

V for vitamin, inverted V for lack of it: A rare case of subacute combined degeneration of cord with bladder symptoms and associated multiple Tarlov cysts

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ABSTRACT

Subacute degeneration of cord (SACD) is a rare presentation of a common disease and it should be kept in mind when the patient develops symptoms of peripheral neuropathy. This is a case of a 65-year-old male who presented with pins and needles sensation of bilateral hands and feet and with difficulty in voiding. Neurological examination showed large fiber neuropathy with an exaggerated knee jerk, absent ankle jerk, extensor plantar, and a positive Romberg's sign. Routine laboratory investigations showed low hemoglobin and low vitamin B₁₂ levels. Magnetic resonance imaging of the spinal cord showed a hyperintense lesion in the posterior aspect of the cervical cord with a classical "inverted V" sign on the axial section which is suggestive of SACD. Coincidentally, there were multiple Tarlov cysts present. Finally, the patient improved with vitamin B₁₂ injections. This case is unique as an association of SACD with Tarlov cysts with a classical inverted V sign has not been reported according to the literature.

Keywords: Subacute Degeneration of Cord; Tarlov Cysts; Inverted V Sign

Anemia is one of the most common prevailing disorders in India. Vitamin B₁₂ deficiency is a largely prevalent disorder in the vegetarian population in our country. Neurological complications like peripheral neuropathy are common.

Subacute degeneration of cord (SACD) is a rare presentation of a common disease and it should be kept in mind when the patient develops symptoms of peripheral neuropathy. While SACD is a debilitating presentation, it is easily treatable and physicians should have a high degree of suspicion [1].


Here, we present the case of a 65-year-old male who presented with pins and needles sensation of bilateral hands and feet and with difficulty in voiding.

CASE REPORT

A 65-year-old male who is a farmer by occupation came to the General Medicine outpatient department with complaints of a tingling sensation in all four limbs in a glove and stocking pattern for 1 month which initially appeared in the upper limb and which slowly progressed to the lower limb. The patient also complained of giddiness while walking during the night

and there was a history of multiple falls. This was followed by difficulty in buttoning his shirts and mixing his food for the past 20 days. The patient also had trouble voiding urine for over a week. He also complained of increased urgency, frequency, and a sensation of incomplete voiding. He also complained of painless bleeding per rectum which, on examination, was diagnosed as grade-4 hemorrhoids. The patient had no comorbidities or no previous surgeries. He was a chronic smoker with 20 pack-year. He consumed a mixed diet and had non-vegetarian food twice a week.

On examination, he was a moderately built man with a body mass index of 21.8 kg/m². The general examination showed pallor. His pulse rate was 84 beats/min which was regular in rate, rhythm, and character with no delays. The blood pressure was 110/80 mm Hg. Higher mental functions and cranial nerve examinations were within normal limits. The motor system showed a distal weakness of the upper limb with a reduced handgrip and power in the wrist region was 4/5. Deep tendon reflexes showed exaggerated knee jerk with normal deep tendon reflexes in others. The plantar reflexes were bilaterally extensor, suggestive of pyramidal tract involvement. Sensory examination showed evidence of posterior column involvement with impaired fine touch and vibration sense up to the wrist in the upper limb and mid-leg in the lower limb. Romberg's sign was positive. The patient had a wide-based gait and no cerebellar signs, suggestive

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of sensory ataxia. These findings were likely in favor of non-compressive myelopathy as the patient had motor and sensory deficits in the form of a dorsal column involvement with a bladder symptom.

Basic laboratory investigations showed hemoglobin of 8.4 mg/dl. Blood indices showed a mean corpuscular volume of 94fl and mean corpuscular hemoglobin of 29 pg. Total counts and platelet counts were within normal limits. The reticulocyte index was 0.78, indicating a hypoproliferative marrow. The peripheral smear shows a dimorphic anemic picture. Vitamin B₁₂ level was 50 picogram/ml. Folate levels were normal. The patient had iron deficiency too, secondary to the bleeding per rectum. While evaluating for Vitamin B₁₂ deficiency, thyroid-stimulating hormone levels were normal. Anti-nuclear antibody testing was negative. Upper gastrointestinal endoscopy was done which showed multiple gastroduodenal erosions with an antral ulcer. Antibodies against intrinsic factor were also sent and found negative. As the patient had symptoms of a spastic bladder, uroflowmetry was done which showed evidence of a neurogenic bladder.

Imaging studies were done which showed chronic lacunar infarct in the right medial temporal lobe. The cervical spine region showed a long segment hyperintensity along the posterior aspect of the cervical and upper dorsal cord with a characteristic “Inverted V” sign on the axial section. The lesion extended from C1 to D2 (Fig. 1a). Multiple nerve root cysts known as Tarlov cysts were noted in the cervical, dorsal, and lumbosacral regions (Fig. 1b). This was most likely an incidental finding as the patient *per se* did not give any history of compressive symptoms but were an interesting finding nonetheless.

Nerve conduction studies showed demyelinating neuropathy of bilateral common peroneal nerves with sensory axonopathy of bilateral superficial peroneal nerves. A neurosurgeon’s opinion was taken in view of the multiple Tarlov cysts and since the patient did not have any compressive symptoms, no surgical intervention was required.

The patient was managed with vitamin B₁₂ supplementation and then he showed considerable and rapid improvement in

symptoms. He also slowly regained function of the bladder with complete recovery. This indicates that a high degree of suspicion is required to diagnose a case of subacute degeneration of the cord as the patient shows good resolution of symptoms when treated early and adequately.

DISCUSSION

Nutritional anemia is an extremely common disease in most developing nations. Almost one-quarter of the world’s population is anemic [1]. While Iron deficiency anemia remains the most common variant, vitamin B₁₂ is an essential vitamin that is primarily procured by a non-vegetarian diet [2]. The Indian population, which mainly has a vegetarian diet, tends to have more incidence of vitamin B₁₂ deficiency [3]. Vitamin B₁₂ deficiency is caused by malabsorption, pernicious anemia, post-ileal resection, post-gastrectomy status, chronic *H. pylori* infections, and certain drugs such as antimetabolites [4].

Vitamin B12 deficiency can have multiple neurological manifestations such as myelopathy, neuropathy, dementia, behavioral changes, optic nerve involvement, paraparesis, quadriparesis, and bladder involvement [5]. SACD is a documented complication of vitamin B₁₂ deficiency, where the posterior and lateral corticospinal tract pathways are involved, leading to neurological manifestations. When there is combined demyelination of cranial nerves and peripheral nerves, it is termed as “Combined system disease” [6]. While it is named subacute combined degeneration of cord, the majority of the cases have predominantly sensory symptoms. The classical clinical features of exaggerated knee reflex, depressed ankle jerk, and depressed extensor plantar reflex are rarely appreciated in clinical practice, and other close differentials in such a scenario such as Fredrich’s ataxia, HIV myelopathy, and Tabes dorsalis should be kept in mind.

Studies have shown that bladder involvement is not unheard of in Vitamin B₁₂ deficiency and there have been isolated cases of bladder involvement in SACD [7]. Studies have shown that SACD with bladder symptoms can have both upper motor neuron and lower motor neuron like features. The involvement of the lateral corticospinal tract would probably indicate detrusor overactivity, while a flaccid bladder would be more common when the posterior column is involved. In our case, the patient had symptoms suggestive of a corticospinal tract involvement.

The T2-weighted hyperintensity image is essentially due to demyelination and gliosis. The usual differentials of a hyperintensity in T2-weighted image in the spinal cord include demyelinating disorders such as multiple sclerosis, HIV, and Herpes virus infection. However, SACD usually presents in the dorsal column with lesions being contiguous over numerous segments. Tarlov cysts are fluid-filled nerve root cysts found mostly at the sacral level of the spine. These cysts typically occur along with the posterior nerve roots [8].

The initial presentation of these cysts was ambiguous, but the neurosurgeon, I. Tarlov found out that some of these cysts can be

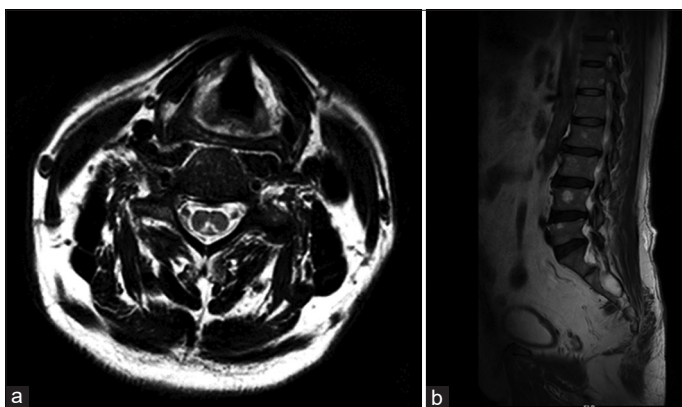


Figure 1: (a) T2-weighted/short tau inversion recovery image along the posterior aspect of the dorsal and cervical column with an inverted V sign on the axial section; (b) well-defined T2-weighted hyperintense cystic lesions noted in sacral region which was suggestive of Tarlov cysts

symptomatic and needed to be corrected surgically. Studies over the years have shown that Tarlov cysts can cause compressive symptoms such as pain, bladder and bowel dysfunction, paresthesia, and cauda equine-like picture. Rarely, there has been an association with autoimmune collagen disorders such as Marfan's disease and Ehler Danlos syndrome [9]. The multiple Tarlov cyst was an incidental finding in this scenario and studies have shown that more often than not, they are asymptomatic, but there have been isolated reports of extradural compressive myelopathies which required immediate surgical intervention. This patient possibly had posterior column involvement in the form of long fiber neuropathy, positive Romberg's sign, and a pyramidal tract pathology in the form of bladder overactivity.

CONCLUSION

The interesting aspect of this case is the presentation of vitamin B₁₂ deficiency as SACD with bladder symptoms and an incidental finding of a Tarlov cyst. The magnetic resonance imaging picture of the classical "inverted V" sign was unique. The importance of having high clinical suspicion of vitamin B₁₂ deficiency-induced SACD is because patients respond well to treatment with cyanocobalamin.

DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate patient consent forms.

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