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IMAGING HIGHLIGHT

Acute acro-paraesthesia and bilateral clumsy hand syndrome rare presenting manifestation of vitamin B_{12} deficiency: a case report highlighting clinico-radiological findings at diagnosis and follow-up

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Subacute combined degeneration of the spinal cord (SCD) is a neurodegenerative disease characterized by subacute progression in the central and peripheral nervous systems mainly caused by vitamin B_{12} deficiency.[1] In the diagnosis of vitamin B_{12} deficiency, measurement of urine methylmalonic acid (MMA) and serum homocysteine levels has been reported as more useful than measurement of total vitamin B_{12} level.[2] We report a rare case of SCD that we diagnosed by determining urine MMA, serum homocysteine levels with significant neurologic symptoms and characteristic magnetic resonance imaging (MRI) findings of the spinal cord, in the absence of characteristic findings of vitamin B_{12} deficiency and anaemia.
Informed written consent obtained and documented.

CASE REPORT

In September 2019, a 25-year lady with a previous history of alopecia areata presented to the neurology clinic with a 2-week history of paraesthesia and numbness over all 4 limbs and the trunk with clumsiness of her hands. She reported electric like sensation over her lower limbs if she bent her neck forwards. There was no weakness or incontinence nor was there a history of recent vaccinations, fever or other neurological symptoms. She consumed a non-vegetarian diet and had no history of weight loss or anaemia. There was no exposure to heavy metals use or to nitrous oxide. On examination, the patient was calm and higher mental functions were preserved. She had impairment of fine touch, position and vibration in bilateral hands and feet and positive Romberg's sign. No sensory level could be ascertained and there was no motor weakness. Visual examination, cerebellar function and gait assessment were normal.

Routine lab investigations showed borderline anaemia with decreased total RBC count.

Magnetic Resonance Imaging of the cervico-thoracic spine revealed increased T2 signal bilaterally and symmetrically in the posterior columns of the cervical cord from C2 to C5 levels. (Figurre 1, 2) No contrast enhancement was seen. MRI of brain and orbits with contrast was unremarkable, CSF examination did not show any significant abnormality including absent oligoclonal bands. The ESR was 5 and CRP 1.7. Auto immune screen (ANA, anti-dsDNA, ANCA, Anti-Ro, Anti-La, lupus anticoagulant) was negative. Anti-AQP4 and anti-MOG antibodies were negative. Infectious screen was negative for HIV, Hepatitis B, Hepatitis C. A pan CT scan to look for occult malignancy was normal. Copper levels were normal and there was no history of nitrous oxide abuse/exposure.

Serum vitamin B_{12} level was 242 (pmol/L/ 133-675) with negative Intrinsic factor and anti-parietal antibody. Urine methyl-malonic acid (44.0 umol/L) and serum homocysteine (18 umol/L) were noted to be elevated suggestive of B12 deficiency.

Based on significant clinico-radiological findings and biochemical suggestion of B_{12} deficiency despite the normal B_{12} levels, diagnosis of subacute combined degeneration of spinal cord was established. Patient was treated with parenteral B_{12} therapy and discharged on oral B_{12} tablets.

Complete clinical recovery was achieved within 6 weeks on follow up. Follow up MRI of the cervico- thoracic spine showed interval decrease in the T2W hyperintensity in the posterior columns

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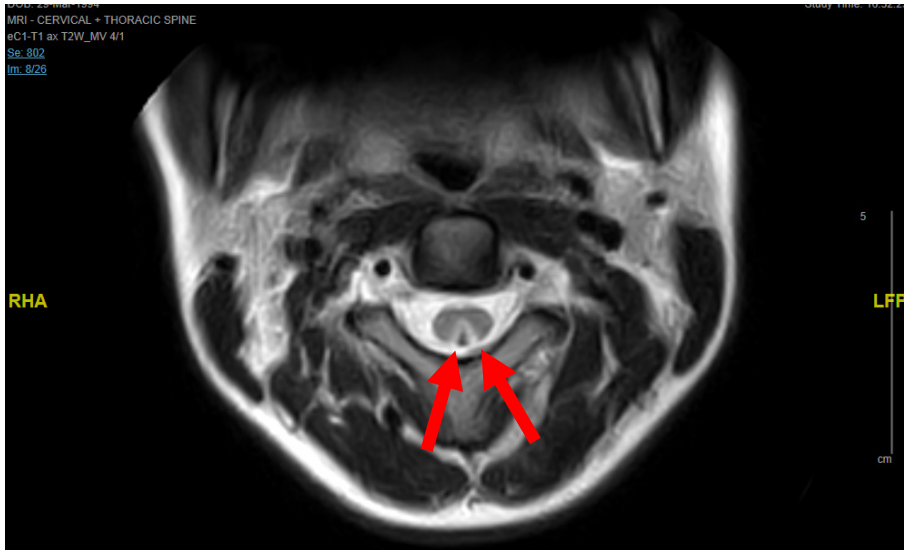

Figure 1. **A.** Axial T2W MRI at C3 vertebral level of the spinal cord showing characteristic "inverted V pattern" hyperintense signal (arrow) in the posterior columns of the spinal cord.

of the cord. There was no contrast enhancement or new lesion seen.

Retrospectively history taking at this point revealed that patient was taking neurobion tablets which contains vitamin B_{12} from GP for paraesthesia which could have been the cause of borderline normal serum vitamin B_{12} level in our patient.

DISCUSSION

Vitamin B12 deficiency may affect both the central and the peripheral nervous system. Spinal cord lesion caused by vitamin B_{12} deficiency is known as subacute combined degeneration of spinal cord (SCD). SCD is a result of myelin damage in posterior and lateral columns of spinal cord caused by deficiency of vitamin B_{12}. Area involved is the lower cervical and upper Dorsal region.

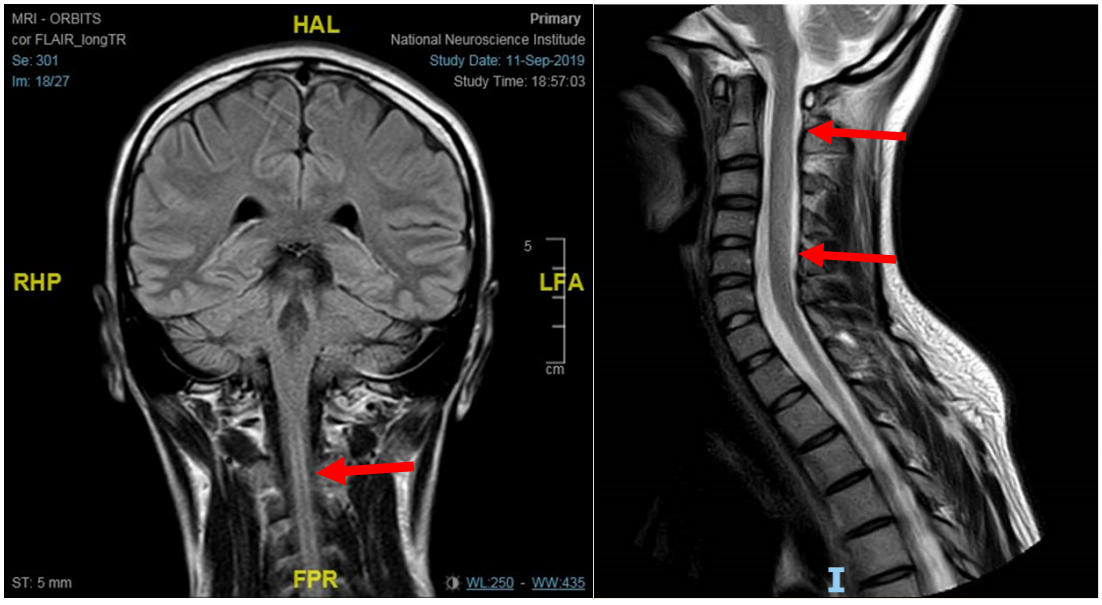

Figure 1 **B**, **C**. Coronal FLAIR and Sagittal T2W MRI showing linear hyperintense signal in (arrows) the cord extending from C2-C5 vertebral levels predominantly in the posterior aspect of the cord with mild swelling.

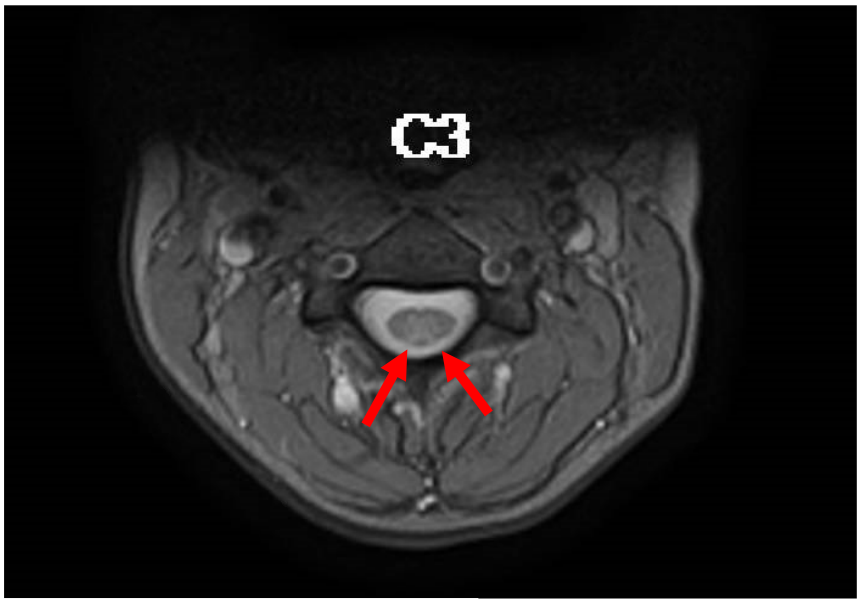

Figure 2. **A**. Axial T2W MRI at C3 vertebral level showing interval decrease in the T2W hyperintensity in the posterior columns of the cord.

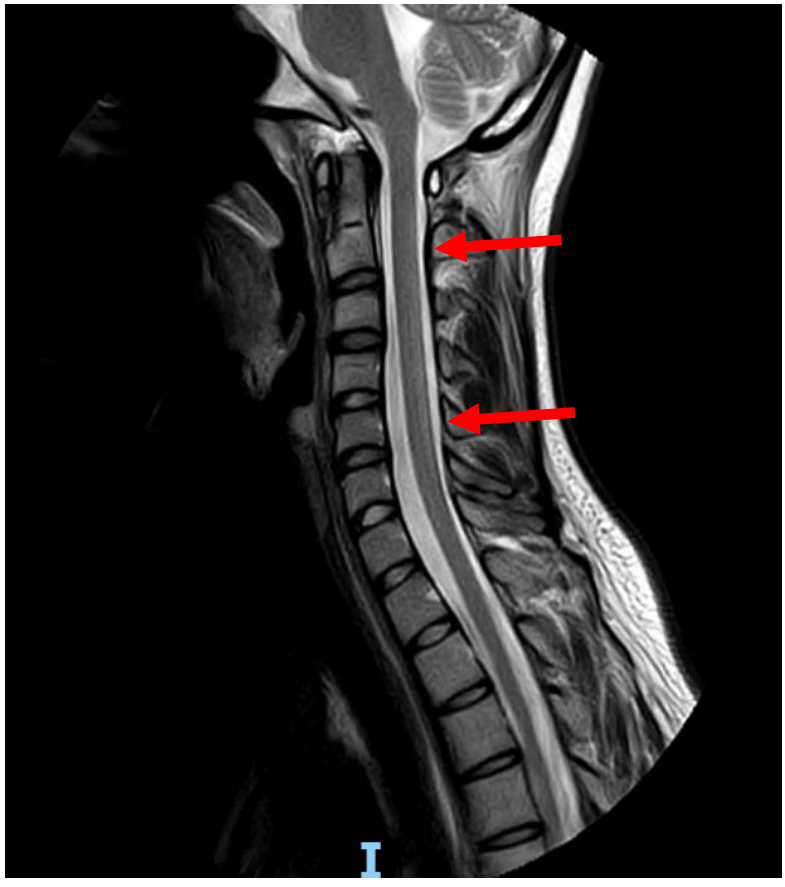

Figure 2. **B**. Sagittal T2W MRI shows interval reduction in hyperintense signal (arrows) with reduced swelling and thickness, although faint hyperintensity is still seen in the previous longitudinal extent of involvement.

The main symptoms of SCD are paraesthesia, stiffness, numbness or tingling of the limbs, sensory ataxia, with impaired vibration and joint position sensation. Spastic paraparesis may develop if remains untreated. Babinski signs may be present, and the deep tendon reflexes are variable. If patients have the above-mentioned common symptoms of SCD and macrocytic anaemia, the possibility of SCD should be highly considered. However, the hematologic abnormalities of vitamin B12 deficiency (macrocytic anaemia) may develop after the neurologic abnormalities. Some SCD patients might have minimal symptoms without hematologic abnormalities initially, such as acroparaesthesia and Lhermitte's sign only.[3] At this moment, in the early stage, in addition to blood vitamin B_{12}, homocysteine levels and urine methyl malonic acid, spinal MRI may be a good diagnostic tool.

A recent meta-analysis showed that up to one-third of subacute combined degeneration patients have normal or elevated serum vitamin B12, and suggests that the diagnosis of subacute combined degeneration should remain based on clinical manifestations and not to exclude patients only based on normal/elevated levels of serum vitamin B12.[4]

MRI is the modality of choice to image the spinal cord. The classic appearance of SCD is a longitudinally extensive involvement of the dorsal columns of the spinal cord and occasionally, involvement of the lateral columns. The cervical and upper thoracic segments are most commonly involved. On axial section, the involvement of the dorsal columns gives an inverted "V" appearance on T2 weighted images.[5] Copper deficiency, which may be associated with B12 deficiency or may occur in isolation, can show a similar MRI appearance.[6] Other non-metabolic myelopathies can potentially involve the dorsal columns of the cord, but tend to be scattered and are not usually longitudinally extensive (i.e. more than 3 vertebral segments in length). Tabes dorsalis or neurosyphilis may involve the dorsal columns but is a relatively rare condition. Spinal cord abnormality in SCD is reversible after treatment. The abnormal MR signal in the spinal cord may either disappear on follow-up after months, or it may persist, especially in cases diagnosed and treated at an advanced stage.[7] Brain lesions of vitamin B_{12} deficiency over the medulla oblongata, pons, mesencephalon and crus cerebelli have also been reported.[8]

Once the diagnosis of SCD is suspected, the treatment with vitamin B_{12} replacement should be started as early as possible to avoid irreversible neurologic damage. The clinical symptoms and MRI abnormalities showed improvement in our patient 6 weeks later, as in previous cases reports. SCD seems to be a reversible disease after vitamin B_{12} therapy. The timing of diagnosis and duration of illness may play an important role in the treatment response and prognosis of SCD, hence SCD patients should be diagnosed early by having a high index of suspicion and use diagnostic tools such as MRI. Spinal MR imaging assists in early diagnosis and follow-up MR imaging findings correlate with clinical outcome after treatment with vitamin B12 supplementation. Early diagnosis and treatment are crucial because SCD represents a potentially reversible cause of non-compressive myelopathy and treatable with vitamin B_{12}.[9] Diagnostic delay and/or late initiation of therapy may result in permanent irreversible injury to the spinal cord with little or no improvement on treatment.

DISCLOSURE

Conflict of interest: None

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