

Brief Communication

Clinical diagnosis of aortic dissection is characterized by acute spinal cord damage

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Aortic dissection (AD) is a rare, life threatening condition. Neurological complications, such as paraplegia, as a presenting manifestation of AD are even rarer.¹ The incidence rate of AD is 5-30 cases per million people each year. The mortality rate of AD is 3.2/100,000 people per year.² Aortic dissection presenting as acute spinal cord damage is often misdiagnosed as other primary spinal diseases, such as acute myelitis or spinal hemorrhage, leading to a delay in its diagnosis and an increase in mortality. Currently, a limited number of studies have been systematically performed on AD presenting as acute spinal cord damage. Here, we carried out retrospective research on 9 cases to study the clinical manifestations and imaging features of AD primarily presenting as acute spinal cord damage. The study of clinical and imaging features of AD will increase the knowledge of clinicians in understanding AD primarily presenting as acute spinal cord damage, which will help to prompt early diagnosis and reduce mortality.

The study group included 9 cases of AD primarily presenting as acute spinal cord damage at the emergency department of the First Affiliated Hospital in Liaoning Medical College, Jinzhou, China during the period between January 1998 and January 2007. The information was obtained from the in-hospital patients' medical records. The symptoms taken into account were typical symptoms of transverse spinal cord damage, sudden onset of pain, and other complications such as acute pancreatic necrosis, acute skeleton muscle ischemic necrosis, acute renal failure, and so on. Five of the 9 patients were male, and the other 4 were female. Ages ranged from 32-70 years, and the mean age of the patients was 51 years. All of the 9 patients had hypertension. Two patients had coronary artery disease, 4 had diabetes, and one had cardiac tamponade. Using the classification of DeBakey, among 9 cases of AD, 3 patients were type I, the youngest was 44-years old; 6 patients were type III, the youngest was 32-years old. The Statistical Package for Social Sciences (SPSS Inc, Chicago, IL, USA) version 17.0 was used for data analysis. Informed consent was obtained from each subject, after receiving approval of the experimental protocol by the local human ethics committee.

Using the classification of DeBakey, AD patients were designated as type I to III according to the site of the tear and the extent of dissection. For type I classification, the tear occurred at the ascending aorta and the dissection extended distally to the descending aorta. For type II classification, the dissection was limited

within the ascending aorta and aortic arch. For type III classification, the tear occurred in the descending artery and the dissection extended along the descending artery into the thoracic and abdominal aorta. The diagnostic criteria for hypertension were systolic pressure ≥ 140 mm Hg and/or diastolic ≥ 90 mm Hg. Acute kidney failure was considered if the glomerular filtration rate (GFR) was ≤ 20 mL/min, and serum creatinine (Scr) was ≥ 451 $\mu\text{mol/L}$.

Transthoracic echocardiography (TTE), (Sonos 5500[®] Echocardiography System, Philips, Berkeley, California, USA), was used (transesophageal echocardiography [TEE] was not used). The frequency of the probe was 3.75 MHz. While on their left side, the echocardiography transducer (or probe) was placed on the chest wall (or beside the thorax) of the subjects. Determination of image planes was made on the long axis of the left ventricle, short axis of the mitral valve, and capillary muscle, and the apical axis of 4-chamber, 5-chamber, and 2-chamber. Diastolic functions of the ventricular septum and free wall were tested using multiplanar and multiangle imaging. M-mode and echo methods were used to determine the ventricular wall thickness and to observe morphology and structures of different ventricles. Transthoracic echocardiography is widely used with its rapid checking time and cheap cost, but one of its major limitations is the frequent appearance of artifacts that mimic a dissection flap. It is therefore, essential that the echocardiographer makes certain to distinguish true dissection flaps from such artifacts. Another examination was performed on a GE signal 1.5T MRI scanner (General Electric [GE], Fairfield, Connecticut USA) using cardiac and electrocardiographic gating. Images were taken by scanning through the ascending and descending aorta at both coronal and sagittal oblique planes. The MRI has been shown to be very accurate in the diagnosis of thoracic aortic disease without either radiation exposure or iodinated contrast, with sensitivities and specificities that are equivalent to or may exceed those of CT and TEE. However, it is mainly because of its prolonged duration of imaging acquisition and lack of widespread availability on an emergency basis or in primary hospital that limits the MRI's wide application. High-speed single spiral CT (GE, Fairfield, Connecticut USA) was also used. Scanning parameters were 150 mA, 120 Kv, and the coil was 1-1.5. Depth and distance of the scanning was 5-10 mm. Depth of image reconstruction was 1-2 mm and 0.5-1 mm. Ioversol (100 mL) was used as a non-ionic contrast agent for CT scanning. A small dose of contrast material was injected through an intravenous (IV) line into a small vein in the arm of the patient (2.0-2.5 mL/s). Scanning was performed 20-30 seconds after injection. A 3-dimensional reconstruction

was created using the scanned images. The CT scanning can be considered at first for its ability to image the entire aorta including lumen and wall, its short time required to complete the imaging process, and its vivid 3-dimensional image.

All patients presented with typical symptoms of transverse spinal cord damage with sudden paraplegia, loss of sensations, and bladder or bowel difficulties. Based on the DeBakey type classification, DeBakey type I was identified in 3 patients and DeBakey type III was identified in 6 patients. Six of the 9 patients had anterior spinal artery syndrome. Three patients had symptoms of anterior and posterior spinal artery damage. The symptoms included sudden onset of pain in the chest, abdomen, and back. However, only one patient had painless AD. Also, one patient had shock syndrome. Moreover, most of the patients had overexcitement, an increased workload, over tiredness, and emotional excitation before admission to the hospital. Of note, most patients in the study had other complications, which were important in distinguishing AD from other diseases: blood in urine (11.1%), acute renal failure (66.7%), a high level of serum enzymes (55.6%), anemia (33.3%), progressive reduction in platelet count (11.1%), and reduction in pulse dorsal pedis artery (22.2%). Physical examination revealed that blood pressure was elevated in all patients and peaked at 260/150 mm Hg, mean pressure was 182/103 mm Hg, and systolic pressure was increased by 30-70 mm Hg compared with normal. Muscle power of the bilateral lower extremities was grade 0 in 5 cases, and grade I-II in 4 cases. Simultaneously, 6 of the 9 patients were complicated by acute renal failure, 5 patients suffered from acute skeletal muscle ischemic necrosis, 3 patients had acute pancreatic necrosis, one patient had pleural

effusion, and one patient had cardiac tamponade. To confirm the diagnosis of AD, TTE, computed tomography angiography (CTA) and MRI tests were conducted to detect the site, size, and number of the aortic tear, true and false lumen, intimal flap, cardiac tamponade, aortic regurgitation, and affected branches of the coronary artery. We found that 3 patients had a widened aorta, the true and false lumens that were divided by an intimal flap, and an enlarged left ventricle with diastolic dysfunction revealed by TTE. In addition, CTA test illustrated that the aorta was divided into true and false lumens by an intimal flap and simultaneously detected the 2 lumens in 3 patients, blood flow in the true lumen was faster than in the false lumen, therefore, the intensity of the signal in the true lumen was higher than that in the false lumen. Moreover, CTA (volume rendering [VR]) showed true and false lumen and intimal flap from the aorta to kidney artery in another patient (Figure 1). The MRI is also an important tool for the rapid diagnosis of aortic aneurysm. The MRI tests in 2 cases revealed formation of the true and false lumens in the aorta, and deformation of the true lumen and thrombus in the false lumen (Figure 1). Although 2 of the 9 patients had obvious paraplegia, spinal MRI tests were normal. It is possible that the MRI test was performed too quickly (within 3 hours of onset of AD) to detect abnormalities. In the 9 cases, the average length of aorta covered was 18.7 cm (range from 8-25.3 cm). The maximum aortic diameter averaged 49 mm. The true lumen diameter averaged 22.6 mm. The false lumen diameter averaged 20.2 mm. Among them, 3 patients were examined by TTE, 2 patients were examined by CTA, another 2 patients were examined by MRI, one patient was examined by CTA (VR) combined with MRI, one patient was diagnosed by autopsy after

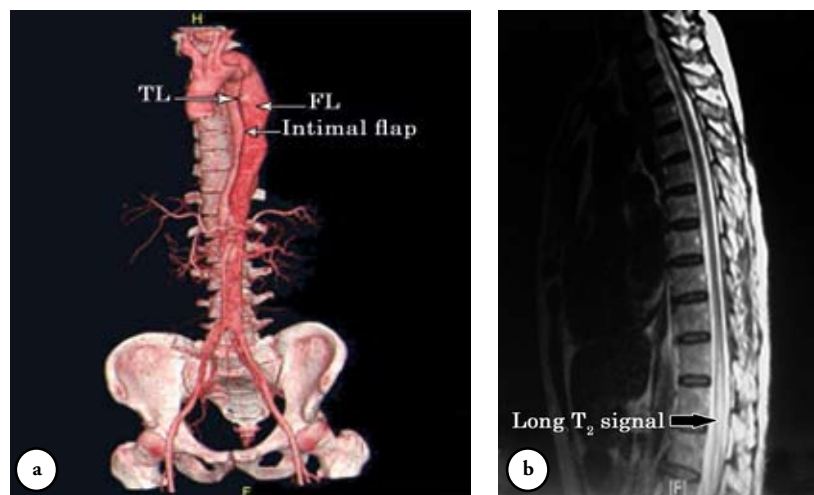


Figure 1 - Computer tomography angiography volume rendering showed a) true lumen (TL) and false lumen (FL) and intimal flap from aorta to kidney artery, and b) the MRI image revealed long T₂ signal at the thoracic level.

death. Typical imaging features of TTE, CTA, and MRI are true and false lumen and intimal flap from the aorta. Selection of the most appropriate imaging study may depend on patient related factors (namely, hemodynamic stability, renal function, contrast allergy) and institutional capabilities (namely, rapid availability of individual imaging modalities, state of the technology, and imaging specialist expertise). For example, for patients who require repeated imaging to follow an aortic abnormality, the MRI, with its minimal radiation exposure, may be preferred to CT. In terms of treatments and prognosis in these AD patients, 7 patients died on the day of admission despite active medical treatment, one survived after aorta aneurysm surgery and artificial vascular grafting, and the conditions of another patient stabilized after anti-hypertension treatment.

Acute AD is the most common and severe aortic disease. Clinical manifestations generally include chest or back pain. Nevertheless, the correct antemortem diagnosis was made in less than half of the cases despite major advances in diagnostic modalities. The acute onset of severe chest, abdomen, or back pain is one of the most common initial presentations. The pain often occurs suddenly and is described as a tearing, ripping, or with a knife-like nature, and pain medication cannot fully relieve the pain. In this report, 8 patients complained of pain in the chest, abdomen, or back, and only one patient had no complaint of pain. However, in some patients, although AD was painless, chest x-ray tests were abnormal.³ Less common presentations of AD include congestive heart failure, syncope, cerebrovascular accident, shock, paraplegia, and lower extremity ischemia. Aortic dissection presenting as acute spinal cord damage is the most severe complication and is very rare. In that case, apart from the typical clinical manifestations of AD, the symptoms of acute spinal cord damage are evident. Aortic dissection is characterized by acute onset, rapid progression, and complicated clinical manifestations. The mortality rate is high if the diagnosis is delayed. Nevertheless, our reports are consistent with previous reports showing that severe neurological complications resulting from spinal cord ischemia, such as paraplegia, loss of part or all forms of sensations,⁴ and difficulty in micturition, can occur in patients with AD.

It has been demonstrated that the spinal cord is supplied by 3 arteries: namely, the anterior spinal artery, the posterior spinal artery derived from the vertebral arteries, and the spinal branches derived from intercostal and lumbar arteries. Anatomically, the clinical symptoms of spinal cord ischemia should occur after mesenteric ischemia. Moreover, the ischemic tolerance in the spinal cord is not as good as in the mesenteric artery. Therefore, the symptoms of spinal ischemia are

much more severe than mesenteric artery ischemia, and the clinical manifestations of AD are characterized by abdominal pain as an initial complaint, but acute paraplegia as the main manifestation. In addition, the anterior spinal artery is the main supplier of the radicular arteries, especially at the T10-12 regions, where blood flow is usually lower than other regions under normal conditions, making the T10-12 spinal cord more reliant on the supply of the anterior spinal artery, and more sensitive to insufficient blood flow. When AD occurs, obstruction of the aortic branches prevents blood supply from the spinal cord and other organs, leading to insufficient tissue perfusion and subsequent ischemia. In this report, we found that all cases had acute renal failure and/or acute pancreatic necrosis and/or acute skeletal muscle ischemic necrosis, suggesting the development of anterior spinal artery syndrome. It has been reported that the incident rate is 18-19%, and the duration of paraplegia usually varies from hours to 2 days.

In addition, it is noted that AD primarily presenting as acute spinal cord damage is difficult to distinguish from other spinal cord diseases, such as acute transverse myelitis (TM) and vascular diseases of the spinal cord. Emergency room physicians should not rule out the possibility of AD when making the differential diagnosis of acute myelitis, vascular myelopathy, or spinal cord compression. Transverse myelitis is a common spinal cord disease, and it often occurs in young adults. The onset of TM is rapid. The patients have a history of previous infection or immunization and experience chest and back pain or thoracic radicular pain, followed by lower limb weakness or paraplegia below the thoracic level. Clinically, damage occurs most often at the thoracic spinal cord; it develops over hours and peaks at one or 2 days. Compared with AD, the progress of TM is relatively slow and there is no sign of any other organs being affected. Another spinal cord disease is hemorrhage resulting from the vascular disease of the spinal cord. It occurs more frequently in males than in females. Patients are typically middle aged or elderly adults. Clinical features are sudden onset, affected bilateral lower limb, and severe chest, or abdominal pain. It is difficult to distinguish from the spinal ischemia induced by AD. However, the former usually does not affect multiple organs, while AD is usually complicated with acute renal failure, reduction in dorsal pedis artery pulse palpation, shock syndrome, platelet reduction, and an elevated level of creatine kinase. Moreover, a higher T1 signal revealed by spinal MRI tests makes the diagnosis easy. On the other hand, the typical characteristic of the patients with spinal vascular malformations is acute onset, the limbs are affected, and intermittent claudication is the main manifestation.

Moreover, MRI reveals vascular steal phenomenon, in which oxygenated arterial blood shunted through the vascular malformation causes the surrounding normal parenchyma to become hypoperfused. Furthermore, spinal angiography can facilitate the correct diagnosis.

After diagnosis of acute AD, the patients are required to be continuously monitored in the intensive care unit, and effective treatments should be taken to ensure that vital signs are stable, including blood pressure, heart rate and rhythm, central venous pressure, urine volume, and measurement of pulmonary capillary pressure and heart blood volume are measured when required. The management for AD includes pain control and controlling the intimal tear and reducing blood pressure. The systolic pressure should be under 100-120 mm Hg, with the average pressure of 60-70 mm Hg. Once the clinical conditions of patients are stable, TTE, CTA, and MRI tests will be performed to further confirm the diagnosis of AD, and surgery will be decided.

If patients with aortic aneurysm are defined as DeBakey type I, or II, especially complicated with incomplete closure of the aorta, surgical repair is usually considered. For the treatment of type III AD patients, a vascular grafting of the descending aorta is required. For the AD patients with multiple organs affected, re-establishment of blood circulation in the intercostal artery, the renal artery, or the superior mesenteric artery should be considered.⁵ In this report, one of the patients had aortic aneurysm surgery and artificial vascular grafting. His muscle power of bilateral lower limbs recovered from grade 2 to grade 4 after the surgery. Another patient and his relatives refused the surgery. His condition stabilized after treatment with sodium nitroprusside combined with a β -receptor blocker, and his muscle power of bilateral lower limbs recovered from grade 1 to grade 2. Of note, the diagnosis of AD in the 2 patients was made relatively earlier (<3 hours) than the rest of patients (>6 hours). Six of the 9 patients died due to delayed diagnosis and the loss of the chance of surgical intervention. One of the 9 patients was considered as a severe clinical condition (namely, the patient had progressively increasing pain in the chest, increased perspiration, was not lucid, and had a decreasing blood pressure), and his family members decided to cease treatment and he died shortly after

discharge from hospital. The findings suggest that early diagnosis and prompt treatment play an important role in reducing mortality. The cause of AD death is mainly due to rupture of the aortic aneurysm, leading to shock syndrome and cardiac tamponade. One autopsy has confirmed the clinical symptoms of AD in this report.

The study summarized clinical manifestations and imaging features of AD primarily presenting as acute spinal cord damage. Taken together, clinical features of AD patients presenting as acute spinal injury can be summarized as follows: the patients suffer from complete transverse spinal cord damage without infection history, the onset of paraplegia occurs within 1-2 hours, accompanied with hypertension, varying degrees of chest or abdominal pain, shock syndrome with high blood pressure, unexplained blood in the urine, acute renal failure, a high level of serum enzymes, anemia, progressive reduction in platelet count, and reduction in pulse dorsal pedis artery. In terms of imaging features of AD, tests of TTE, CTA, and MRI, which could detect the site, size, and number of the aortic tear, true and false lumen, intimal flap, were the 3 main methods in confirming the diagnosis of AD.

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