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A case of subacute combined degeneration: MRI findings

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Introduction

Subacute combined degeneration (SCD) is a neurological disorder of the spinal cord caused by vitamin B12 deficiency. Neuropathological investigation typically reveals lesions in the posterior columns of the spinal cord. Until recently, these lesions could only be demonstrated at autopsy. With MRI, however, they can be observed in vivo. There have been a few case reports of MRI findings of SCD which have demonstrated abnormal high T2 signal in the posterior columns [1–5]. There has, however, been only one reported case of SCD with contrast-enhanced MRI [5]. We describe a second case of SCD in which gadolinium-enhanced cervical MRI was performed before and after treatment.

Case report

A 19-year-old black male with an unremarkable past medical history presented with 4 weeks of gradually progressing tingling in his

Abstract The specific spinal cord lesion caused by vitamin B12 deficiency is known as subacute combined degeneration (SCD). Neuropathological studies of SCD show lesions mainly in the posterior and lateral columns, involving the cortico-spinal and spino-cerebellar tracts.

We report a case of SCD in a 19year-old man who presented with 4 weeks history of gradually progressing tingling in both hands. MRI of the cervical spine demonstrated symmetrical areas of T2 signal abnormality involving the dorsal columns of the cervical cord from the C2 through C5 levels associated with spinal cord expansion. He was treated with vitamin B12 supplements and experienced gradual improvement in his clinical symptoms. Repeat MRI of the cervical spine after 2 months revealed slight decrease in the area of abnormal signal.

Key words Spine · Cervical spinal cord · Posterior column · Subacute combined degeneration · Vitamin B12 deficiency · Degenerative process · Magnetic resonance imaging

hands and feet. At the time of admission he was unable to use his hands to feed or dress himself.

Clinical examination revealed profound loss of proprioception and vibration sense in the upper extremities. The patient was unable to place his finger on his nose with his eyes closed. Romberg sign was positive but proprioception in the lower extremities was relatively spared.

Laboratory tests on admission revealed macrocytic anemia with hemoglobin of 8.0 mg/dl. The mean corpuscular volume (MCV) was 107 fl and mean corpuscular hemoglobin (MCH) was 36 pg/cell. The patient also had increased bilirubin (total bilirubin of 2.2 mg/dl, direct bilirubin of 0.5 mg/dl), increased LDH (4445 IU/l), and mildly elevated erythrocyte sedimentation rate (34 mm/h).

From the clinical examination, pathological involvement of the posterior columns of the cervical spinal cord was suspected, and cervical MRI demonstrated a symmetrical area of T2 signal abnormality involving the dorsal columns of the cervical cord at levels C2 to C5 associated with spinal cord expansion (Fig. 1 a, b). The medial portions of the dorsal columns appeared unaffected. Neither abnormal T1 signal nor abnormal contrast enhancement was present.

Based on the clinical, laboratory and imaging findings, a diagnosis of vitamin B12 deficiency was suggested. Serology revealed

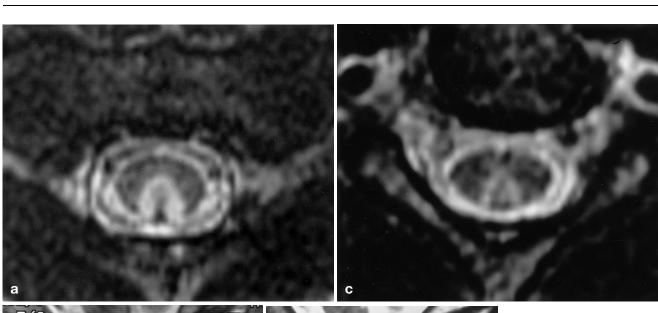




Fig.1 a,b Initial MRI of the cervical spine (a TR/TE/NEX 4500/95/2, **b** 4000/108/2) demonstrates a symmetrical area of T2 signal abnormality with cord expansion involving the dorsal columns of the cervical cord from C2 to C5. c,d Follow-up MRI of the cervical spine (c 4500/95/2, d 400/100/ 2) reveals a slight decrease in the extent of the abnormal T2 signal area in the posterior columns. There is no sign of contrast enhancement (not shown) and there is no longer any evidence of cord expansion

decreased vitamin B12 level (less than 100 pg/ml), and a subsequent Schilling's test revealed decreased percent urinary clearance (4.3%) of the orally administrated cobalt 57 cyanocobalamin, which indicated decreased absorption of vitamin B12.

The patient was treated with intravenous and intramuscular injections of vitamin B12 and experienced gradual improvement in his clinical symptoms. At the time of a follow-up MRI 2 months

after the initial presentation, he was able to dress and eat with his hands without significant difficulty. The repeat images of the cervical spine revealed a slight decrease in the extent of the abnormal T2 signal area in the posterior columns (Fig. 1c, d). There was no evidence of contrast enhancement and the cord swelling had diminished significantly.

Discussion

The spinal cord lesion of vitamin B12 deficiency is known as subacute combined degeneration (SCD). Deficiency of vitamin B12 can occur by impaired absorption due to lack of intrinsic factor (idiopathic), total or partial gastrectomy, blind loop syndrome, celiac disease, Crohn's disease, or chronic pancreatic insufficiency. Strict vegetarians and patients with infestation of fish tapeworm (*Diphyllobothrium latum*) may also develop serious B12 deficiencies. Increasing prevalence of vitamin B12 deficiency has been reported in patients infected by human immunodeficiency virus, particularly in those with acquired immunodeficiency syndrome [6].

The earliest symptoms and signs of vitamin B12 deficiency are paresthesia and areflexia. These are related to damage to the peripheral nerves, rather than the spinal cord. Spinal cord lesions may subsequently occur and are characterized by dysesthesia and disturbance of deep sensation. In an advanced case, subsequent degeneration of the corticospinal tracts and dorsal columns may lead to paraplegia. The spinal cord damage may not improve significantly with vitamin B12 therapy and may be made dramatically worse if folic acid is given instead of vitamin B12. Early diagnosis is, therefore, essential so that patients may be treated before significant cord damage has occurred. Treatment with hydroxycobalamin or cyanocobalamin results in an arrest of disease progression and some degree of reversal of symptoms, with complete recovery in almost half of the patients [7].

Neuropathological studies of SCD show that the main lesions are present in the posterior and lateral columns, involving the cortico-spinal and spino-cerebellar tracts. Pathological findings are characterized by cord demyelination with eventual axonal loss.

There have been several case reports of the MRI findings in SCD [1–5]. They typically describe symmetrical high T2 signal bilaterally in the posterior columns of the spinal cord. Cases with follow-up studies showed improvement of the abnormal T2 signal with vitamin B12 supplementation therapy [1–4]. Our case also showed this typical pattern of posterior column involvement and interval improvement after vitamin B12 supplementation. Sparing of the medial dorsal columns in our case probably accounts for the relative sparing of the proprioceptive sense in the lower extremities.

In only one other reported case did the patient receive gadolinium-DTPA. In that case the post-gadolinium-DTPA images revealed enhancement in the posterior columns [5]. Our case did not show enhancement, and this probably reflects the absence of the breakdown of the blood nerve (spinal cord) barrier. Our case also demonstrated cord expansion on the initial MRI, which is, to our knowledge, a previously unreported finding. Since it is known that the earliest change of demyelination is the fusiform expansion of the myelin sheaths, this cord expansion presumably represents the acute stage of demyelination [8].

Differential diagnoses of abnormal signal lesions in the posterior columns of the spinal cord include infectious or post-infectious myelitis, peripheral neuropathy, carcinomatous radiculopathy, radiation myelitis, multiple sclerosis, arterial or venous ischemia, traumatic cord injury, metabolic disease (including vitamin E deficiency) and acute transverse myelitis [9–11]. Preferential involvement to the posterior columns may be seen in all of these conditions, and therefore the radiological finding of posterior column involvement is not specific for SCD. However, the diagnosis of SCD should be suggested so that appropriate laboratory testing may be performed.

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