

CASE REPORT

Acute bilateral useless hand syndrome: a rare presenting manifestation of vitamin B₁₂ deficiency

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SUMMARY

We report a case of bilateral useless hand syndrome, a rare presenting manifestation of vitamin B₁₂ deficiency. A 38-year-old man, a strict vegetarian and a teacher by occupation, presented with acute onset clumsiness of both hands while performing fine movements. Detailed history-taking, examination of the patient and relevant investigations (complete blood count, serum vitamin B₁₂ and MRI of the cervical spinal cord) were carried out. Laboratory analysis was suggestive of vitamin B₁₂ deficiency and MRI demonstrated a lesion involving the posterior columns of the cervical cord. The patient was diagnosed as a case of non-compressive cervical myelopathy predominantly involving the posterior column due to vitamin B₁₂ deficiency. Acute bilateral useless hand syndrome can be a rare presenting feature of vitamin B₁₂ deficiency.

BACKGROUND

Useless hand syndrome is defined as difficulty in executing distal fine movements of the hands in the absence of any weakness (pyramidal) or extrapyramidal involvement. It is predominantly attributed to involvement of the posterior columns of the spinal cord. Patients with useless hand syndrome have difficulty using their fingers to manipulating objects. This phenomenon has been reported in patients with multiple sclerosis (MS)¹⁻⁵ and may rarely be due to cervical compressive myelopathy.⁶

Common neurological manifestations of vitamin B₁₂ deficiency are subacute combined degeneration of cord, behavioural abnormality, optic nerve involvement, peripheral neuropathy and myelopathy.^{7,8} Acute bilateral useless hand syndrome as a presenting feature of vitamin B₁₂ deficiency has not been reported in the literature to date.

CASE PRESENTATION

A 38-year-old man presented with difficulty in performing fine motor movements with his hands, such as buttoning and unbuttoning of shirts, tearing chapatias, writing and manipulating objects with his fingers, for the past 7 days. He had no history of weakness and/or stiffness in any of the limbs or tingling or numbness in the hands or feet. There was no history of exposure to toxins or drugs, and no high-risk behaviour. There was no history of similar episodes and no chronic illness in the past. He had no diabetes, no hypertension and was a strict vegetarian with no addictions.

The patient was thin-built, and his vitals and general examination were normal. On neurological examination, higher mental function, cranial nerves

and fundus examination were normal. Nutrition, tone and power in all four limbs were normal. There was impairment of dexterity in both hands, loss of joint position and sense of vibration in both feet and hands, while pain and temperature sensations were intact. Astereognosis and pseudoathetosis were present in both hands. Deep tendon reflexes were normal in all four limbs except bilateral ankle jerks, which were absent with bilateral flexor plantar response. Romberg's test was positive. Gait was normal, and systemic examination including skull and spine was normal.

INVESTIGATIONS

On investigations, haemoglobin was 12.4 g/L, total leucocyte count (TLC) 8000/mm³, platelet count 50 000/mm³ and mean corpuscular volume (MCV) 107.6 (normal range 80-96 fL). Peripheral blood smear showed presence of macrocytes and reduced platelet counts. Renal and liver function tests, random blood sugar, serum electrolytes and thyroid function tests were normal. ELISA for HIV, HBsAg and hepatitis C virus was normal. Serum vitamin B₁₂ was low (130 pg/mL). Routine cerebrospinal fluid (CSF) examination was normal and CSF virology study for herpes simplex virus,^{1,2} cytomegalovirus, Epstein-Barr virus and mumps was negative. Nerve conduction studies showed absent sural sensory nerve action potentials (SNAPs) and decreased amplitude of bilateral median and ulnar SNAPs. Pattern reversal visual-evoked potentials were normal.

On the basis of history, examination and laboratory features, a provisional diagnosis of pseudoathetosis due to non-compressive myelopathy (vitamin B₁₂ deficiency) was made. The patient was subjected to cervical spine MRI, which showed abnormal hyperintensity on sagittal T2-weighted (T2W) images in the cervical spinal cord, which was localised to posterior columns from C3 to C7, not contrast enhancing and, on axial T2W MRI, there was a classical 'inverted V sign' in posterior columns (figure 1A, B). T2W and fluid-attenuated inversion recovery sequence on MRI of the brain did not show any white matter hyperintensity.

After neuroimaging, a diagnosis of cervical non-compressive predominantly posterior myelopathy due to vitamin B₁₂ deficiency was made.

TREATMENT

The patient received treatment with intramuscular injection of vitamin B₁₂ (cyanocobalamin) 1000 µg daily for the first 7 days, then weekly for 4 weeks, followed by monthly for 12 months.



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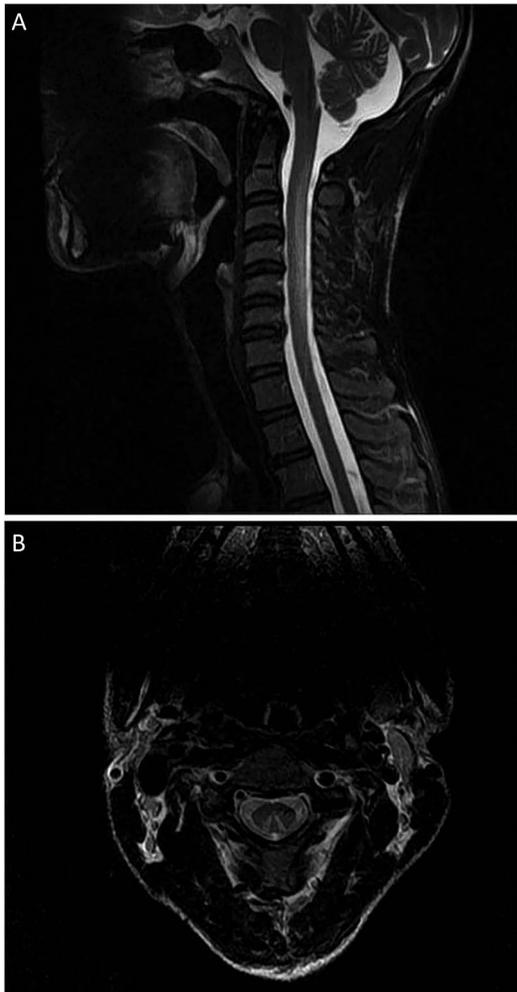


Figure 1 (A) T2-weighted (T2W) sagittal section MRI of the cervical spine showing hyperintensity involving the posterior columns from C2 to C7 level. (B) Axial T2W cervical spine MRI showing a typical inverted 'V'-shaped hyperintensity.

Along with this, as he was strictly vegetarian, the patient was also advised to have vegetarian food products rich in vitamin B₁₂, such as legumes, as well as milk and milk products.

OUTCOME AND FOLLOW-UP

The patient started showing significant improvement on follow-up at 4 weeks and, when examined at 8 weeks, he was completely relieved of his symptoms. When he was examined at 9 months he was doing well.

DISCUSSION

Useless hand syndrome in patients with MS has been described by Oppenheim.⁹ It is a phenomenon in which the hand is functionally impaired due to posterior column deficits leading to clumsiness of complex finger movements and loss of manual dexterity. This phenomenon has been reported in patients with MS and cervical spondylosis. Unilateral involvement has been noted to be more common than bilateral involvement in patients with MS.¹⁻⁵

The pathophysiology behind useless hand syndrome due to posterior cervical cord involvement remains unclear. It has been hypothesised that there is a centre for sensorimotor integration in the high cervical cord; this centre works to convey kinaesthetic and discriminative sensations necessary to guide fine finger movements. The propriospinal neurons in the cervical cord receive afferent signals from cutaneous forelimbs and

efferent signals from the corticospinal tract, and integrate these signals, contributing to executing purposive forelimb movements at a premotoneuronal level.^{9 10}

Neurological involvement is very common in vitamin B₁₂ deficiency, reported in 40–70% of patients. The most commonly affected sites are the brain, optic nerve, spinal cord and peripheral nerves, in varying combinations; among these, the most common and important is involvement of the posterior and lateral columns of the cervical and thoracic spinal cord. These patients can have gradual or subacute onset paraesthesias in the feet, followed by the hands; later, patients may develop spastic weakness of both lower limbs and have significant ataxia. Behavioural and visual symptoms may also be present, along with myeloneuropathy. The mechanism behind myeloneuropathy is due to accumulation of methyl malonyl CoA and abnormal fatty acids that constitute the myelin sheath, leading to defective myelination of the neural sheath. Neuropathological study has demonstrated spongiform changes with foci of myelin and axonal destruction mainly in the posterior and lateral columns. The other features of vitamin B₁₂ deficiency are macrocytic anaemia, skin pigmentation and atrophic glossitis, however, acute onset clumsiness of both hands due to involvement of the posterior column as the presenting symptom of vitamin B₁₂ deficiency is not commonly seen.^{7 8 11 12}

Vitamin B₁₂ deficiency is quite common in pure vegetarians, as few vegetarian products are good sources of vitamin B₁₂. So it is very important for vegetarians to have foods rich in vitamin B₁₂, such as legumes and milk products. This measure can protect vegetarians from various clinical manifestations due to vitamin B₁₂ deficiency.

In our patient, there was an acute history of clumsiness in both hands with significant deterioration on eye closure, without any pyramidal or extrapyramidal findings, and MRI findings were suggestive of abnormal T2W hyperintensity in the posterior and lateral cervical cord, without contrast enhancement. Axial T2W imaging showed a classical inverted 'V' sign.^{13 14} Our patient had a classic MRI picture of vitamin B₁₂ deficiency, which was further corroborated by increased MCV and low vitamin B₁₂ level. MRI of the brain was normal and the patient had significant improvement after supplementation of vitamin B₁₂. Early recognition and treatment can reverse all symptoms of this condition affecting the nervous system, so timely diagnosis of vitamin B₁₂ deficiency is very important.

Learning points

- ▶ Useless hand syndrome is defined as difficulty in executing distal fine movements of the hands in the absence of any weakness (pyramidal) or extrapyramidal involvement. It is predominantly attributed to involvement of the posterior columns of the spinal cord.
- ▶ Common causes of this syndrome are multiple sclerosis and cervical spondylosis.
- ▶ Vitamin B₁₂ deficiency can present as acute onset useless hand syndrome due to involvement of the posterior cord early on in the disease course.
- ▶ MRI of the spinal cord shows classic abnormalities in the posterolateral region suggestive of demyelination of the posterior and lateral columns.
- ▶ Early diagnosis and treatment play an important role in the reversibility of neurological deficits; delayed treatment can result in irreversible disabling neurological impairment.

Competing interests None declared.

Patient consent Obtained.

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