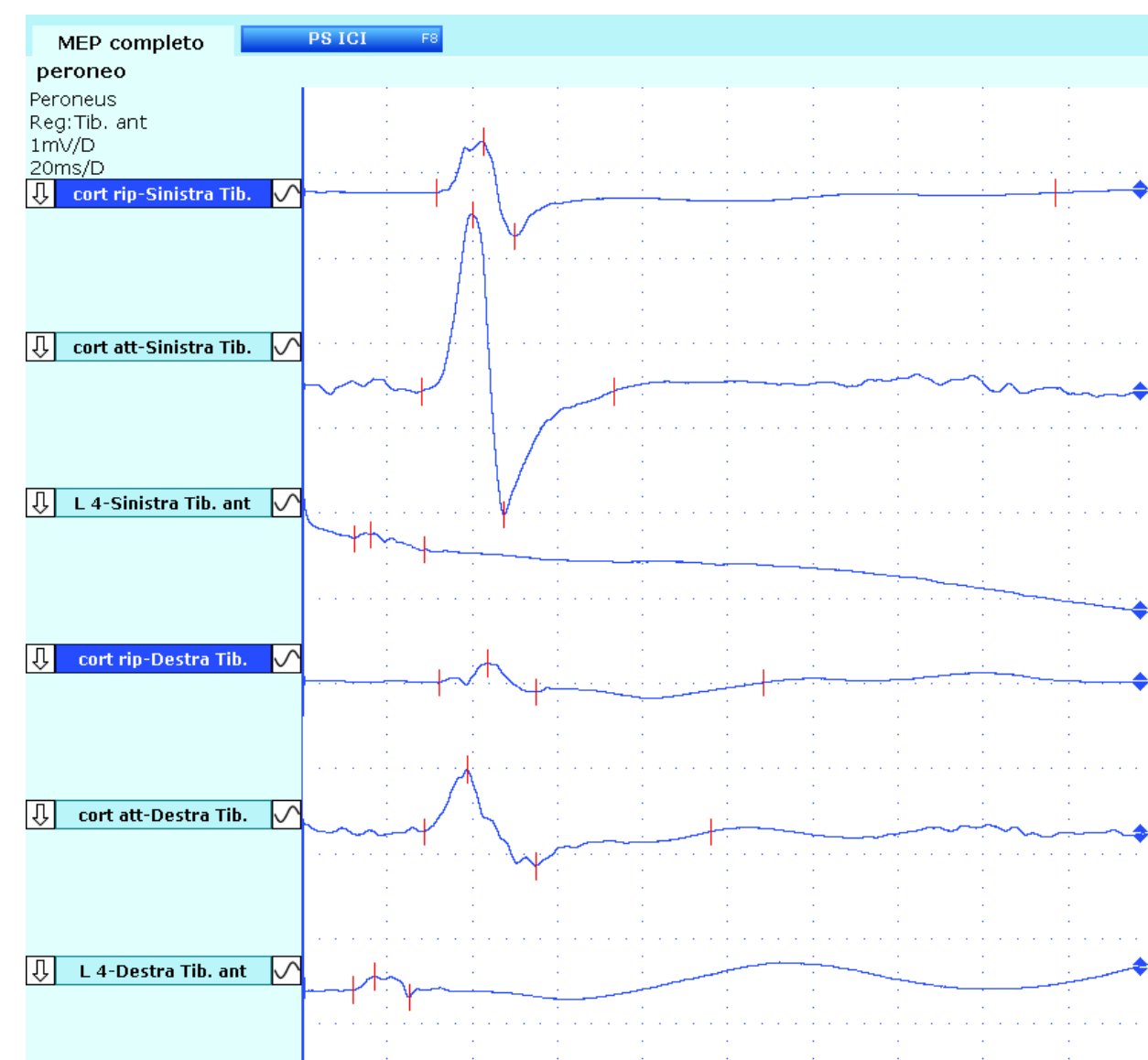
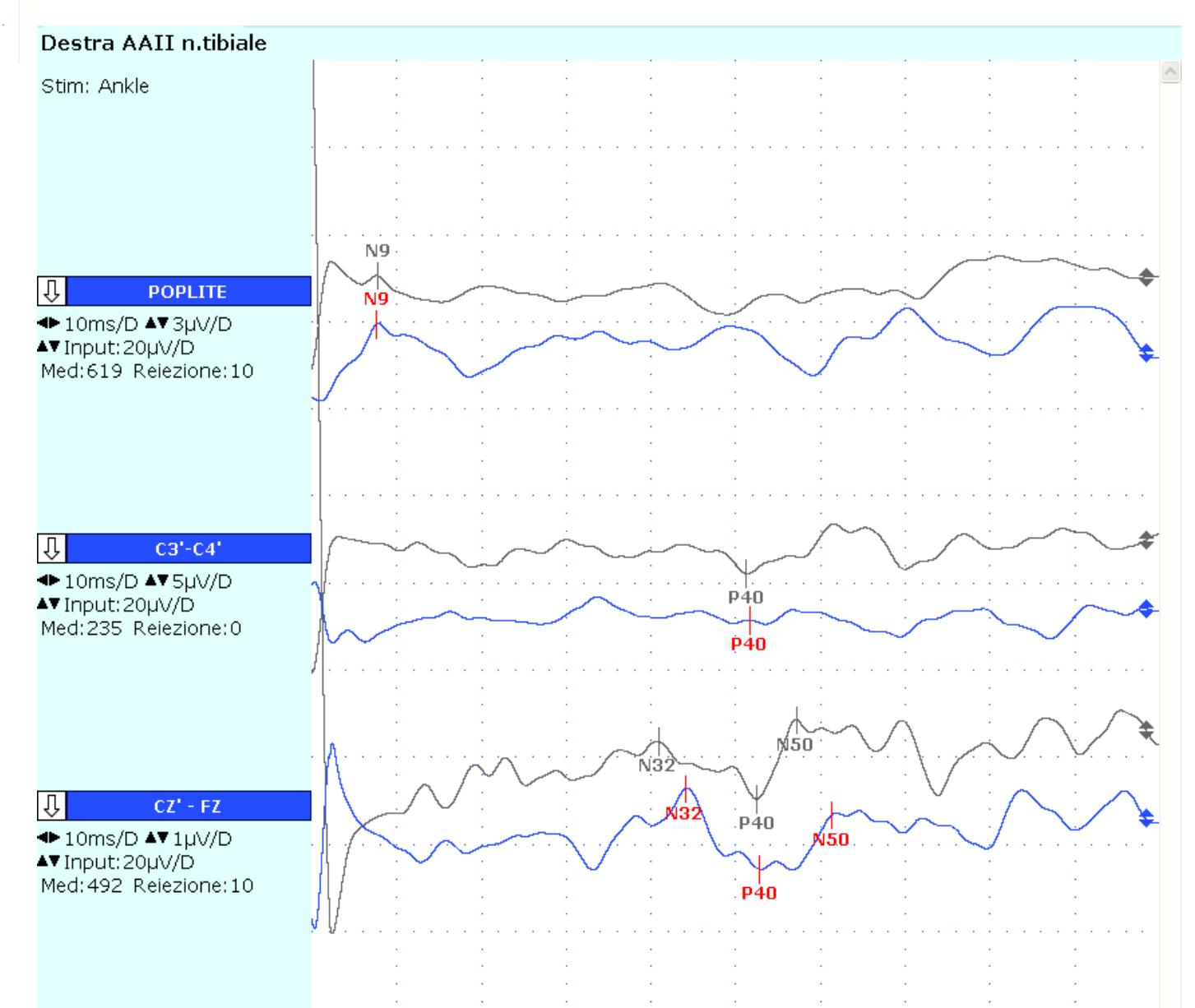


Fig 2: Reduction of MEP amplitude with increased CCT and impaired central sensory conduction in lower limb SSEP.



Discussion: Clinical, neurophysiological and neuroradiological findings in copper deficiency myelopathy may overlap with those of subacute combined degeneration due to vitamin B12 deficiency. However, in our case, neurological symptoms developed very quickly, compared to previously reported cases. A very mild B12 deficiency was considered disproportionate with the neurological impairment and neuroradiological features, suggesting us to search for alternative causes. Indeed, in every patient with subacute or acute history of sensory ataxia and spastic gait, serum copper deficiency should be kept in mind. In case of hypocupremia, antibodies screening for coeliac disease and upper gastrointestinal tract endoscopy should be performed, even in patient without symptoms of coeliac disease.

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