



Copper myeloneuropathy

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Abstract

Myeloneuropathy is a frequently encountered condition and often poses a diagnostic challenge¹. Myeloneuropathy arises from a variety of nutritional, toxic, metabolic, infective, inflammatory, and paraneoplastic disorders⁴. Vitamin B12, folic acid, copper, and vitamin E deficiencies may lead to myeloneuropathy resembling as that of subacute combined degeneration of the spinal cord⁶. Chikungunya viral infection has been shown to produce a syndrome similar to myeloneuropathy. Human immunodeficiency virus (HIV) infection may resemble subacute combined degeneration⁹. Magnetic resonance imaging (MRI) in subacute combined degeneration of the spinal cord typically reveals characteristic signal changes on T2-weighted images of the cervical spinal cord. Once myeloneuropathy diagnosis is suspected, all these patients should be subjected to a battery of tests¹¹. Vitamin B12, folic acid, vitamins A, D, E, and K serum levels along with levels of iron, methylmalonic acid, homocysteine, and calcium should be assessed¹³. Clinical features resembling myeloneuropathy along with the battery of biochemical tests often helps in establishing the correct diagnosis.

Keywords: *Myeloneuropathy, Posterolateral syndrome, Demyelination.*

Introduction

Copper deficiency may lead to anaemia, neutropenia and ringed sideroblasts in the bone marrow⁵. Copper deficiency may also lead to myelopathy presenting with a spastic gait ataxia and sensory ataxia. It may also be associated with myopathy, demyelination, peripheral neuropathy⁸. Increase in signal intensity in T2 weighed images that involve the dorsal column is seen in copper myeloneuropathy³. Nerve conduction studies may show varying degrees of peripheral neuropathy picture¹⁵.

Case Report

A 15 -year-old girl came to OPD with complaints of lower limb numbness and gait unsteadiness for 3 months. These symptoms rapidly worsened a week back and she lost the ability to ambulate and stand independently. Bowel, bladder and cognitive functions were normal. The patient's mental status, speech and language were normal. Cranial nerve examination were normal. Her distal lower limb muscles were weak comparatively than her proximal limb muscles. Deep tendon reflexes were brisk in the lower limbs. Joint position sense

was absent in the ankle with complete loss of vibration sense in her lower limbs. Plantar responses were elicited which were found to be extensors. The patient was unable to stand and she was unable to ambulate because of her severe proprioceptive deficits. Diagnostic testing revealed anemia and leucopenia, without thrombocytopenia. Vitamin B12, folate, and methylmalonic acid levels were sent and was found to be normal. Other infectious and inflammatory serologic markers were negative for inflammation. Cerebrospinal fluid examination were found to be normal. Serum copper and ceruloplasmin levels were markedly decreased at 0.05 ng/mL (0.75–1.45 ng/mL) and 1.37 g/mL (22.9–43.1g/mL), respectively. MRI Brain with whole spine screening were normal. The patient was treated with oral copper. Within 1 month of copper supplementation, the patients anaemia resolved completely. When seen 4 months following the initiation of copper supplementation the patient reported feeling stronger. Neurological examination revealed resolution of the patients power in proximal and distal group of muscles.

Discussion

Copper deficiency is an cause of neurologic degeneration and is also an cause of anemia and myelodysplastic syndrome. Copper is an important cofactor in several enzymatic processes important in the function of the central nervous system, including dopamine hydroxylase, cytochrome oxidase, copper zinc super oxide dismutase⁷. Known causes of copper deficiency include malabsorption, excess zinc ingestion, parenteral nutrition without adequate copper and nephrotic syndrome. Menkes disease is an X-linked inherited disorder of copper malabsorption seen in infants with various neurologic and systemic manifestations¹³. Copper malabsorption in ruminant animals is a cause of a progressive ataxic myelopathy, which is called as “swayback.” In “swayback,” necropsy has revealed demyelination in spinal cord white matter. The myelodysplastic syndrome due to copper

deficiency rapidly reverses with copper supplementation. The mechanism of copper deficiency of our patient is not known. The patient had no prior history of gastric surgery, denied excess ingestion of zinc and did not have any other symptoms to suggest generalized malabsorption syndrome¹¹. In the absence of any other obvious mechanism, copper deficiency may be due to a defect in the luminal copper transport or trafficking of copper and zinc. Increase in T2 signal intensity in the dorsal columns in will be seen with copper deficiency myelopathy². Other than subacute combined degeneration of the cord, the differential diagnostic possibilities for abnormal increased T2 signal intensity in the dorsal cord include HIV myelopathy, Herpes viral infection, Demyelinating disorders (eg, multiple sclerosis). Demyelinating disorders like multi[le sclerosis does not involve greater than two vertebral bodies. HIV and Herpes results in expansion of the cord and enhancement.

Treatment

Treatment of copper myeloneuropathy is supplementation of oral copper (2-4 mg/day) for 4-12 weeks⁵. Treatment results in significant improvement in power and patient will be symptomatically better.

Conclusion

This type of cases are very rare in our country. Hence these patients must be counseled and reassured. The overall goal of therapy in Copper myeloneuropathy is to control symptoms and to maintain function. Target medical therapy to arrest the extent of disease activity. Hence, treatment should be started early to prevent further neurological damage,

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