Subacute Combined Degeneration Of The Cord

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Citation


Abstract

Vitamin B<sub>12</sub> deficiency may be the consequence of several pathological conditions. The most frequent neurological manifestation is the subacute combined degeneration (SCD) of the spinal cord and polyneuropathy, rarely dementia and damage of the optic nerve occur. Numbness of the limbs and trunk is an early symptom; weakness, clumsiness and spasticity, abnormal reflexes, gait ataxia develop later. MRI reveals hyperintensities involving dorsal columns of spinal cord in cervical and upper dorsal region. Timely parenteral Vit B12 can result in complete resolution clinically as well as radiologically.

INTRODUCTION

Subacute combined degeneration of spinal cord is a potentially reversible cause of neuropathy if timely diagnosis and appropriate management is provided. This specific type of neuronal degeneration results from Vitamin B<sub>12</sub> deficiency. There is specific predilection for involvement of dorsal columns, lateral corticospinal and lateral spinothalamic tracts of spinal cord. Patients usually complains of insidious onset of sensory symptoms followed by involvement of motor involvement which may progress to paraplegia in neglected cases. The posterior column involvement is very well demonstrated by Magnetic Resonance Imaging (MRI) which shows hyperintense signal intensity in the posterior part of spinal cord. Cervical spinal cord is usually involved, however thoracic spinal cord may be involved in some cases.

CASE REPORT

A 72 year old strict vegetarian female presented in the out patient department with complaints of paresthesias involving both upper limbs followed by lower limbs for the past 2-3 months with mild abnormality of the gait. It was followed by motor weakness of all four limbs. Patient was non-diabetic, there was no history of trauma or fever. Neurological examination revealed numbness in both lower and upper limbs. Proprioception, vibration and position senses were minimally disturbed. Mild motor weakness in upper limbs was noted. All reflexes were brisk in both the legs and arms but plantars were flexor. Nerve conduction test and electromyography were within normal limits. Hematological examination revealed normal hemoglobin level(12gm), mean corpuscular volume was elevated. Serum Vitmin B<sub>12</sub> level were low (75 pg/ml ) whereas serum folate level were elevated (25ng/ml). Gastric endoscopy was normal with no evidence of atrophic gastritis. Cerebrospinal fluid (CSF) examination does not reveal any abnormality.

Diagnosis of subacute combined degeneration of cord was considered and patient was reffered for MRI. MRI of spine was performed on 1.5 Tesla Magnet which revealed hyperintense signal in the cervical cord in the posterior columns extending from C2 to C6 level. No signal abnormality was seen on T1 weighted images. Cord was not expanded. No evidence of any enhancement was seen on post gadolinium images. No evidence of any neural compression was seen. The MRI and laboratory findings were diagnostic of subacute combined degeneration of cord. Vitamin B<sub>12</sub> was administered (1000 µg/day for five consecutive days, intramuscularly). Within 3 weeks the gait improved, ataxia, weakness and numbness of the limbs decreased.
**Figure 1**
Figure 1: T2 weighted sagittal image of cervical spine shows hyperintense signal intensity involving the dorsal columns of spinal cord (arrows). The abnormal intensity is extending from C2 to C6 level. The cord is not expanded and visualised dorsal spinal cord shows normal signal. No evidence of any abnormal compression on cord is seen.

**Figure 2**
Figure 2: T2 weighted axial image reveal symmetrical hyperintensities involving posterior columns in the spinal cord. CSF is seen all around the cord with no evidence of any compression on spinal cord.

**DISCUSSION**
Subacute combined degeneration of the spinal cord is the most common neurological manifestation of Vitamin B₁₂ deficiency. Pernicious anemia with atrophic gastritis is the most common cause of its deficiency in the western countries, however, in India, alcoholism, malnutrition and ileocecal tuberculosis are the common causes. Hematologic, gastrointestinal and neurological manifestations are also common. The diagnosis of B12 deficiency is made by low serum B₁₂ level, or if the B₁₂ level is borderline, elevated levels of the metabolites homocysteine and methylmalonic acid. The hematologic changes, most notably megaloblastic anemia, are not reliable markers for B12 deficiency. More than one quarter of patients with neurologic syndromes will have a normal complete blood count.

Decreased B12 levels impair function of Methionine Synthetase and Methylmalonyl CoA mutase. This leads to production of abnormal fatty acids and elevated levels of Methylmalonic acid, which are toxic to myelin. Swelling of myelin sheaths is followed by astrocytic gliosis.

The neurologic manifestations of vitamin B12 deficiency are the result of its effects on the brain, optic nerves, peripheral nerves, and spinal cord. Patients with subacute combined degeneration present with paresthesias, which may progress to sensory deficits, gait disturbances, ataxia,
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and muscle weakness especially in the legs. If left untreated it may progress to ataxic paraplegia. Neurological findings include numbness, loss of vibratoty and joint position sense, sense of proprioception, hyperreflexia, spasticity, weakness and extensor plantar responses. Myelopathy is usually diffuse and signs are asymmetric. Loss of reflexes due to associated peripheral neuropathy may be seen. Optic atrophy and disturbed mental function (megaloblastic madness) may also be associated. Visual disturbance due to optic neuritis may also be seen.

Histologically, multifocal demyelination and vacuolization may be found in the posterior, lateral, and, occasionally, anterior columns. Axonal degeneration and, ultimately, axonal death is characteristic. The progression of demyelination is often from dorsal columns to lateral columns, with subsequent Wallerian degeneration.

MRI findings reveal increased signal involving the posterior columns of cervical and thoracic cord. Minimal cord expansion and enhancement can be seen in some cases. Lateral column involvement on MR imaging is usually not apparent. Although these findings are consistent with those of subacute combined degeneration, they are nonspecific. MR imaging is useful in distinguishing the different causes of intramedullary myelopathy. Differential diagnosis is wide including multiple sclerosis (MS), inflammatory disorders (sarcoidosis), infections (HIV & herpes simplex), ischemia and neoplasms (astrocytomas and ependymomas). Presence of long segments of hyperintensities involving posterior columns along with characteristic history and laboratory findings clinches the diagnosis. Timely diagnosis and treatment can prevent irreversible neurological damage and MR can help in monitoring the response to treatment.

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