Case Report

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# Early treatment of subacute combined degeneration of the spinal cord in pernicious anemia will improve the outcome

## Abstract

Vitamin B12 deficiency is probably more common than what is reported in the literature. B12 is an essential vitamin, and its deficiency can lead to neuropsychiatric manifestations such as peripheral neuropathy, myelopathy, cerebellar ataxia, dementia, delirium, even psychosis and mood disorders. We report a case of 45 year old polish male presented with symptoms and signs suggestive of SCD of the spinal cord, due to severe vitamin B12 deficiency as a result of pernicious anemia. Cervical spine MRI showed enhancement in the bilateral posterior columns of the cervical spinal cord with a classic inverted V appearance. Metabolic workup revealed severe vitamin B12 deficiency, complete blood analysis indicated megaloblastic anemia. Intramuscular injection of cobalamin was started on a timely manner with complete resolution of symptoms.

**Keywords:** vitamin B12 deficiency, subacute combined degeneration, pernicious anemia, malabsorption

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## Introduction

Subacute combined degeneration (SCD) is characterized by demyelination of the dorsal and lateral spinal cord, the degeneration as a result of vitamin B12 deficiency which is the most common cause. It is usually associated with autoimmune gastritis or pernicious anemia[1] but can also be caused by other nutritional deficiencies like vitamin E deficiency or copper deficiency.

## Case presentation

45 year old Polish male patient with a past medical history of hyperthyroidism who presented to the emergency room complaining of two weeks history of upper and lower extremity numbness and stiffness involving his palms up to the arms and his feet. He reported fatigue and muscular pain all over his body for the last two months, and yellowish discoloration of his eyes that noticed few days ago. He denied any dietary restrictions. Physical examination revealed impaired position sense, impaired vibration sense and 'glove and stocking' peripheral paresthesia.

Laboratory findings were significant for macrocytic anemia (with hemoglobin of 9, MCV of 109), indirect hyperbilirubinemia 5.1, high LDH (1630), high retic count (3.7 %) and low level of vitamin B12 (159). Antibodies profile showed positive anti intrinsic factor antibody, high thyroid peroxidase antibody (270) and high thyroglobulin antibody.[2] Cervical Spine MRI showed heterogeneous T2 hyperintensity with scattered enhancement in the bilateral posterior dorsal columns of the cervical cord with an inverted "V" appearance extending from C2-T1 as shown in Figure 1 & 2. Thoracic cord MRI demonstrates mild posterior column hyperintensity without enhancement.

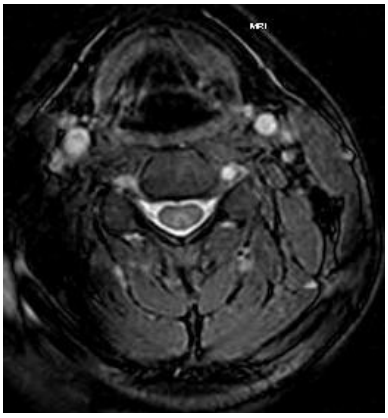

**Figure 1** Cervical MRI w/wo contrast, showing T2 hyperintensity with scattered enhancement in the bilateral posterior dorsal columns of the cervical cord.

MRI T2 Axial demonstrating homogeneous hyperintense signal changes of the dorsal columns and the corticospinal tract due to vitamin B12 deficiency.

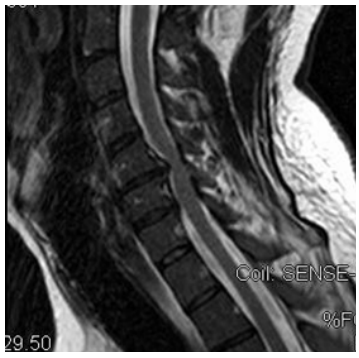


**Figure 2** T2 Sagittal MRI demonstrating hyperintense signal changes.

## Discussion

### Pathophysiology

At the gut level, cobalamine is bound to intrinsic factor released in the stomach, a glycoprotein produced by the parietal cells of the

stomach. The cobalamine-intrinsic factor complex is transported to the terminal ileum, where it binds to receptors on the brush border of enterocytes and is absorbed. The common setting of vitamin B12 deficiency is pernicious anemia, an autoimmune disorder caused by antibodies against gastric parietal cells and the intrinsic factor.[1] Vitamin B12 deficiency may also result from Helicobacter pylori gastritis, surgical resection, tumors, and other conditions involving large parts of the stomach or the lower ileum.[3,4]

### Clinical presentation and natural progression

B12 deficiency can present with a wide range of symptoms and signs; hematological and neurological resulting in megaloblastic anemia, motor-sensory deficits with peripheral neuropathy and myelopathy being the most common,[4] in addition to autonomic nervous system dysfunction. Hemolysis and pancytopenia could be detected as well.[5]

Pernicious anemia can present with severe hematologic conditions and even neuropsychiatric illness known as megaloblastic madness, the severity of neuropsychiatric involvement is inversely related to the hematological manifestations and bone marrow suppression.[6] This neuropsychiatric sequela can range from delusions, mania, paranoia and even psychosis.[7]

### Diagnosis

Low vitamin B12 levels in the serum warrants measurement of both methylmalonic acid and homocysteine to confirm vitamin B12 deficiency. Methylmalonic acid is more sensitive and specific for the diagnosis if found elevated. Serum B12 measurement has both false negative and false positive values that are in common in almost 50% of cases, due to the fact that B12 is highly protein bound.[1] Hematological derangements with anemia and macrocytosis are usually present also.[6] It is advised to consider early spinal MRI imaging due to its high value in early identification, diagnosis and treatment follow up of the disease.[8,9]

### Treatment

In patients with severe neurological abnormalities; Intramuscular injection of vitamin B12 1000 µg multiple times per week for 1 to 2 weeks, then weekly until clear clinical improvement, followed by monthly injections.[1] Lifelong therapy with vitamin B12 is indicated for all patients with pernicious anemia or malabsorption. Stopping treatment will result in recurring symptoms within a short period of time. After replacement with vitamin B12 supplements, follow up with MRI of the spine correlatedvery well with the clinical outcomes for patients with B12 deficiency.[8,10–13]

## Conclusion

Our patient has neurologic abnormalities that are consistent with vitamin B12 deficiency, which was confirmed with a low level of vitamin B12. In the absence of dietary restriction or a known cause of malabsorption, further evaluation is warranted; testing for pernicious anemia (anti– intrinsic factor antibodies) was positive. Parenteral vitamin B12 treatment (8 to 10 loading injections of 1000 µg each, followed by monthly 1000 µg injection) is an effective therapy.

Effective vitamin replacement will correct blood counts in two months and correct or improve neurologic signs and symptoms within six months. After three months of parenteral injections the patient reported significant improvement in his symptoms, with almost complete resolution in six months. Early identification of vitamin B12 deficiency and prompt diagnosis of SCD could avoid any irreversible neurologic damages and prevent disability by early parenteral vitamin B12 treatment.

## Acknowledgments

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## Conflicts of interest

The author declares that there is no conflict of interest.

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