

# Paraparesis as the Major Initial Presentation of Aortic Dissection: Report of Four Cases

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**Abstract-** Aortic dissection, which typically manifests as sudden tearing or migratory pain, is a well-known medical emergency. However, in 5% of aortic dissection patients, there is no pain. In these patients, the diagnosis depends on the development of neurologic complications. After analyzing the initial symptoms of a series of patients with aortic dissection, we found 4/211 (1.9%) patients suffered from paraparesis. We suggested that the mid- or low thoracic cord be most vulnerable site during acute aortic dissection. This report highlighted the importance of considering the diagnosis of aortic dissection in a patient with a history of acute onset of transient or permanent neurological symptoms in the lower limbs. Whether paraparesis can be an indicator of the prognosis of aortic dissection requires further researches.

**Key Words:** Aortic dissection, Paraparesis, Spinal cord ischemia

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## INTRODUCTION

Acute aortic dissection is a catastrophic disease and requires very prompt treatment. In 1972, Anagnostopoulos et al.<sup>(1)</sup> reviewed 963 cases and found the mortality rate to be 50.1% within 48 hours of onset and 79.9 % within 2 weeks. Patients with aortic dissection typically present with sudden onset of catastrophic, migratory, and tearing pain localized in the substernal, interscapular, and mid-back areas. It can also radiate to the abdomen, lower back, flank, neck or extremities, thus mimicking myocardial infarction, cholelithiasis, pancreatitis, or even renal colic. However, in approxi-

mately 5% of aortic dissection patients there is no pain<sup>(2)</sup>. For these patients, neurologic symptoms, including an altered mental status, numbness in limbs, hemiparesis, paraparesis, and Horner's sign, become important clues for diagnosis. The incidence of neurological deficits in painless aortic dissection ranges widely from 7.8 to 46%<sup>(3)</sup>. In a large-scale analysis including 1805 patients with painless aortic dissection, 4.2 % (2 to 8 %) presented with acute paraparesis or paraplegia<sup>(4)</sup>. However, these data may underestimate the true conditions because some critically ill patients could not be adequately examined.

On the other hand, in clinical practice, an elderly

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patient with the initial presentation of acute paralysis and at a sensory level is most readily diagnosed with spinal cord compression, without taking other possibilities into consideration. Aortic dissection can easily be missed when the history is difficult to clarify and pain is absent. In this report, we describe four aortic dissection patients with an initial presentation of paraparesis and also propose the possible pathophysiology.

To analyze the initial symptoms of aortic dissection, we searched the admission registration database of National Cheng Kung University Hospital over the past 5 years according to the key words of aortic dissection and paraparesis. The charts and images were carefully reviewed. The inclusion criteria were an acute episode of paraparesis that was localized to the spinal cord and a definite aortic dissection evidenced by image studies. The exclusion criteria were recurrent episodes of aortic dissection, previous paraparesis, and quadriplegia due to other causes. Two hundred and eleven patients were collected. As shown in Table 1, four patients presented acute paraparesis as the initial symptom.

## CASE REPORTS

### Case 1

HSY, a 55-year-old man, had hypertension for three years without regular control. He suffered from a sudden onset of severe low back pain while sitting in a chair. The pain ascended to the upper back in one minute. He tried to stand up but failed due to bilateral lower limb weakness, though he still could move his legs in the horizontal plane. He also felt numbness in his entire lower limbs about ten minutes later. Initial neurological examination revealed normal cognitive and cranial nerve functions. Both the sensory and motor components of the

upper limbs were normal. Muscle power of the lower extremities was 2/0 (R/L) by MRC grading and the deep tendon reflexes (DTRs) were all areflexic. The pinprick test revealed a symmetric L1-2 sensory level. The vibration test was moderately impaired in the lower limbs, whereas the joint position sense was intact. Sphincter function was impaired. The thoracic-lumbar (T-L) spine X-ray showed only mild degenerative change in the L4-5 vertebrae. T-L spine magnetic resonance imaging (MRI) with gadolinium (Gd) showed an abnormal signal at T10 to the conus and relative dilation of the cord (Fig. 1). An abnormal signal was also noted near the aortic wall. Aortic dissection and myelopathy was highly suspected. Thus, chest and abdominal computed tomography (CT) with contrast medium was arranged immediately. The examination showed a dissecting aneurysm involving the ascending and descending aorta (Fig. 2). The patient refused an operation and he was admitted to the cardiac intensive care unit for conservative treatment. The vital signs and general medical condition were stabilized but paraplegia persisted. His condition did not change over the following 3 years.

### Case 2

CYF, a 64-year-old female, had hypertension for five years without regular control. She denied other underlying diseases. She was suddenly unable to stand up due to weakness of the lower limbs. She denied any accompanying back pain, sensory abnormality, dizziness, or upper limb deficit, except for mild and transient nausea. The paraplegia lasted for only 10 minutes and then completely resolved. The results of neurological examinations at the emergency room (ER) were normal. No similar episode recurred thereafter. T-L spine MRI with Gd showed an intact spinal cord but an abnormal thoracic aorta wall. Chest and abdomen CT with contrast medium proved an aortic dissection at the ascending aorta, and graft replacement of the ascending aorta was performed immediately. She did not have any post-operational complication and was discharged several days later. She was stable and did not have any recurrent paraparesis during the recent 5 years.

**Table 1.** Major initial symptoms of 211 cases of aortic dissection

Initial symptom	Cases (%)
Migratory pain	39 (18.5)
Chest pain	130 (61.6)
Abdominal pain	40 (19.0)
Acute paraparesis	4 (0.9)

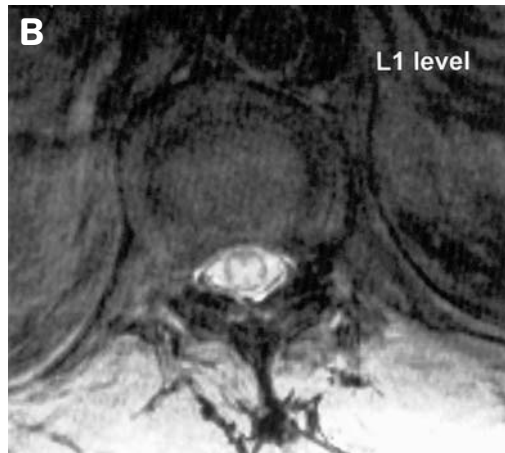


Figure 1. Lumbar spine MRI with and without Gd showed (A) an abnormal signal at the central part of the cord from T11 to the conus medullaris on a sagittal view, and (B) in a transverse view (L1 level), a relative dilation of the gray matter at the cord level of L1 on a transverse view.

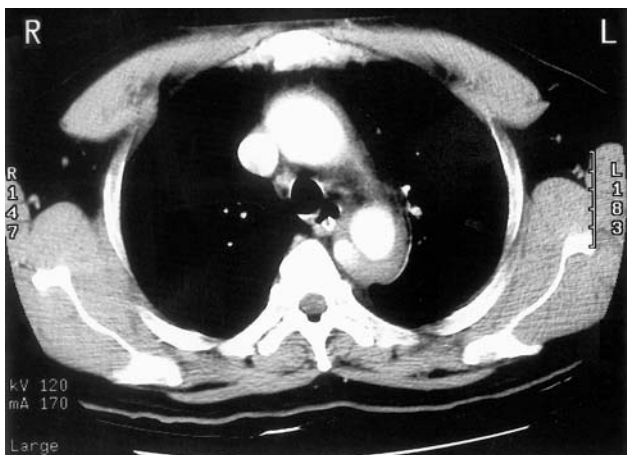


Figure 2. CT of the chest and abdomen with contrast medium revealed an intramural hematoma at the ascending and descending aorta on a transverse view.

### Case 3

ZCS, a 67-year-old male, had hypertension with a history of congestive heart failure. The hypertension has been under regular medical control for eight years. He complained of a sudden onset of bilateral lower limb weakness and numbness while working on the farm. The

tingling sensation affected both lower limbs. The patient did not report any ascending of numbness, incontinence, or back pain. Initially, only the left lower limb was weak. Five minutes later, the right leg also became equally weak. At our ER, his consciousness and cognitive functions were normal. There was no motor and sensory involvement in the upper limbs. Muscle power of the lower limbs was 2/2 (R/L), and the DTRs were all areflexic. The superficial abdominal reflex decreased in the area below the umbilicus. The cremaster reflex disappeared and the anal tone decreased. The pinprick test revealed a symmetric sensory level at T10-11, and the joint position sense was equally impaired in both lower limbs. Chest CT with contrast medium revealed a type I aortic dissection. While waiting for the results of CT, his blood pressure dropped abruptly and he became unconscious. Though emergent subxyphoid pericardiectomy was performed, the patient expired 4 hours later.

### Case 4

KCH, a 68-year-old male, had hypertension, diabetic mellitus, and old myocardial infarction with regular medical control. He had a sudden onset of sharp abdomi-

nal pain that radiated to the chest and upper back (interscapular area) while he was sitting and chatting. Acute weakness and hypoesthesia of the lower limbs appeared at the same time. His consciousness was clear. The cranial nerves and upper limbs were spared. Muscle power of lower limbs was 0/0, and DTRs were all areflexic. The pain and temperature sensations were completely impaired below T4, and the joint position and vibration sensations showed only a mild decrease from toe to knee joints. Chest CT with contrast medium revealed a distal type aortic dissection. The spinal cord MRI was not available. His paraplegia remained when his consciousness was still clear. He was then admitted to the intensive care unit due to a recurrent myocardial infarction, and then died of sepsis 3 months later.

## DISCUSSION

We described four cases of aortic dissection with acute paraparesis as the major initial symptom. The aortic dissection, revealed by contrast CT study, was located in the thoracic segment in one case and involved both the thoracic and abdominal segments in the other three cases. Neurological examination indicated that the lesion was at the spinal cord level, T10, T11 and T4 respectively in three cases and not localizable in one case. Spinal cord MRI was performed in two patients (cases 1 and 2) and showed a lesion from T10 downward in case 1 and no lesion in case 2. Two cases (cases 3 and 4) expired, one case (case 1) had persistent neurological deficits, and one case (case 2) recovered promptly and completely (Table 2). In case 1, the findings that the vibration sensation was impaired while the joint position sense was intact seem contradictory. One explanation is that manual testing of the vibration sensation could be graded

by changing the magnitude of vibration amplitude and was, thus, more sensitive. The second possibility is that joint position sense is mainly carried by the dorsal columns while the vibration sense is conveyed in several pathways. Because the anterior part of the spinal cord is more frequently involved, it is possible that the two mentioned senses were affected to different degrees. The third possibility was that the deficit in vibration sense might have been due to an unnoticed existing peripheral neuropathy.

Many differential diagnoses have to be taken into consideration with acute paraparesis. Among them, thromboembolism of the spinal artery, shock, epidural or subdural hematoma, aortic dissection with spinal cord involvement, rupture of arteriovenous malformation, and tumor bleeding are the most frequent etiologies<sup>(5,6)</sup>. To evaluate an acute spinal cord lesion, an MRI is usually first arranged. Not only the spinal cord but also the aorta must be well evaluated. On the other hand, depending on the site of dissection, aortic dissection may result in many neurological sequelae, such as cerebral ischemia, ischemic peripheral neuropathy, and spinal cord infarction. Patients with acute aortic dissection can develop either anterior spinal artery syndrome or complete transverse myelopathy, depending on the degree of disruption to spinal circulation<sup>(7)</sup>. In one previous study of 44 patients with spinal cord infarction, two (4.5%) had acute aortic dissection as the cause of their spinal cord ischemia<sup>(8)</sup>. Furthermore, Horner's syndrome can occur if an aneurysm compresses the superior cervical ganglion.

The incidence of aortic dissection with paraparesis in our study was 1.9 % (4/211), which is slightly lower than the rate reported in one previous study (2-8%, total 1805 patients)<sup>(4)</sup>. The incidence of painless aortic dissection was 1.0% (2/211), which is also lower when com-

**Table 2.** Initial symptoms, types of aortic dissection, sensory levels and durations of neurological deficits in our four patients

Case no.	Sensory level	Type of aortic dissection	Initial symptoms	Neurological deficits
1	T10 #	Type I	Paraparesis, migratory pain	Persistent
2	Nil	Type I	Paraparesis	Transient
3	T11	Type I	Paraparesis	Persistent
4	T5 #	Type III	Paraparesis, chest pain	Persistent

# Cases 1 and 4: manifested as anterior spinal artery syndrome

pared to a previous study (5%)<sup>(2)</sup>. The discrepancy may be related to the small case number in our study. Painless aortic dissection with paraparesis was first reported in 1986<sup>(9)</sup>. The physical signs may be subtle, and the chest X-ray may be normal. The sudden onset of weakness and paraesthesia results from interruption of blood flow to the spinal cord. Lower extremity motor and sensory loss may be the only symptoms. Zull and Cydulka emphasized that an acute aortic dissection should be assumed if a patient with an acute paraparesis had one of the following symptoms or signs, e.g., chest pain, migratory pain, pulse deficit, aortic insufficiency murmur, cardiac temponade, stroke, altered consciousness, and aortic bruits. They also suggested that aortic dissection should be considered in patients with paraparesis and hypotension, unless the etiology was known to be otherwise<sup>(10)</sup>.

The simultaneous existence of acute paraparesis and aortic dissection does not automatically confirm any correlation between the two conditions. In fact, there are no universally accepted criteria that prove or disprove the relationship. The causal relationship was assumed in most of the past reports. In the present report, because the sharp chest pain and the paraparesis occurred almost simultaneously in two patients (cases 1 and 4), the myelopathy in these cases could be strongly correlated to the aortic dissection. In the other two cases, however, only paraparesis was noted with no painful sensation. The aortic dissection might be an incidental finding. Since no other causes could be found, we favored that aortic dissection causing insufficient blood supply at the spinal cord level as being the best explanation for the symptoms.

The pathophysiology of aortic dissection and the associated hemodynamic change is the key to understand the relationship between the aortic dissection and spinal cord ischemia. Dissection of the aorta begins with a tear in the intimal layer. This tear permits blood to enter the aortic wall, creating an intramural hematoma progressing distally in the aorta and its branches<sup>(11)</sup>. A common site for the initiation of an intimal tear is at the proximal portion of the ascending aorta due to the thrust of blood ejecting from the left ventricle. At least three hypotheses concerning ischemia of the spinal cord in the aortic dis-

section have been proposed<sup>(3,12,13)</sup>. All are based on the temporary loss of blood flow to the spinal cord. First, the dissecting blood flow stretches, narrows, and completely or partially occludes the intercostal arteries. Further stretch causes extensive tearing of intercostal arteries, which causes permanent spinal cord damage. The other hypotheses include spasm of the intercostal arteries and temporary obstruction of the orifice of the intercostal arteries. The latter two mechanisms are more appropriate for transient ischemia in the spinal cord. The anterior spinal artery syndrome, which spares the posterior column, is more frequent than the posterior spinal artery syndrome. In our cases, two cases presented as anterior spinal artery syndrome, while the other two cases indicated complete transverse myelopathies.

In the present report, two cases were diagnosed as "painless" aortic dissection. There are three possible mechanisms for the lack of pain. First, the dissecting hematoma only causes the intima to bulge inward and re-enters the true aortic lumen without displacing the adventitia outward and causing pain. Second, the involvement of cerebral vessels may dull the patients' perception of pain. Third, the loss of visceral and spinothalamic perception of pain caused by the preceding severe spinal ischemia may dull the perception of pain. The aortic dissection without any painful sensation may mislead the diagnostic process. Thus acute aortic dissection must always be considered as one of the differential diagnoses for all patients with a sudden onset of painless paraplegia.

Although the proximal part of aorta is more prone to dissection in theory, distal dissections appear to have a slightly greater incidence of spinal cord ischemia<sup>(14)</sup>. It is the branches in this aortic segment, in particular the artery of Adamkiwicz which is sheared off in aortic dissection. When the interruption of blood flow occurs, the mid-thoracic cord is the most vulnerable part because it is the watershed zone between the blood supplies of the upper and lower cord. In our cases, three were localized at T10, T11, and T5 level respectively, and the other was unable to be localized due to the transience of the symptoms. The origin of the artery of Adamkiwicz is variable (T6-T12 or L1-3), and arises more frequently on the

left from an inferior intercostal or a superior lumbar artery. This variability might explain the "high" paraparesis in the case 4. In a previous study<sup>(15)</sup>, painless aortic dissection always involved the descending thoracoabdominal aorta, where the artery of Adamkiewicz originates.

In conclusion, this report highlights the importance of considering the diagnosis of aortic dissection in patients with an acute onset of transient or permanent neurological symptoms in the lower limbs. Pain can be absent. Whether paraparesis is an indicator of prognosis in aortic dissection requires further research.

## REFERENCES

1. Anagnostopoulos CE, Prabhakar MJ, Kittle CF. Aortic dissections and dissecting aneurysms. *Am J Cardiol* 1972;30:263-73.
2. Slater EE, DeSanctis RW. The clinical recognition of dissecting aortic aneurysm. *Am J Med* 1976;600:625-33.
3. Tanaka T, Uemura K, Sugiura, et al. Transient paraplegia caused by acute aortic dissection. *Neurol Med Chir (Tokyo)* 1990;30:54-8.
4. Zull DN, Cydulka R. Acute paraplegia, a presenting manifestation of aortic dissection. *Am J Med* 1998;84:765-70.
5. Thompson GB. Dissecting aortic aneurysm with infarction of the spinal cord. *Brain* 1956;79:111-8.
6. Howell JM, McFarling DA, Chisholm CD. Ischemic injury to the spinal cord as a cause of transient paraplegia. *Am J Emerg Med* 1987;5:217-9.
7. Gilles FH, Nag D. Vulnerability of human spinal cord in transient cardiac arrest. *Neurology* 1971;21:833-7.
8. Cheshire WP, Santos CC, Massey EW, et al. Spinal cord infarction: etiology and outcome. *Neurology* 1996;47:321-30.
9. Greenwood WR, Robinson MD. Painless dissection of the thoracic aorta. *Am J Emerg Med* 1986;4:330-3.
10. Rosen SA. Painless aortic dissection presenting as spinal cord ischemia. *Ann Emerg Med* 1988;17:840-2.
11. Joo JB, Cummings AJ. Acute thoracoabdominal aortic dissection presenting as painless, transient paralysis of the lower extremities: a case report. *J Emerg Med* 2000;19:333-7.
12. Moersch FP, Sayre GP. Neurologic manifestations associated with dissecting aneurysm of the aorta. *JAMA* 1950;144:1141-8.
13. Donovan EM, Seidel GK, Cohen A. Painless aortic dissection presenting as high paraplegia: a case report. *Arch Phys Med Rehabil* 2000;81:1436-8.
14. Prendes JL. Neurovascular syndromes of aortic dissection. *Am Fam Physician* 1981;23:175-9.
15. Inamasu J, Hori S, Yokoyama M, et al. Paraplegia caused by painless acute aortic dissection. *Spinal Cord* 2000;38:702-4.