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Case Report

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Sub-acute combined degeneration of the spinal cord - Inverted ' V' sign a clue to avoid morbidity.

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Abstract

SCD(Subacute combined degeneration) also known as Lichtheim's disease, refers to degeneration of the posterior and lateral columns of the spinal cord due to various causes - vitamin B12 deficiency (most common), copper deficiency or vitamin E deficiency. The clinical manifestation of this entity lagging behind spinal Magnetic resonance (MR) imaging, demands MRI to be used as a screening tool. However it is not possible to use a costly screening tool in a large scale and for a entity which is not a frequent clinical encounter. Hence early imaging diagnosis has a useful role, when incidentally detected thus allowing the clinician to intervene early using this clinic-radiological lag time to benefit the patient and reduce morbidity and by any luck completely prevent it. Inverted V sign / Rabbit ear sign is classical MR imaging feature for SCD and the appearance is due to selective involvement of the dorsal columns and progressive involvement of lateral and anterior column of the cord. However always complete Inverted V sign may not be seen in all cases with few variants as seen in our 3 cases, of which 2 patients were asymptomatic. Various Biochemical investigations were performed which confirmed the MR imaging findings of SCD in all three cases. Two asymptomatic patients were followed up post treatment(Vitamin B12 injections) after 2 months with Axial T2 screening of the cervical cord which showed complete resolution in the form of absent inverted v sign seen earlier.

Keywords: SCD, Vitamin B12 deficiency, Magnetic resonance imaging (MRI), inverted V sign.

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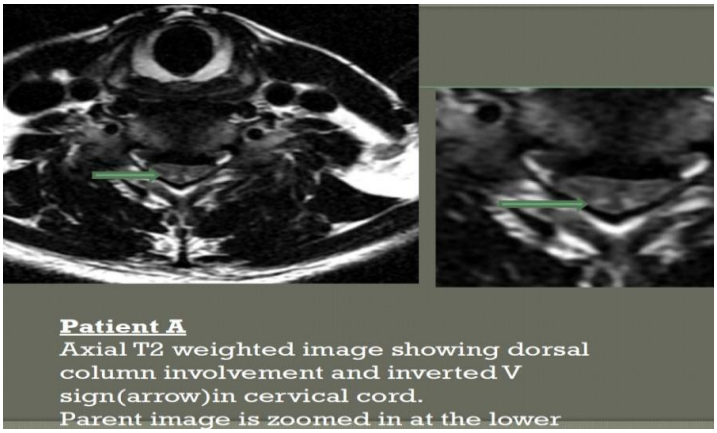
INTRODUCTION

Vitamin B12 deficiency causes a wide range of hematological, gastrointestinal and psychiatric manifestations. Myelopathy, neuropathy, dementia, behavioral changes, and optic nerve involvement are common neurologic manifestations of vitamin B12 deficiency. Most common causes of vitamin B12 deficiency include pernicious anemia, gastrectomy, intestinal parasitic infections, tropical sprue, cystic fibrosis, medications and increased requirement (in hyperthyroidism and alpha thalassemia), and inadequate intake with food (e.g., vegetarianism and veganism). The spinal cord changes caused by vitamin B12 deficiency are known as subacute combined degeneration (SCD).

Case presentations:

We have analyzed and retrospectively compared our findings in 3 cases that were referred to department of radio diagnosis by different departments at SSIMS & RC medical college. Middle aged male patients (age group 40 – 50yrs), with only one patient presented with varying sensory symptoms like paraesthesia & loss of joint position. Other two patients were asymptomatic and were referred for lumbar spondylosis and were detected incidentally. Asymptomatic patients were reexamined retrospectively by the concerned doctors which elicited no obvious findings.



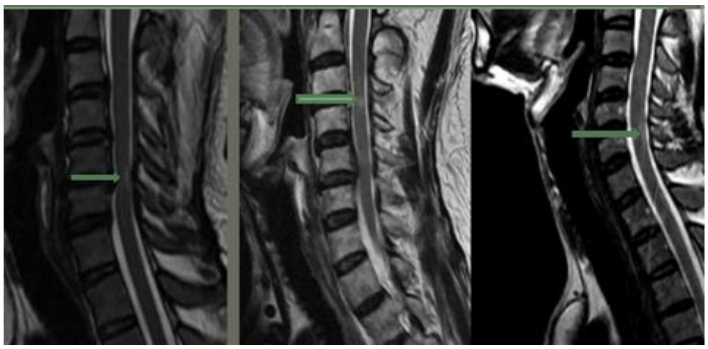
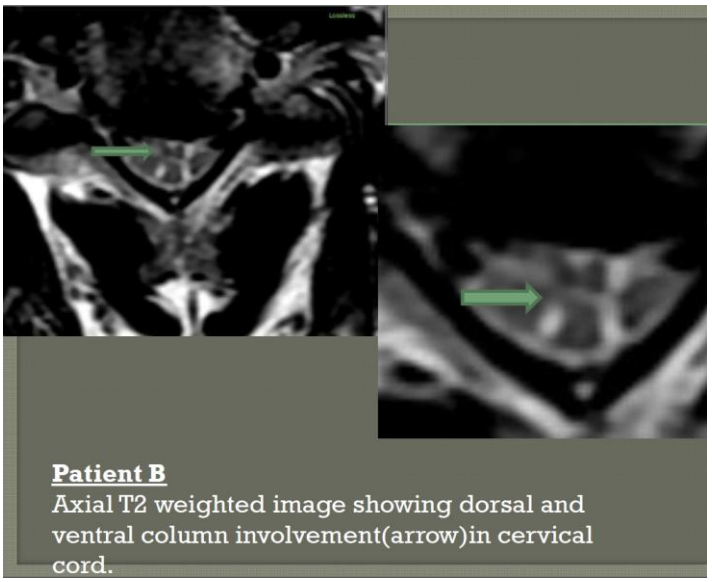


Figure 1. A – Axial T2 images showing hyperintense signal at the dorsal column of the cervical cord. B – Sagittal images of the cervical cord in different patients showing short segment posterior column high signal.



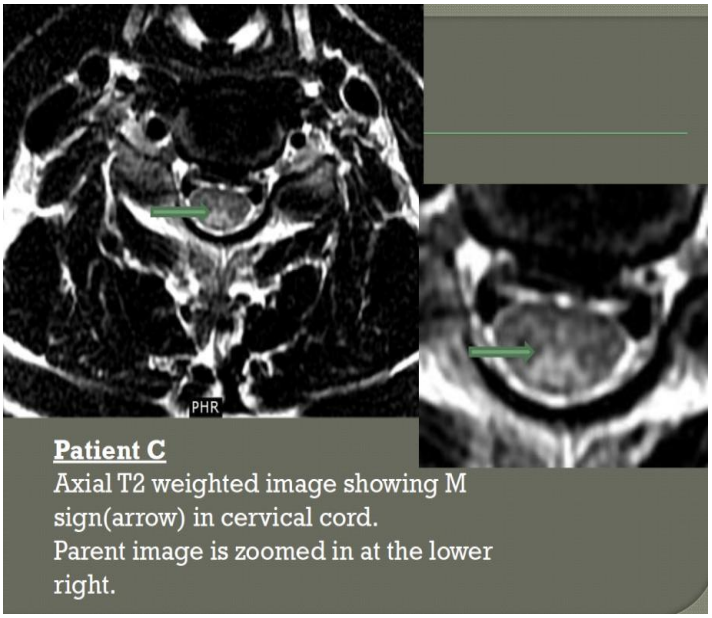


Figure 2.Axial T2 images showing hyperintense signal at the dorsal column of the cervical cord

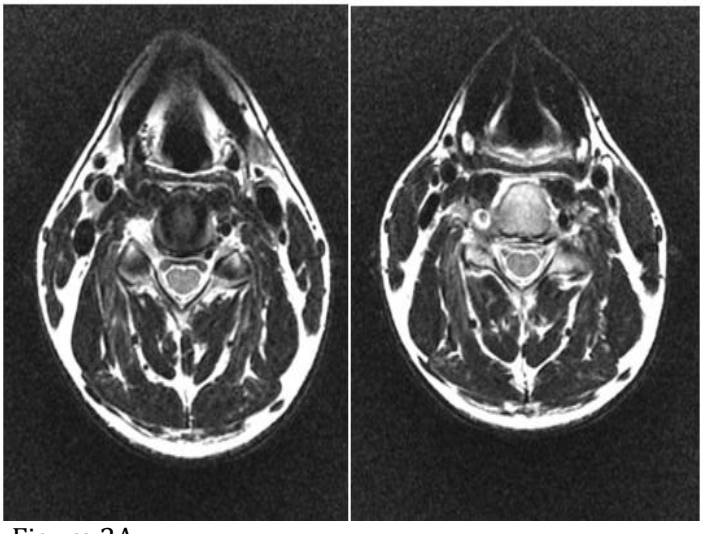


Figure 3A.

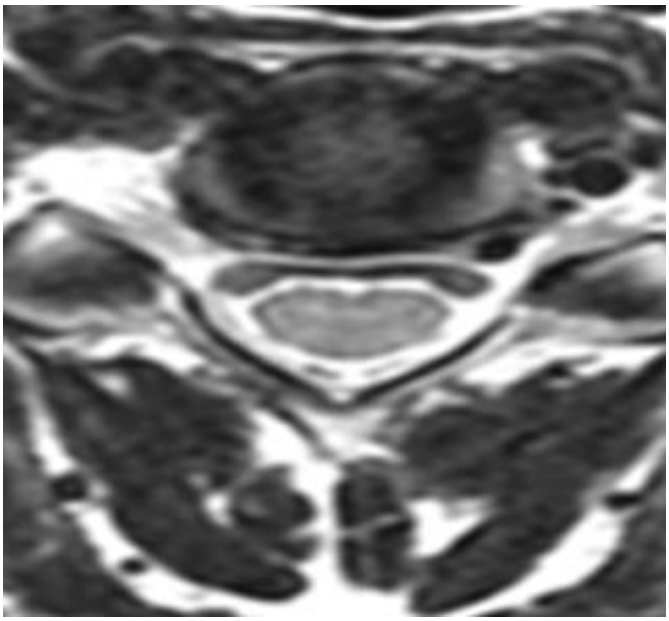


Figure 3. B.

Figure 3. A & B -Post treatment resolution of hyperintense signal in the cervical cord.B - Zoomed image.

Patients were followed up with imaging in the two asymptomatic cases. However the other symptomatic case could not be followed up on imaging due to financial restrains of the patient, but showed significant improvement symptomatically.

Discussion& results:-

Sub-acute combined degeneration of spinal cord, also known as Lichtheim's disease[1], refers to degeneration of the posterior and lateral columns of the spinal cord as a result of vitamin B12 deficiency (most common), copper deficiency[2] or vitamin E deficiency.

The myelopathy of vitamin B12 deficiency (or SCD) is characterized neuropathologically by degeneration of myelin and axonal loss[3]. It is clear now that the neuropathological lesions in SCD are due to overproduction of myelinolytic tumor necrosis factor α (TNF-α) and to the reduced synthesis of the two neurotrophic agents epidermal growth factor (EGF) and interleukin-6. This deregulation of the balance between TNF-α and EGF synthesis is induced by cobalamin deficiency[4].The MRI findings of the spinal cord in SCD are high-signal intensity on T2-weighted images within the posterior or lateral columns. Brain lesions of vitamin B12 deficiency over the medulla oblongata, pons, mesencephalon and crus cerebelli have also been reported[5]. SCD can also result from common variable immunodeficiency syndrome, paraneoplastic malabsorption, folate deficiency, acute monoblastic leukemia and nitrous oxide anesthesia[6].

The main symptoms of SCD are paresthesia, stiffness, numbness or tingling of the limbs; sensory ataxia; and impaired vibration and joint position sensation. Spastic paraparesis may develop later with anterior column involvement if SCD is left untreated. Babinski's sign may be present, and the deep tendon reflexes are variable[7]. When these symptoms are associated with macrocytic anemia, the possibility of SCD could be definitely considered. Since hematologicalfeatures(macrocytic anemia) may present later after neurological manifestations, blood vitamin B12 levels becomes the gold standard to evaluate for any early depletion. Thus spinal MRI is a good diagnostic tool[8] for early detection to be further confirmed with biochemical tests. Once the diagnosis of SCD is suspected, treatment with vitamin B12 injection should be started as early as possible to avoid irreversible neurologic damage. Improvement in myelopathy may occur if vitamin B12 therapy is started early in the course of the disease. The resolution of the MRI changes in our case correlated well with the biochemical tests[7,9].

Vitamin B12 cobalamin can be found in food from animal and some plant sources. The average adult ingests 5–30 g of vitamin B12 per day, of which 1–5 g are absorbed. There is a total body store of 2–5 mg of B12, of which 1 mg is stored in the liver. Consequently, the effects of vitamin B12 deficiency may not be appreciated until several years later, when these stores are depleted. Several laboratory tests are used to directly or indirectly measure the amount of cobalamin and enable detection of a deficiency[10].Serum B12, Malonic Acid, Methylmalonic Acid, Schilling Test, Complete Blood Count – to evaluate for megaloblastic anemia. Therapy with vitamin B12 results in partial to full recovery, depending on the duration and extent of neurodegeneration.

In the three cases we have reviewed , there is one complete inverted V sign, one M like appearance and one discontinuous inverted V sign- all the abnormal signal are involving the dorsal columns of the cord and other two cases where there was coexisting focal minimal signal at lateral columns in one and anterior column in the other.

Patients were followed up with imaging in the two asymptomatic cases which showed complete resolution. However the other symptomatic case could not be followed up on imaging due to financial restrains of the patient, but showed significant improvement symptomatically.

Biochemistry tests revealed reduced serum vitamin B12 levels in both symptomatic & asymptomatic patients, macrocytic hypochromic anemia in only symptomatic case &normal folic acid levels in all our patients. Reactive status was checked for and ruled out.

Conclusion

The presentation and clinical features of subacute combined degeneration (SCD) is no mystery to clinicians. It is but the incidentally detected cases on MRI imaging that need special attention here, as it is

well documented that if imaging has its say in screening, we could well prevent SCD a grave complication of vitamin B12 deficiency from a taking a deep bite before its too late. This article is to highlight that if screening could be performed in cases in patients with known causes, statistics on morbidity will definitely be changing in coming years.

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