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Case Report
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Subacute Spinal Cord Degeneration Presented with Cerebellar Tiger Stripes: A Case Report

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Abstract

Background: Subacute combined spinal cord degeneration is a nervous system degenerative disorder caused by vitamin B12 deficiency in the body, typically involving the posterior and lateral cords of the spinal cord, as well as peripheral nerves, optic nerves, and brain, but is uncommon in patients with cerebellar tiger stripes and increased cerebrospinal fluid protein.

Case Report: A 47-year-old man presented with progressive gait disturbance, tingling of hands and feet, and progressive memory impairment for 4 weeks. T1-Weighted Imaging (T1WI) showed a hypointense signal with stripes and other slightly hypointense signals. T2-Weighted Imaging (T2WI), Fluid Attenuated Inversion Recovery (FLAIR) showed an overall hyperintense signal with stripes and other hypointense signals (tiger stripes sign). The cervical spinal cord showed signal changes in the posterior column on T2-weighted MRI scans. After three weeks of vitamin B12 therapy, the patient's clinical symptoms, signs, imaging abnormalities, and elevated cerebrospinal fluid proteins recovered. To facilitate early identification and treatment, we reported a case of spinal cord injury combined with a typical "tiger stripes" sign due to vitamin B12 deficiency to achieve a good prognosis.

Conclusion: We present a case of spinal cord injury combined with a typical "tiger stripes" sign and increased cerebrospinal fluid protein due to vitamin B12 insufficiency. Despite its rarity, vitamin B12 deficiency needs to be considered as one of the etiologies in patients with clinically manifest cerebellar ataxia for early diagnosis and prompt treatment to improve patient outcomes.

Keywords: B12 vitamin deficiency; Tiger stripes; Cerebellum; Combined degeneration of the subacute

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Background

Subacute combined degeneration of the spinal cord is a disorder of the central and peripheral nervous system caused by insufficient intake, absorption, fixation, transportation, or metabolism of vitamin B12. Most of the lesions involve posterior cord, lateral cord, and peripheral spinal cord nerves [1]. Here we report a case of a 47-year-old male with predominant cerebellar signs, impaired proprioceptive sensation with correlated imaging in a case with hyperhomocysteinemia and vitamin B12 deficiency. In the meantime, Cerebrospinal Fluid (CSF) examination revealed increased protein levels and reversed after therapy. Till now, reports focus on hyperhomocysteinemia and vitamin B12 deficiency in cerebellum and spinal cord are limited. Moreover, there is no report concentrating on the reversion of CSF protein levels consistent with treatment outcomes.

Case Presentation

A 47-year-old man was admitted to the Neurology Department with progressive gait disturbance, tingling of hands and feet, and progressive memory impairment for 4 months. For 2 years, he presented with intermittent nausea, increased abdominal bloating, and anorexia. Second, he underwent Esophagogastroduodenoscopy 2 years before the paroxysmal crisis in another hospital, which revealed a morphologic change in gastric folds consistent with erosive gastritis. There was no standardized and regular treatment focus on erosive gastritis. A year later, after a poor appetite, his weight dropped by approximately 15 KG. He had difficulty ambulating independently 4 months ago, intermittent tingling in both hands and feet, and progressive memory impairment. As his symptoms worsened, he was unable to walk on his own in the dark and fell several times.

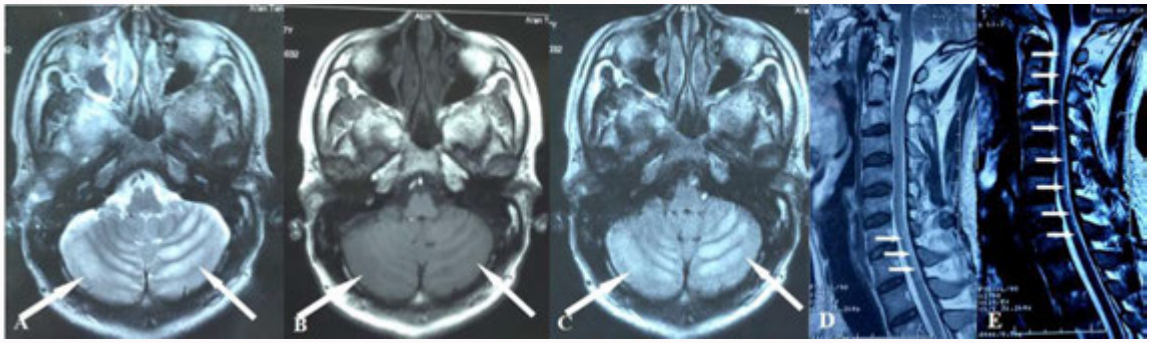

Figure 1: A. T2-weighted imaging (white arrow, tiger striped sign), B. T1-weighted imaging (white arrow, tiger striped sign), C. FLAIR (white arrow, tiger striped sign), D, E. T2-Weighted Imaging (T2WI) showed an hyperintense signal in the posterior column of the cervical spinal cord, lesions progress over time. (D 2016-08-27, E 2016-08-30).

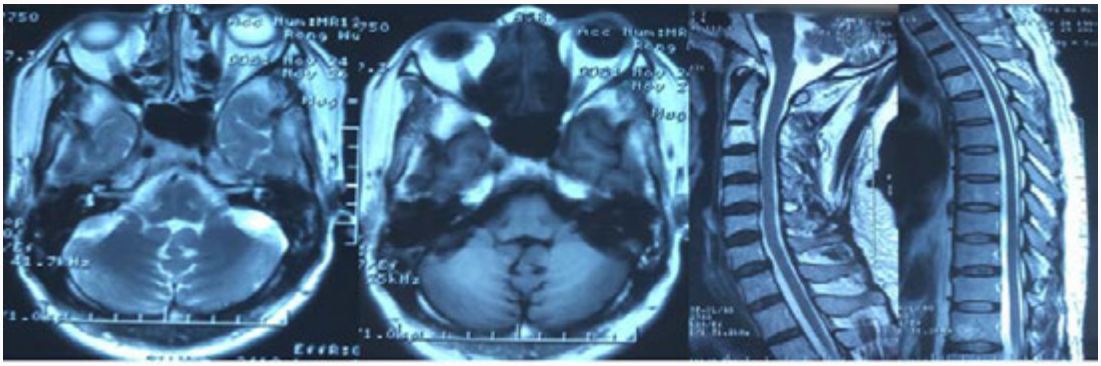

Figure 2: Magnetic Resonance Imaging (MRI) images shows the reversion of bilateral patchy hyperintensities in the cerebellum and cervical spinal cord after treatment.

The patient did not improve significantly after visiting a local clinic and receiving experimental treatment with oral vitamin B 12 and folic acid intake.

Physical examination revealed he was conscious with no memory impairment whose advanced cortical function was assessed by Mini-Mental State Examination (MMSE) (MMSE score 29). His vital signs were normal, cardiopulmonary findings were normal, and 4 out of 5 strengths were symmetrical in the extensor muscles of the lower extremities. In addition, he had features of ataxia, decreased vibration, proprioception, and algesia in both lower extremities. Deep tendon reflexes of arms, knee, and ankle were reduced with a positive Chaddock's sign on both sides.

Investigations in blood suggested macrocytic anemia, elevated serum B12, and higher folate levels detected (>2000 pg/mL), elevated serum homocysteine (66.7 μmol/L), nonreactive serology for HIV and syphilis, normal thyroid, renal, and liver function tests, tumor markers, and negative rheumatoid antibody. The CSF scan revealed an increase in protein level (671 mg/L) with normal cell count and differential concentration of glucose and chloride.

T1-Weighted Imaging (T1WI) showed a hypointense signal with stripes and other slightly hypo intense signals. T2-Weighted Imaging (T2WI), Fluid Attenuated Inversion Recovery (FLAIR) showed an overall hyperintense signal with stripes and other hypointense signals (tiger striped sign) (Figure 1). On T2 weighted MRI, the cervical spinal cord showed signal changes in the posterior column. Lesions develop over time (Figure 2).

Spikes Evoked Somatosensory (SEPs) and extremity electromyography suggests multiple motor and sensory nerve injuries and impaired central afferent fibers, responsible for ataxia due to sensory ataxias and cerebellar damage.

The patient was treated with intramuscular vitamin B12 (500 mg daily) for 2 weeks, then followed by intramuscular 500 mg vitamin B12 once two weeks for 8 weeks. Patients had anemia, radiological signal (Figure 2), EMG findings, and clinical symptoms resolved within 3 months of therapy. According to the combination of gastritis, encephalopathy, myelopathy, peripheral neuropathy, and prompt vitamin B12 substitution effect, Subacute Combined Degeneration (SCD) diagnosis was established.

Discussion and Conclusion

Poor dietary vitamin intake and malabsorption result in Vitamin B12 deficiency and deficiency of folic acid. B12 deficiency is caused by a low intake, gastrectomy, pernicious anemia, end ileal disorders and probably secondary to cobalamin II trans deficiency [2]. Here in our case the patient had erosive gastritis without standardized therapy for years and rapid weight loss implying low vitamin intake. Deficiency of vitamin B12 and folic acid is the most common reason for the Hyperhomocysteinemia required (Hhcy) because it is an essential cofactor in the demethylation of Hcy to methionine and the trans-sulphuration process [3].

Vitamin B12 deficiency mediates swelling of the myelin sheath initially followed by axonal switching that typically commences in the thoracic cord. This affected site may extend to other levels with myelin sheath and axonal degeneration leading to Wallerian type degeneration [4]. The dorsal and lateral spinal cord columns and

periventricular white patterns demyelinating the brain are classic MRI findings, due to the lowest carboxymethyl transferase protein activity in the cerebellum and the highest in cortical white matter compared to other brain regions [5]. For this reason, limited reports have been published that focus on vitamin B12 deficiency leading to

isolated cerebellar involvement in both adult and pediatric populations [6,7]. The cerebellar cortex clearly divides into three layers from the outer to the inner layer: the molecular layer, the Purkinje cell layer (ganglion cell layer) and the granule layer [8]. The formation of MRI signal of tiger stripe sign is related to cerebellar white matter atrophy, thickening of granular cell layer and molecular layer [9]. In our case, the patient was treated with methylcobalamin for 4 months following the initiation of symptoms and ideally recovered clinically and radiologically. The value confirmed the relationship between the essentials of early treatment and the effect of therapy.

The elevated protein in CSF and its reversion consistent with decreased homocysteine and clinical symptoms have also not been reported previously. It is well known that there is accumulating evidence to support the postulate and that its spontaneous oxidation, HCA, is neurotoxic both directly and indirectly [10]. Hhcy pathway mediates endothelial cell injury and disruption of blood brain barrier [11]. And, also hay, which is considered as a pro-inflammatory molecule, activated several cytokines and induced oxidative stress in brain and vascular tissues as well. They are the structural basis for the high protein levels in the CSF. Hhcy has also been reported to be an induction of apoptotic and necrotic cell death [12], mediating apoptosis *via* activation of neuronal death *via* the pro-apoptotic pathway and overstimulating excitotoxic effects in glutamate receptor expressing cells. Hhcy also induces DNA damage and enhances the generation of Reactive Oxygen Species (ROS) in cells [10], which is also a major component that precipitated apoptosis. In both vascular dysfunction and neuronal necrosis, protein levels may be elevated, which requires further investigation.

In conclusion, we present a case of spinal cord injury combined with a typical "tiger stripes" sign and increased cerebrospinal fluid protein due to vitamin B12 insufficiency. Despite its rarity, vitamin B12 deficiency needs to be considered as one of the etiologies in patients with clinically manifest cerebellar ataxia for early diagnosis and prompt treatment to improve patient outcomes [12]. Vitamin B12 is a major risk factor for developing ataxias.

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