Human Swayback Due to Copper Deficiency

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Citation

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Abstract

A 74-year-old female presented with distal extremity burning paresthesias and gait imbalance of 3 months duration. Her past medical history includes gastrojejunostomy 5 years ago. No history suggestive of malnutrition or alcohol use. Physical examination revealed ataxic myelopathy. Her blood works revealed copper and ceruloplasmin levels, which are significantly reduced, consistent with copper deficiency. CSF analysis was normal. Nerve conduction studies demonstrated symmetric sensory motor axonal polyneuropathy. MRI of the cervical and thoracic spine revealed areas of abnormal, non-enhancing intramedullary T2 and FLAIR sequence hyper intense signal in the posterior cord. Patient was treated with intravenous copper and improved in few weeks. Copper deficiency may result from inadequate intake, malabsorption of any cause, including postgastrectomy, bacterial overgrowth or sprue. Manifestations include: neutropenia, thrombocytopenia and anemia and/or neurological symptoms of large fiber/posterior column and corticospinal tract dysfunction. Differential diagnosis of ataxic myelopathy should include copper deficiency in patients with predisposing conditions.

BACKGROUND

Copper is an essential nutrient in humans, and is readily available in the diet and is rapidly absorbed through the stomach and duodenum into the portal circulation for subsequent uptake by the liver. The known causes of acquired copper deficiency include prior gastric surgery, excessive zinc ingestion, and malabsorption, however often the cause is unclear. Copper deficiency can present with either hematological or neurological manifestations.

CASE PRESENTATION

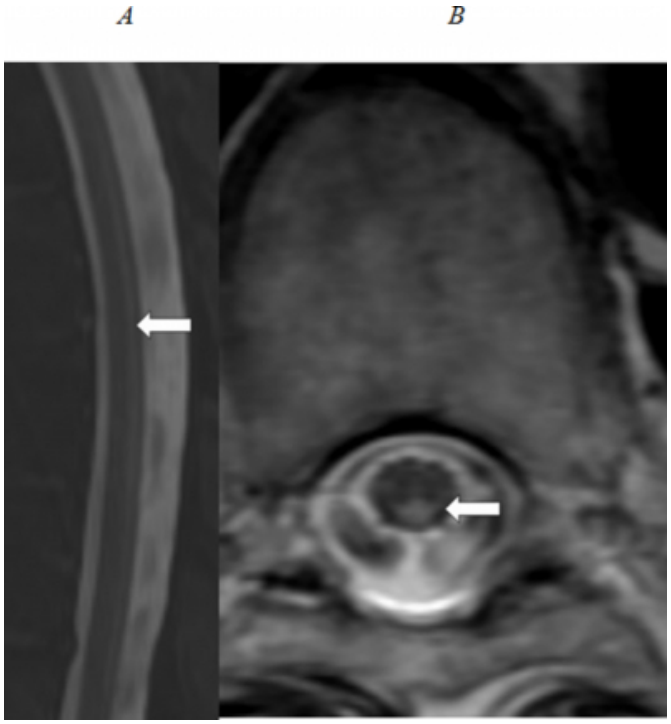
A 74-year-old female presented with distal extremity burning paresthesias, and gait imbalance of 3 months duration. She denied visual changes, bowel or bladder disturbance. Past medical history was notable for gastrojejunostomy 5 years ago. There was no history of alcohol or tobacco use, and family history was non-contributory. Neurologic examination revealed normal mentation and normal cranial nerves. She had moderate distal upper and proximal lower extremity weakness, stocking-glove pattern pinprick loss to the wrists and ankles, distal vibratory and joint position impairment below the ankles. Deep tendon reflexes are exaggerated in all the four limbs; bilateral extensor plantar responses. Gait was broad based and ataxic, with a positive Romberg. Coordination was intact.

Her Complete Blood Count and comprehensive metabolic panel were normal. Her TSH, ESR, ANA, Serum Protien electrophoreses, HIV, Vitamin B12, Folate, Homocysteine,methylmalonic acid, Anti Hu antibodies, RPR, Vitamin E and Zinc were all normal. Nerve conduction studies demonstrated symmetric sensorimotor axonal polyneuropathy. CSF analysis was normal and negative for viral titers. MRI of the cervical and thoracic spine revealed areas of abnormal, non-enhancing intramedullary T2 and FLAIR sequence hyper intense signal in the posterior cord at the C3-5 and T4-9 levels (see figure 1). Serum copper was low at 12mcg/dl (normal 70-155); ceruloplasmin was low at 5mg/dl (normal 18-53).

She received copper sulfate IV (2mg/day) for 1 week, then maintenance oral dose of 3mg daily and within 2 weeks her copper was 83mcg/dl and cerulopasmin 24 mg/dl; a year later they were 153 mcg/dl and 37 mg/dl respectively. Patient showed symptomatic improvement within 2 months.

Figure 1

Figure 1: T2-weighted thoracic MRI demonstrating a high T2 signal lesion (denoted by white arrows) in the dorsal cord in sagittal (A) and axial (B) views



DISCUSSION

Copper deficiency associated myelopathy has been well described in various animal species, commonly referred as "sway back" or enzootic ataxia[2]. More recently, copper deficiency has become recognized as a previously under-diagnosed etiology of myeloneuropathy in humans, with a presentation similar to that of Vitamin B12 deficiency. Copper deficiency may result from inadequate copper intake, or more commonly malabsorption of any cause, including postgastrectomy, bacterial overgrowth or sprue[1]. More recently, cases of secondary hypocupremia due to hyperzincemia have been reported. High zinc levels interfere with copper absorption. The most commonly reported manifestations of copper deficiency include: hematological abnormalities (neutropenia, thrombocytopenia and anemia) and/or neurological symptoms of large fiber/posterior column and corticospinal tract dysfunction[3]. Similar to the case presented in our study, there are previous reports of high T2 signal intensity in the dorsal spinal cord on MRI of patients with copper deficiency, a pattern also noted in subacute combined degeneration from B12 deficiency[4]. The neurological signs and symptoms of copper deficiency may be present without the hematological manifestations[3]. The timing of development of clinical signs and symptoms of copper deficiency following bariatric surgery is not clear, although some studies have suggested that it may take years for total body stores of copper to become depleted [5]. Relapse has been reported on presumably adequate copper supplementation[2] and it has been recommended that serum copper level to be monitored periodically to gauge the therapeutic response to treatment[5]. No studies have addressed the most appropriate dose, route and duration of supplementation. The RDA for adults is 900 mcg per day for both males and females. Prior cases in the literature were treated with oral or IV supplementation equivalent to 1.5–3 mg/day of elemental copper[6, 7]. The recovery of the neurological deficits by copper replacement therapy is quite variable with residual neurological deficits present in many patients [2, 4].

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