

Subacute combined degeneration associated with vitamin E deficiency due to small bowel obstruction

A case report

Jong Burm Jung, MD^a, Yuntae Kim, MD^{a,*}, Kiyoung Oh, MD^a, Soo A Kim, MD^a, Joung Hyun Doh, MD^a, Hye Jeong Oh, MD^b, Jin Myoung Seok, MD^b

Abstract

Rationale: There have been a few reported cases of subacute combined degeneration (SCD) associated with vitamin E deficiency, but the period of intestinal malabsorption was more than several years. We present a rare case of acute onset SCD that occurred in a relatively short period of several weeks with vitamin E deficiency related to small bowel obstruction.

Patient concerns: A 50-year-old woman had abdominal pain. A small bowel obstruction was suspected and conservative treatment was performed. She underwent bowel surgery after 2 weeks without any improvement. Following the operation, she was in a state of reduced consciousness. She was treated in an intensive care unit. Her consciousness level gradually recovered to alert in a week, but other symptoms such as ataxia, weakness on limbs, severe dysarthria, and dysphagia occurred. Since then, she had spent nearly 6 weeks in a bed-ridden state without improving.

Diagnosis: SCD associated with vitamin E deficiency was confirmed by laboratory investigations, electrophysiologic test, and whole spine magnetic resonance imaging scans.

Interventions: For vitamin E supplementation, she was administered a dose of 1200 mg/d. Physical therapy was focused on strengthening exercise, balance, and walker gait training. Occupational therapy was focused on activities of daily living training and dysphagia rehabilitation.

Outcomes: After 6 weeks, her muscle strengths and functional level were substantially improved. The vitamin E level was recovered to normal range.

Lessons: This case suggests that if neurological symptoms occur in patients with intestinal obstruction, clinicians need to consider a deficiency of micronutrients such as vitamin E and vitamin B12. Patients with short clinical courses suffer less neurological damage and achieve faster recovery.

Abbreviations: APCT = abdominopelvic computed tomography, BMI = body mass index, MRC = Medical Research Council, MRI = magnetic resonance imaging, SCD = subacute combined degeneration.

Keywords: ataxia, intestinal obstruction, malabsorption, subacute combined degeneration, vitamin E deficiency

Editor: N/A.

This research was supported by the Soonchunhyang University Research Fund.

The authors have no conflicts of interest to disclose.

^a Department of Physical Medicine and Rehabilitation, Soonchunhyang University Cheonan Hospital, Soonchunhyang University College of Medicine, Cheonan, Republic of Korea, ^b Department of Neurology, Soonchunhyang University Cheonan Hospital, Soonchunhyang University College of Medicine, Cheonan, Republic of Korea.

* Correspondence: Yuntae Kim, Department of Physical Medicine and Rehabilitation, Soonchunhyang University Cheonan Hospital, Soonchunhyang University College of Medicine, 31 Suncheonhyang 6-gil, Dongnam-gu, Cheonan-si, Chungcheongnam-do, Republic of Korea (e-mail: simon108@naver.com).

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How to cite this article: Jung JB, Kim Y, Oh K, Kim SA, Doh JH, Oh HJ, Seok JM. Subacute combined degeneration associated with vitamin E deficiency due to small bowel obstruction. *Medicine* 2019;98:36(e17052).

Received: 19 March 2019 / Received in final form: 8 July 2019 / Accepted: 12 August 2019

<http://dx.doi.org/10.1097/MD.0000000000017052>

1. Introduction

Subacute combined degeneration (SCD) involves degeneration of the posterior and lateral columns, which may include limb numbness, weakness, and gait disturbance.^[1,2] It is usually caused by vitamin B12 deficiency, and is caused in rare cases by vitamin E, folic acid, and copper deficiency.^[3] Among them, vitamin E deficiency involves not only peripheral nerves but also the spinocerebellar long tract.^[4] It may be caused by chronic fat malabsorption associated with cystic fibrosis, hepatic cholestasis, and abetalipoproteinemia.^[5] In addition, intestinal obstruction can cause severe malabsorption, which can lead to several micronutrients deficiency, such as vitamin E deficiency.^[6,7]

There have been a few reported cases of SCD due to vitamin E deficiency, but the period of intestinal malabsorption in one case was >20 years.^[3] In another reported case of vitamin E deficiency associated with malabsorption, the period of intestinal malabsorption was >4 years.^[5] However, there has been no reported case of SCD in just 2 months after intestinal obstruction. In this case report, we present a rare case of acute onset SCD in an adult with vitamin E deficiency associated with small bowel obstruction.

2. Case presentation

The patient was a 50-year-old woman living in China who was engaged in design work. She underwent surgery for a small bowel obstruction in China in 2016, and then returned to her normal life while maintaining a body mass index (BMI) of 18.75 kg/m². She had abdominal pain in early February 2018. A small bowel obstruction was suspected and conservative treatment was performed in Chinese hospital. However, she did not show any improvement for 2 weeks, so she underwent the surgical treatment of adhesiolysis and small intestinal suture. Following the operation, she was in a state of reduced consciousness and continued to have a fever. She was treated in an intensive care unit. Her consciousness level recovered to alert in a week, but other symptoms such as ataxia, limb weakness, and dysarthria gradually developed over several weeks. She had spent nearly 6 weeks in a bed-ridden state without improving in China. She then came to Korea and was admitted to the neurology department of our hospital to determine the underlying cause.

A physical examination revealed that her cranial nerves were intact. Vision was normal, and pupil was 3 mm isocoric and reactive to light. Ocular movement, hearing, and tongue movements were normal. Facial sensation and symmetry were intact, but severe dysarthria and dysphagia were observed. According to the Medical Research Council (MRC) grade, the muscle strengths of bilateral upper extremities were 2/5 in proximal and 3/5 in distal muscles. The muscle strengths of bilateral lower extremities were 2/5 in both proximal and distal muscles. Deep tendon reflex test revealed increased biceps and triceps jerk, but decreased ankle jerk. She also showed decreased proprioception in bilateral lower extremities. A cerebellar examination showed bilateral abnormal findings in finger-to-nose and rapid alternative movement tests. Bilateral limb and truncal ataxia were observed as well. BMI was 14.45 kg/m², and severe malnutrition was suspected (Fig. 1). She had difficulty in voiding and so a Foley catheter was inserted.



Figure 1. Atrophic changes were observed in both lower limbs.

Complete blood count, chemistry, thyroid function, and lipid panel tests were normal except for mild leukocytosis and normocytic normochromic anemia. Serum electrolytes were within normal limits. Abdominopelvic computed tomography (APCT) showed distension in the colorectum, dilatation of duodenum first and second portion with abrupt narrowing between the aorta and superior mesenteric artery. Brain magnetic resonance imaging (MRI) revealed no specific findings. The electrophysiologic test showed sensorimotor peripheral polyneuropathy, mainly in the lower extremities. Somatosensory-evoked potential studies showed absent bilateral tibial nerves response, suggesting abnormalities in the dorsal column pathway. Whole spine MRI showed normal T1 and abnormal high T2 signal in the bilateral posterior and lateral horn of spinal cord at the C5 level (Fig. 2). The electrophysiologic test and whole spine MRI scans were consistent with SCD at the cervical spinal cord. Additional tests were conducted to determine the cause of the SCD. Laboratory investigations revealed serum ceruloplasmin of 21.7 mg/dL (normal: 16–45 mg/dL), serum copper of 70.69 ng/dL (normal: 64–134 ng/dL), serum vitamin B12 of 649 ng/dL (normal: 197–771 ng/dL), and serum vitamin E of 3.71 mg/dL (normal: 5.0–20.0 mg/dL). Serum vitamin E levels showed deficiency.

Finally, it was diagnosed as SCD associated with vitamin E deficiency. For vitamin E supplementation, she was administered a dose of 1200 mg/d. She was transferred to the physical and rehabilitation department 3 weeks after admission, as she could not sit up alone. She could independently maintain a sitting position in bed, but she could not overcome resistance. It was impossible for her to stand alone. Physical therapy was focused on sitting, standing balance, and walker gait training. Occupational therapy was focused on an upper extremities strengthening exercise and activities of daily living training.

She was on a soft diet, but an aspiration symptom was observed when ingesting liquid. A video fluoroscopic swallowing study showed during and postswallowing aspiration of semisolid and liquid materials with decreased laryngeal elevation and closure. Therefore, rehabilitation therapy for swallowing disorders was performed as well. The Foley catheter was removed and she attempted self-voiding, but there was no voiding sense. Clean intermittent catheterization was performed every 6 hours. In addition, neurogenic bladder was controlled with alpha blocker, anticholinergics.

After 6 weeks of vitamin E supplementation, her functional level was substantially improved. The muscle strengths of the bilateral upper and lower extremities were improved to 3/5 in the proximal muscles. She was able to gait on a walker with moderate assistance. Meals were available as a general diet, and liquid was supplemented with food thickener without aspiration symptom. She was able to urinate well without medication. Her vitamin E level improved to 17.34 mg/dL. She then returned to her hometown in China for continuous rehabilitation. Patient has provided informed consent for publication of the case. In our case, the patient accepted regular examinations and proved therapy, so the ethical approval was not necessary.

3. Discussion

We present a rare case of acute onset SCD in an adult with vitamin E deficiency associated with small bowel obstruction, which was improved with vitamin E supplement and rehabilitation. There have been cases reported of SCD caused by vitamin E deficiency. For example, Harding et al reported that a 57-year-old

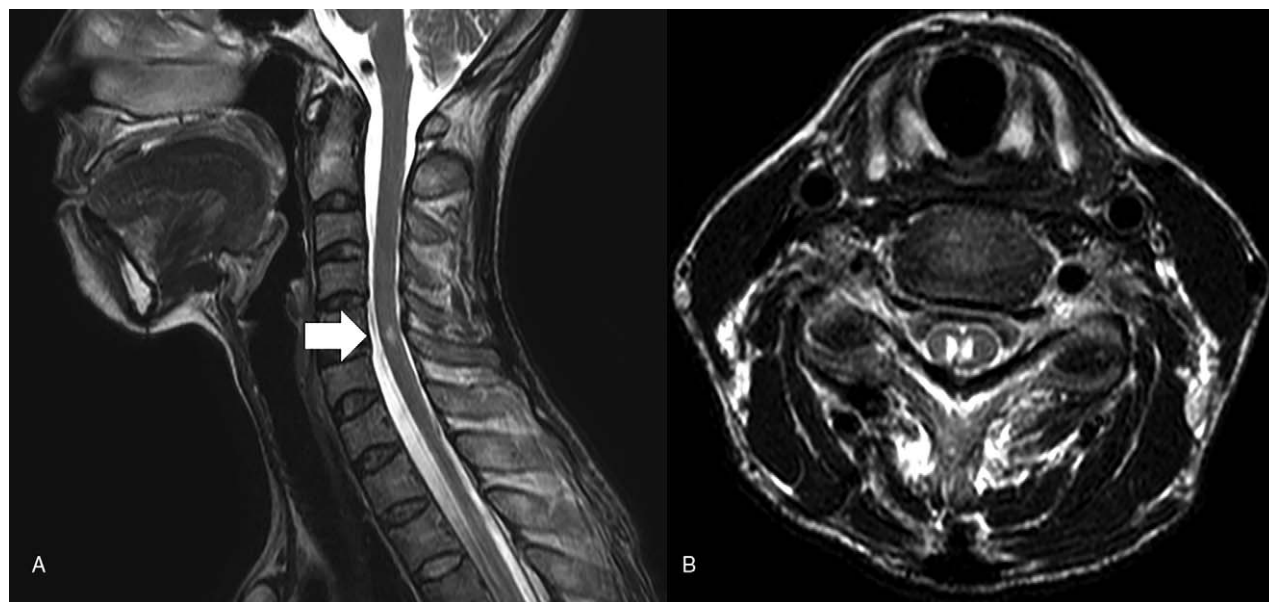


Figure 2. Sagittal (A) and axial (B) T2-weighted magnetic resonance imaging of the cervical spine shows high signal in bilateral posterior and lateral horn of spinal cord, C5 level (arrow).

woman and 41-year-old man had SCD secondary to chronic intestinal malabsorption.^[3] They each had a progressive neurologic disorder for >20 years, but did not know the cause. Finally, they were each diagnosed as SCD due to vitamin E deficiency, and improved clinically and electrophysiologically following oral therapy with vitamin E. Furthermore, Federico et al reported that a 32-year-old woman underwent a bowel resection due to subacute bowel obstruction.^[5] After 4 years, symptoms such as ataxia and general weakness were observed, which had worsened for about 17 years. Doctors diagnosed a vitamin E deficiency and symptoms improved after supplementing vitamin E for 2 months. The previous cases of SCD due to vitamin E deficiency usually involved a long period of intestinal malabsorption. Gabasi et al reported an average of 13.7 years to develop symptoms of vitamin E deficiency.^[8] In this case, the patient had a vitamin E deficiency associated with small bowel obstruction in a relatively short period of several weeks, leading to SCD.

Patients with SCD present the following clinical features; limb numbness (86.8%), limb weakness (67.6%), gait disturbance (63.2%) and bladder and/or bowel dysfunction (14.7%).^[1] The most commonly involved region is the cervical (70%) and thoracic spine (53.3%).^[1] The symptoms of the patient were consistent with the typical symptoms of SCD.

Vitamin E deficiency can have several causes. Naturally, it can be caused by abetalipoproteinemia^[9,10] and the mutation of α -tocopherol transfer protein gene.^[11,12] For acquired cases, it can be caused by chronic malabsorption associated with metabolic syndrome,^[13] cholestatic liver disease,^[14] cystic fibrosis,^[15] and extensive intestinal resection.^[3,16] The patient did not undergo intestinal resection in China, but received adhesiolysis and small intestinal suture for small bowel obstruction. APCT showed distension in the colorectum, dilatation of duodenum first and second portion after 6 weeks of bowel surgery. Although bowel surgery was performed, adhesiolysis seemed to be unable to resolve obstruction sufficiently. Intestinal obstruction can also cause severe malabsorption.^[6,7] The patient's BMI decreased from

18.75 to 14.45 kg/m², and we assumed that the patient suffered from severe malabsorption with poor oral intake in a short time.

Diagnosis of the disease can be made based on low levels of serum E in combination with clinical features.^[17] These tests help to early diagnosis, which is important for the prognosis of patients. The early diagnosis of micronutrient deficiency, such as vitamin E, along with replacement as soon as possible can stop disease progression and improve ataxia, limb weakness, and dysarthria.^[18,8] There is a report that the vitamin E level is selectively low in patients with short bowel syndrome who underwent parenteral nutrition.^[19] Therefore, it is recommended to monitor micronutrients including serum α -tocopherol in the case that malabsorption syndromes are suspected.^[20]

For malabsorption syndromes, vitamin E supplementation was considered to be beneficial.^[4] Ataxia with vitamin E deficiency is responsive to vitamin E supplementation at 800 to 1500 mg/d.^[21] We decided to supplement vitamin E, and her symptoms were shown to be improved after 6 weeks of vitamin E therapy. When supplementing vitamin E, it is necessary to regularly measure the plasma vitamin E concentration every 6 months to maintain a normal range.^[21]

In this case, the cause of ataxia and limb weakness was not found for 6 weeks. As SCD occurred over a relatively short period, it was difficult to identify the cause in the early stage of the disease. If neurological symptoms occur in patients with intestinal obstruction, clinicians need to consider a deficiency of micronutrients such as vitamin E and vitamin B12. If the micronutrient deficiency is confirmed, patients need to be treated as soon as possible. Patients with short clinical courses suffer less neurological damage and achieve faster recovery.

Author contribution

Conceptualization: Hye Jeong Oh, Jin Myoung Seok.

Supervision: Yuntae Kim, Hye Jeong Oh, Jin Myoung Seok, Soo A Kim, Kiyoung Oh, Joung Hyun Doh.

Writing – original draft: Jong Burm Jung.
Writing – review and editing: Yuntae Kim.
 Yuntae Kim orcid: 0000-0003-4063-4692.

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