LETTER TO THE EDITOR
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A Report of Congestive Myelopathy as a Result of Chronic Chylous Reflux Syndrome

Parichart Junpaparp, Ambiga Samiappan, Saranya Buppajarntham, George Newman

Department of Internal Medicine, Einstein Medical Center, Philadelphia, PA, USA

Dear Editor,

We would like to share an interesting case of congestive myelopathy involving a 26-year-old female with chronic gynecological chylous reflux disease who presented with bilateral lower extremities weakness, decreased sensation, and urinary incontinence of 2 months duration. A medical review revealed chronic vaginal discharge and lower-extremity swelling due to chronic chylous reflux disease. Upon examination, the patient had normal muscle strength in the upper extremities, motor power grades of 4 in the left lower extremity (LLE) and 1 in the right lower extremity (RLE), and loss of rectal tone. Sensory examination revealed decreased touch and vibratory sensation in both lower extremities. All cranial nerve and cerebellar functions were intact. Spinal MRI revealed T2-weighted hyperintense signals and atrophy of the thoracic cord with enhancing curvilinear T2-weighted hypointense signals within the thecal sac and flow voids anterior and posterior to the conus medullaris (Fig. 1). The findings were suggestive of a dural arteriovenous malformation (AVM). Surprisingly, subsequent spinal angiography did not reveal any evidence of either a dural AVM or a fistula. We therefore diagnosed this patient with congestive myelopathy secondary to venous drainage dysfunction as a result of chylous reflux syndrome.

Primary chylous reflux syndrome is a rare lymphatic disorder that is characterized by retrograde flow of chyle from its normal route. Hypothesized pathophysiologies include an incompetent lymphatic valvular system, lymphatic hypoplasia, and dilated incompetent megalymphatics. However, the definite pathophysiology remains unclear.[1,2] Approximately 20 cases of primary chylous reflux syndrome have been reported during the past century. The common manifestations are lymphedema, chyluria, and chylous genital discharge. Some patients may experience complications such as chylothorax, chyloperitoneum, and chylopericardium.[3] However, neurological deficits have never been reported in association with this condition.

This patient had been diagnosed with primary chylous reflux syndrome as a young child, and was conservatively managed with dietary control. At the age of 26 years she gradually developed venous congestive myelopathy or Foix-Alajouanine syndrome. Given that the most common cause of venous congestive myelopathy is spinal dural arteriovenous fistula, extensive investigations including MRI/MRA and spinal angiography were conducted.[4] The results were negative for concurrent vascular malformations. To date, only 14 cases of venous congestive myelopathy without concurrent vascular malformation have been reported,[5] none of which had underlying chylous reflux syndrome. Chyle is normally absorbed through the lacteals, drains into the thoracic duct, and empties into the venous system. In patients with chylous reflux disorder, the retrograde flow of lymph may contribute to retention of lymph within the pelvic cavity, with efflux into the vagina, causing vaginal discharge. These can ultimately lead to the development of aberrant vascular channels and can increase the lymph absorption through the venous system, as evidenced in the present patient by increased lower-extremity swelling. This can result in chylous reflux disorder eventually involving the spinal venous plexus or Batson venous plexus and causing chronic venostasis, contributing to venous congestive myelopathy.

The treatment of choice for primary chylous reflux disor-

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Correspondence
Parichart Junpaparp, MD, Department of Internal Medicine, Einstein Medical Center, 5501 Old York Road, Philadelphia, PA 19141, USA
Tel +1-310-382-6703, **Fax** +1-215-456-7926
E-mail junpapap@einstein.edu

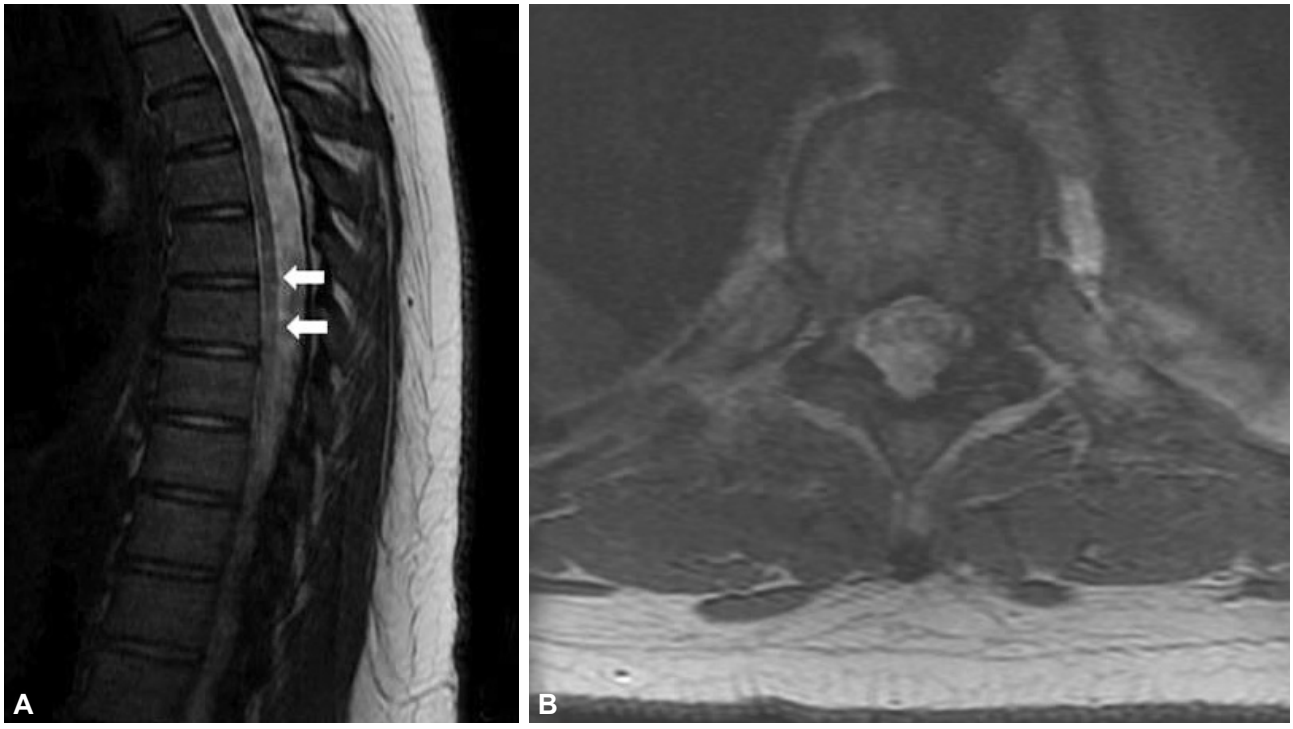

Fig. 1. Thoracic spine T2-weighted MRI. Sagittal view (A) and cross-sectional view (B) showing hyperintense signal and atrophy of the thoracic spinal cord (arrows).

der remains lymphangiography- or lymphoscintigraphy-guided surgery. Removal of the pelvic megalymphatics can theoretically redirect the chylous lymphatic flow toward the thoracic duct.[6] However, there is no recommendation regarding the treatment of venous congestive myelopathy resulting from chylous reflux syndrome. Our patient has been refusing surgery since she was young, and at this admission again selected conservative treatment with rehabilitation. The paraplegia was slightly improved in approximately 8 weeks (motor power grades: 5/5 in LLE and 2/5 in RLE) and the patient could finally ambulate with crutches.

Conflicts of Interest

The authors have no financial conflicts of interest.

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