대한임상신경생리학회지 7(1):49~51, 2005 ISSN 1229-6414

길랑-바레 양상으로 발현한 무통성 대동맥 박리 1례

인제대학교 의과대학 일산백병원 신경과

오은진 · 정상욱 · 박종관 · 홍근식

Painless Aortic Dissection Simulating Guillain-Barré Syndrome

Eun-Jin Oh, M.D., Sang-Wuk Jeong, M.D., Jong-Kwan Park, M.D., Keun-Sik Hong, M.D.

Department of Neurology, Inje University College of Medicine, Ilsan Paik Hospital, Goyang, Korea

A 61-year-old man with an antecedent febrile illness presented with progressive flaccid paraparesis, but no sensory or sphincter involvement. Magnetic resonance imaging (MRI) of the spine was negative and nerve conduction study (NCS) showed the absence of F-waves in his legs, suggesting Guillain-Barré syndrome (GBS). However, abdominal pain after admission led to the consideration of the spinal cord ischemia secondary to aortic dissection confirmed by computed tomography. We report the rare condition of painless aortic dissection simulating GBS.

Key Words: Paraparesis, Aortic dissection, Guillain-Barré syndrome

The aortic dissection may result in spinal cord ischemia and subsequent paraparesis. However, the presentation of aortic dissection with both paraparesis and the absence of characteristic pain is very unusual. Our patient had the progressive flaccid paraparesis without characteristic pain. The initial impression was GBS because of the antecedent history of febrile illness, the absence of sensory or sphincter involvement, the negative spine MRI, and the loss of F-wave responses in initial electrophysioloic studies. The diagnostic pitfall will be discussed.

Case Report

A 61-year-old man presented with paraparesis noted on the day of admission. He did not complain of chest or back pain. On past medical history, he had no history of hypertension and his blood pressure was normotensive. Two months ago, he had the history of flank pain with spontaneous remission. Two weeks prior to admission, he had been investigated for febrile illness of unknown origin. However, diagnostic studies did not reveal any remarkable findings and fever was subsided spontaneously. On neurologic examination, the motor power was grade $4^{+}/4^{+}$ in both lower extremities. The power of both upper extremities was normal. Deep tendon reflexes were decreased in his legs and plantar responses were down going. The sensation of in his legs and the sphincter function were normal. On the day of admission, MRI of the whole spine was obtained to investigate the spinal cord lesion, but did not reveal any remarkable findings (Fig. 1A). The chest X-ray showed no remarkable findings. The initial NCS obtained on the day of admission showed the absence of F-waves in both legs. The amplitude of the compound motor action potential (CMAP) was decreased only in the left peroneal and posterior tibial nerve. The nerve conduction velocities of both lower extremities were normal. The NCS of both upper extremities were completely normal. The laboratory examination revealed mild leukocytosis (10,560/ul) and elevat-

Address for correspondence
Keun-Sik Hong, M.D.
Department of Neurology, Ilsan Paik Hospital, College of medicine, Inje University, 2240 Daewha-dong, Ilsan-gu, Goyang-si, Gyeonggi-do, 411-706, Korea
Tel: +82-31-910-7277 Fax: +82-31-910-7368
E-mail: nrhks@ilsanpaik.ac.kr

ed ESR (46 mm/hr). The patient refused the CSF study. After admission, the paraparesis was slightly aggravated and we started intravenous immunoglobulin therapy under the impression of GBS. On the third hospital day, he complained of abdominal pain and physical examination revealed the abdominal bruit and absence of arterial pulsation in both lower extremities which had been ignored in the initial examination. The chest and abdominal computed tomography (CT) scan was performed under the impression of aortic dissection. The CT scan revealed the dissection of the descending aorta, involving from T8 to T9 and from L2 to the bifurcation of common iliac arteries (Fig. 1B, C). The immunoglobuin therapy was discontinued and beta-blocker was started. The follow-up NCS after 4 days did not show any significant interval changes except for the appearance of F-wave responses in the right peroneal and posterior tibial nerve and the improvement in the amplitude of CMAP in the left peroneal nerve. The paraparesis persisted with some fluctuation. The surgical intervention was consulted, but refused by the patient.

Discussion

In our patient, paraparesis might be caused by the spinal cord ischemia secondary to aortic dissection because it involved the lower thoracic and lumbar area and might interrupt the blood flow through the Adamkiewicz artery. The blood flow of thoracolumbar area has been known to be greatly dependant on the Adamkiewicz artery and this area may be vulnerable to ischemia in this situation. MRI is sensitive method to detect the

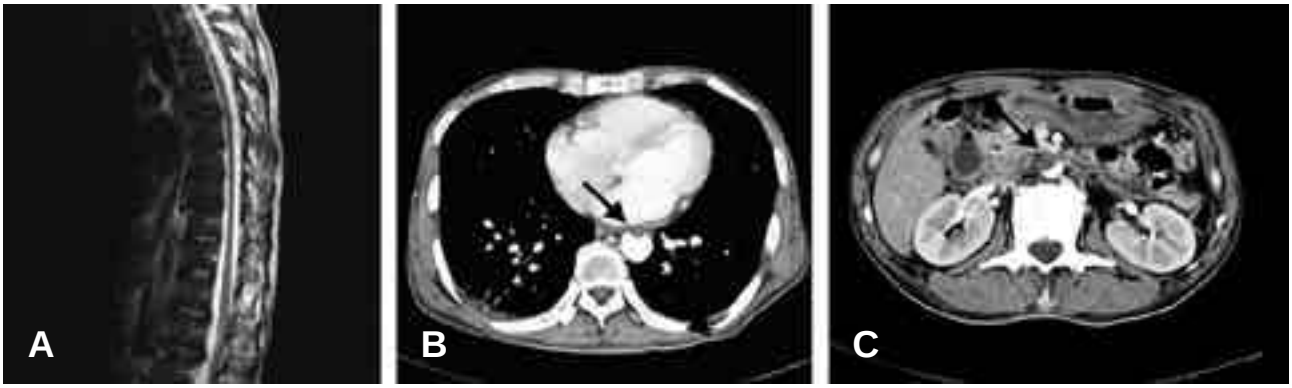

Figure 1. MRI of the whole spine did not reveal any remarkable findings in spinal cord (**A**). CT scan of the abdomen shows the dissection of the descending aorta (arrows), involving from T8 to T9 (**B**) and from L2 to the bifurcation of common iliac arteries (**C**).

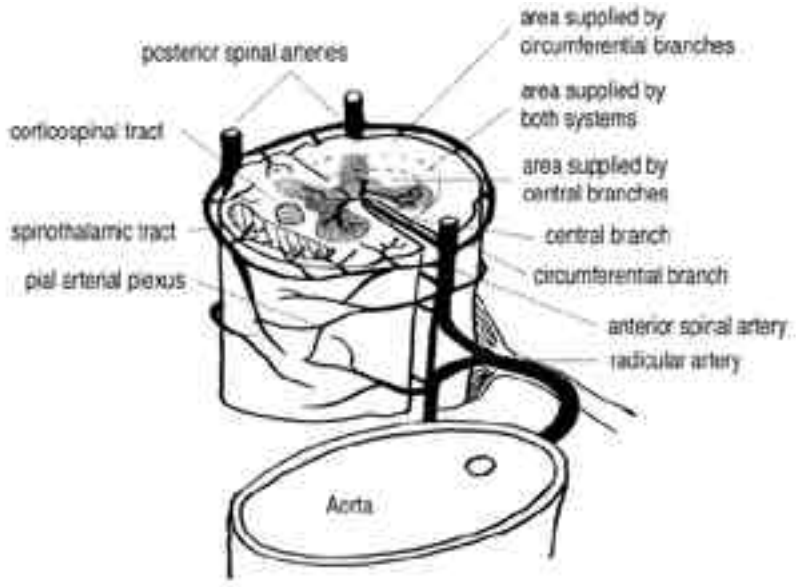

Figure 2. The intrinsic blood supply of the spinal cord.

spinal cord infarction, but could be negative in some cases with incomplete infarction or if obtained in early stage.[2,3]

Spinal cord infarction secondary to aortic dissection could be confined to the gray matter and adjacent white matter, in which cases there is motor weakness but no sensory involvement as in our patient. Previous pathologic studies showed the predominant involvement of gray matter and adjacent white matter with relative sparing of the peripheral white matter.[4] Intrinsic blood supply of the spinal cord could explain the selective vulnerability of these areas. The two systems consist of the intrinsic blood supply of the spinal cord (Fig. 2). The first system is the central branches and supplies the gray matter and adjacent white matter. The second system is the pial arterial plexus and supplies from one-third to one-half of the outer rim of spinal cord, including the spinothalamic tracts. The central branches arise from the anterior spinal artery which is reinforced by the radicular arteries originated from aorta. This reinforcement is crucial and could be seriously affected by the aortic dissection. However, the pial arterial plexus are formed by the circumferential branches from one anterior spinal artery and two posterior spinal arteries and interconnected by the anastomotic channels. Thus the pial arterial plexus is less affected by the aortic dissection and the spinal cord ischemia can present only with weakness but no sensory involvement. The branches of lumbar, iliolumbar, and lateral and median sacral arteries supply the sacral segment of spinal cord and also constitute the anastomotic loops with posterior spinal arteries. Thus the sacral area is relatively resistant to ischemia and this might be related to the sparing of sphincter function.

In our patient, the antecedent febrile illness commonly associated with GBS led us to considerable diagnostic confusion. In the literature, the cases of aortic dissection presenting with fever of unknown origin have been reported.[5,6] The subacute inflammatory response due to continuing dissection may be responsible for fever.

The initial NCS of our patient showed the absence of F-waves with normal nerve conduction velocities in both lower extremities. These findings are consistent with the early findings in patients with GBS.[7] However, we can rule out the possibility of GBS because the follow-up NCS showed the reappearance of F-waves and normal conduction velocities. It has been reported that the absence of F-wave can be the early findings and potential source of diagnostic error in acute myelopathy.[8,9] The F-waves represent the back-firing of the activated motor neurons in response to antidromic stimulation. The excitability of motor neurons is under the control of descending excitatory pathway and local inhibitory system. Thus the increased local inhibition secondary to the lesion of descending excitatory pathway may be the one of postulated mechanisms of decreased F-wave response in acute myelopathy.[9] Direct ischemic injury of the spinal motor neuron could also contribute the absence of F-waves.

In this patient, thorough examination of abdominal auscultation and arterial pulsation might have avoided the delay in diagnosis and the unnecessary therapy.

Acknowledgements

This work was supported by grant (00012001239) from Inje University, 2001.

REFERENCES

1. Satran R. Spinal cord infarction. *Stroke* 1988;19:529-532.
2. Fortuna A, Ferrante L, Acqui M, Trillo G. Spinal cord ischemia diagnosed by MRI. Case report and review of the literature. *J Neuroradiol*. 1995;22:115-122.
3. Castro-Vilanova MD, de Toledo M, Mateos F, Simon R. Spinal cord infarction/ischemia. *Rev Neurol*. 1999;29:977-980.
4. Hughes JT. Vascular disorders of the spinal cord. In: Vinken PJ, Bruyn GW, Klawans HL, eds. Handbook of Clinical Neurology, Vol. 55. Amsterdam: Elsevier, 1989;95-106.
5. Giladi M, Pines A, Averbuch M, Hershkoviz R, Sherez J, Levo Y: Aortic dissection manifested as fever of unknown origin. *Cardiology* 1991;78:78-80.
6. Schattner A, Klepfish A, Caspi A: Chronic aortic dissection presenting as a prolonged febrile disease and arterial embolization. *Chest* 1996;110:1111-1114.
7. Ropper AH, Wijdicks EFM, Shahani BT: Electrodiagnostic abnormalities in 113 consecutive patients with Guillain-Barre syndrome. *Arch Neurol* 1990;47:881-887.
8. Syme JA, Kelly JJ: Absent F-waves early in a case of transverse myelitis. *Muscle & Nerve* 1994;17:462-465.
9. Marras C, Midroni G: Transient absence of F-waves in acute myelopathy: a potential source of diagnostic error. *Electromyogr Clin Neurophysiol* 2000;40:109-112.