

## Case Report

# Painless Dissecting Aneurysm of the Aorta Presenting as Simultaneous Cerebral and Spinal Cord Infarctions

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Authors report a case of a painless acute dissecting aneurysm of the descending aorta in a patient who presented with unexplained hypotension followed by simultaneous paraplegia and right arm monoparesis. To our knowledge, case like this has not been reported previously. Magnetic resonance imaging of the brain and spine revealed hemodynamic cerebral infarction and extensive cord ischemia, respectively. Computerized tomography angiography confirmed a dissecting aneurysm of the descending aorta. The cause of the brain infarction may not have been embolic, but hemodynamic one. Dissection-induced hypotension may have elicited cerebral perfusion insufficiency. The cause of cord ischemia may be embolic or hemodynamic. The dissected aorta was successfully replaced into an artificial patch graft. The arm monoparesis was improved, but the paraplegia was not improved. In rare cases of brain and/or spinal cord infarction caused by painless acute dissecting aneurysm of the aorta, accurate diagnosis is critical because careless thrombolytic therapy can result in life-threatening bleeding.

**Key Words :** Aorta · Dissecting aneurysm · Cerebral infarction · Spinal cord ischemia.

## INTRODUCTION

An acute dissecting aneurysm (DA) of the aorta is a rare vascular event with an incidence of 5-30 cases per million people per year<sup>12)</sup>. A DA is an uncommon, but potentially catastrophic pathologic change with an extremely high mortality and morbidity. The classic, well-known, typical symptom of DA is severe chest or abdominal pain radiating to the back. This abrupt, lancinating pain can easily make physicians alert and cautious. However, it is important that not all DAs elicit typical pain and the correct diagnosis followed by urgent management cannot always be made. In one report, a DA was missed in 38% of patients on presentation, with 28% of cases diagnosed during autopsy<sup>16)</sup>. The frequency of pain-free DAs ranges between 5% and 15%<sup>7,9,13,16)</sup>. Such patients with DAs demonstrate signs and symptoms dependent on the involved arterial branches from the ascending to the descending aorta.

The neurologic symptoms associated with DA are often dramatic and may completely dominate the clinical features. The frequency of neurologic involvement, including transient deficits, varies between 17% and 40%<sup>1,7,9,13)</sup>. Cerebral ischemia and

paraplegia have been reported in 6-16%<sup>6,9-11,17)</sup> and 2-8%<sup>2)</sup> of patients with DAs, respectively. If these neurologic manifestations occur without pain, although rare, the possibility of DA can be missed without a high index of suspicion<sup>1,7,13)</sup>. We managed a patient with a painless DA in which cerebral hemodynamic infarction and thoracic spinal cord ischemia occurred concurrently. Herein, we have focused on the important diagnostic features and therapeutic considerations of DA with a review of the literature.

## CASE REPORT

A 51-year-old man was referred for further evaluation and treatment of paraplegia and right arm monoparesis. At the referral clinic, he was hypotensive (80/50 mmHg) and diagnosed as septic shock. In the emergency room, the blood pressure was 110/80 mmHg and the pulse was 88 beats/min. The duration of hypotension was less than 24 hours. On physical examination, there were no abnormal findings and he denied any pain in the chest, neck, back, or abdomen. On neurologic examination, the patient was somnolent, but could communicate with the examiners. There was right upper extremity weakness (motor grade II/V) and lower extremity weakness bilaterally (motor grade I/V). Below the T6 dermatome, anesthesia to pain and hypesthesia to light touch was noted. Below the T11 dermatome, total anesthesia were demonstrated to all types of stimuli. Perianal sensation, and voluntary and involuntary anal sphincter tone could not be elicited. On a plain chest posteroanterior (PA) ra-

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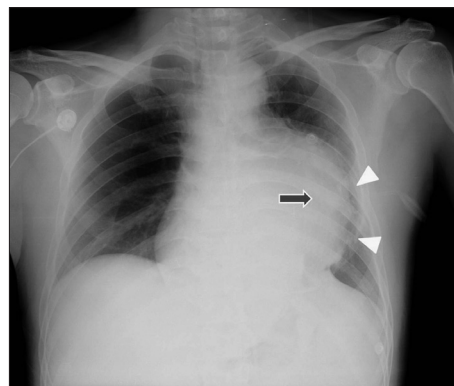
diograph, there was a relatively well-defined homogeneous radiopaque mass obliterating the left cardiac border and left lateral wall of the descending aorta, suggesting a post-mediastinal mass. The small curvilinear calcifications at the lateral border of the mass were highly suspicious findings of thoracic aorta pathology (Fig. 1). Chest computed tomography (CT) and CT angiography revealed extensive aneurysmal dilatation of the descending thoracic aorta with an intramural hematoma from the level of the carina to the level of the aortic hiatus, which was consistent with a Stanford type B (involving only the aorta distal to the origin of the left subclavian artery) DA (Fig. 2). The brain CT showed no abnormalities; however, the brain diffusion magnetic resonance imaging (MRI) showed multifocal hemodynamic infarctions in the supra- and infra-tentorial areas (Fig. 3). The brain magnetic resonance angiography was normal (Fig. 4). Routine cardiac evaluations, including echocardiography and Holter monitoring, were also negative. A thoracic MRI revealed a diffusely swollen spinal cord with patch high-signal intensity from T2 to the lower lumbar area. The lesion proved to be cord ischemia secondary to a DA (Fig. 5). To avert catastrophic rupture of a DA, we did not prescribe anti-coagulants or anti-platelet medications. To improve rheologic perfusion, conservative hydration therapy was done for 7 days. The mental status became alert and the right upper extremity weakness improved (grade IV/V); however, the paraplegia did not improve. The cardiovascular surgeons performed a resection of the DA of the descending aorta, followed by replacement with a 22-mm Vasctek graft (Terumo Cardiovascular System Corporation, Ann Arbor, MI, USA). The patient had no notable events or newly developed complications, but the pre-existing neurologic deficits (paraplegia, and neurogenic bladder and bowel) did not improve. The post-operative chest PA radiograph and CT angiography showed a normalized cardiac silhouette and no DA involving the thoracic aorta, respectively (Fig. 6). The patient was transferred to the Department of Rehabilitation Medicine for specialized treatment.

## DISCUSSION

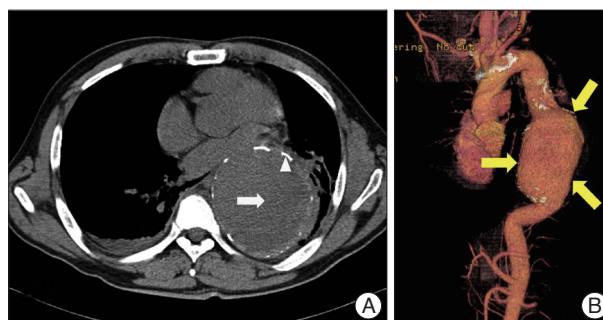
The purpose of this case report was to stress the importance of thorough surveillance of the patient's general condition in addition to neurologic abnormalities. The stroke physicians are prone to focus on ischemic lesions, duration after symptom onset, or decision of therapeutic modalities. In rare conditions, as in this report, routine thrombolytic procedures can be extremely harmful to patients. If this patient manifested a cerebral or spinal ischemic lesion without any complaints of severe pain, a DA would not be likely. Despite the painless attack, simultaneous cerebral and spinal cord

ischemic lesions led to a thorough re-evaluation of the patient, and a diagnosis of life-threatening DA.

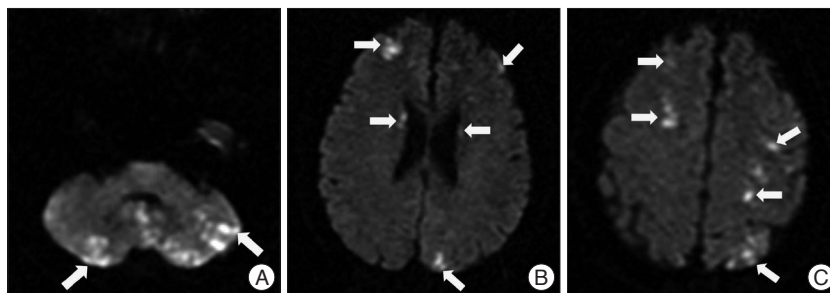
Although pain is one of the most common presenting symptoms of DA, 5-15% of patients deny painful attack episodes<sup>15</sup>. Little is known about the reasons that patients with DA do not experience pain, although three possible hypotheses have been proposed. First, a dissecting hematoma only causes the intima to bulge inward and re-enters the true aortic lumen. In this case,



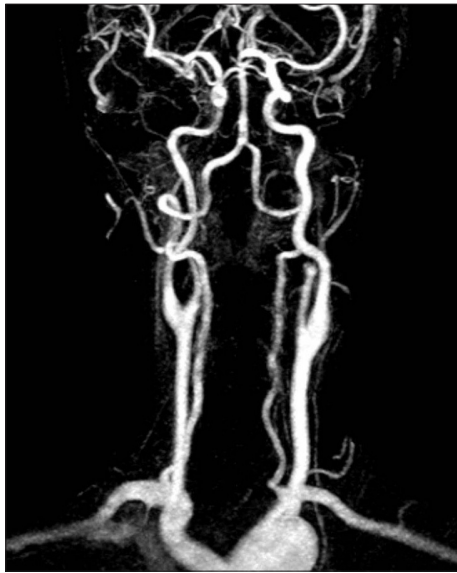
**Fig. 1.** The initial chest PA radiograph shows relatively well-defined homogeneous convex radiopaque mass, which obliterates the left cardiac border and left lateral wall of the descending aorta (arrow). The faint curvilinear calcifications along the lateral border of the mass suggest thoracic aorta pathology (arrowheads). PA : posteroanterior.



**Fig. 2. A :** The axial cut of the non-enhanced chest computed tomography (CT) shows extensive aneurysmal dilatation of the descending thoracic aorta with intramural hematoma (arrow) and surrounding calcified wall (arrowhead). **B :** The 3-dimensional (3D) surface volume rendering image of aorta CT angiography shows a large aortic aneurysm at the thoracic aorta (stanford type B aneurysm; arrows). Note that no aneurysm is visible at the ascending aorta.



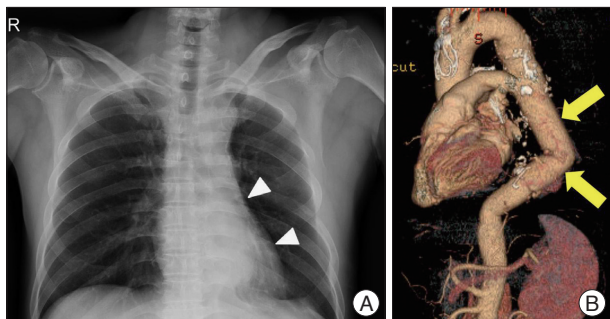
**Fig. 3.** The brain diffusion weighted magnetic resonance images (MRIs) show multifocal infarctions at both cerebellar (A) and cerebral hemispheres with parallel hemodynamic insufficiency patterns (B and C).



**Fig. 4.** The brain and neck magnetic resonance angiography (MRA) shows no definite abnormality.



**Fig. 5.** The T2-weighted sagittal image of thoracic MRI shows diffuse intramedullary high signal and cord swelling from thoracic to lower lumbar spinal level, suggesting cord ischemia (arrows). MRI : magnetic resonance imaging



**Fig. 6.** A : Postoperative chest PA shows normalized cardiac border. B : The 3-dimensional (3D) surface volume rendering image of aorta CT angiography shows no more aneurysm at thoracic aorta. PA : posteroanterior.

the adventitia, which is the source of pain, is not violated or displaced<sup>14,16</sup>. Second, the involvement of cerebral vessels may dull the patient's perception of pain<sup>8,15</sup>. Third, loss of visceral and spinothalamic perception of pain caused by the preceding severe spinal ischemia may block the normal neuro-pathway of pain perception<sup>3</sup>. Nevertheless, an established theory has not been advanced.

In 17-40% of patients with DAs, neurologic symptoms can be manifested<sup>6,10,11,17</sup>. The causes of ischemic neurologic deficits are occlusions of main feeding arteries, such as the carotid, vertebral, and spinal arteries, or vasa nervorum of the peripheral nerves, or hypotension-induced perfusion failures<sup>12</sup>, which can induce ischemic stroke, spinal cord ischemia, ischemic neuropathy, hypoxic encephalopathy, or syncope. The two possible causes of arterial occlusion are extension of dissection into major branches, such as the brachiocephalic trunk or common carotid arteries, and thromboembolism from the aorta to the major arteries<sup>5</sup>. Ischemic stroke has been reported to occur in 5-10% of all patients.

In 2-8% of patients with DA, one of the other etiologies of neurologic deficits is spinal cord ischemia<sup>2</sup>. Cheshire et al.<sup>3</sup> reported that 2 of 44 patients (4.5%) with spinal cord infarctions had DAs as the cause of spinal ischemia. Spinal cord involvement in patients with DA can be secondary to obstruction of the intercostal and lumbar arteries, the artery of Adamkiewicz (arteria radicularis magna), or the thoracic radicular arteries<sup>12</sup>. In this case, based on signal changes of MRI, the cause of spinal cord infarction was obstruction of the medial medullary branch of the anterior spinal artery originated from the artery of Adamkiewicz. As in the current case, considering a MRI signal change of the gray matter of the spinal cord, it can be assumed that a DA is the cause of obstruction of the secondary medial medullary branch of the anterior spinal artery due to obstruction of the artery of Adamkiewicz.

Anatomically, an ischemic cerebral stroke can be elicited much easier in cases of Stanford type A (involving the ascending aorta) dissection by means of extension of dissection or direct thromboembolism, and most of the reported cases of ischemic stroke have Stanford type A. Conversely, an ischemic spinal lesion is much more common in Stanford type B (involving the aorta distal to the origin of left subclavian artery) dissection<sup>4</sup>.

With a hemodynamic point of view, in Stanford type B DA retrograde thromboembolism to intracranial vessels is unusual. Terasaki et al.<sup>18</sup> reported two cases of brain infarction associated with dissection of the thoracic aorta. Unlike our case presentation, the infarction territories of these patients were restricted to middle cerebral arteries territories and the authors speculated that the pseudolumen of the dissected thoracic aorta was the causative factor of occlusion of the right internal carotid artery. However, in our case, several issues should be pointed out for evaluation of the cause of cerebral infarction. First, our patient had severe hypotension at the time of presentation, which made the primary physician suggest septic shock. Second, the infarc-

tion areas were not restricted to one arterial territory, but scattered to multiple vessels territories, with far distal points. Thus, the cause of multiple ischemic cerebral stroke was not embolic or pseudolumen-induced, but hemodynamic compromise. The DA should be considered as one possible cause of cerebral or spinal cord ischemia. With review the reported literatures till now, this seems to be the first case of a painless Stanford type B DA manifested by concurrent hemodynamic compromise-induced cerebral infarction and emboli or dissected intimal flap-induced spinal cord ischemia.

## CONCLUSION

The brain and/or spinal cord infarction caused by painless acute dissecting aneurysm of the aorta is rare. But, early accurate diagnosis is essential to prevent thrombolytic therapy induced life-threatening bleeding.

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