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Case Report

Paraplegia caused by painless acute aortic dissection

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Objectives: Painless acute aortic dissection in which paraplegia is the only presenting sign is rare, with limited reported cases.
Case report: The authors report a patient with painless acute aortic dissection who presented with sudden onset paraplegia. Ischemic diseases of the spinal cord were suspected as the cause. MRI revealed extensive acute aortic dissection with an intramural hematoma. The patient was treated conservatively by strictly controlling his blood pressure. The treatment was successful, although the motor function of the lower extremities could not be rescued. Although 3% to 5% of patients with acute aortic dissection present with paraplegia as a result of spinal cord infarction, most of these patients experience severe pain prior to presentation.
Conclusion: Painless acute aortic dissection in which paraplegia is the only presenting sign is very rare. However, aortic diseases, including acute aortic dissection, should always be considered as a differential diagnosis of patients with sudden onset, painless paraplegia.
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Keywords: acute aortic dissection; painless; paraplegia; spinal ischemia

Introduction

The presenting signs of acute aortic dissection (AAD) are protean, and vary considerably depending on the portion and the extent of the aorta involved as well as the organs affected by ischemia. However, patients with AAD have a common presenting symptom of severe pain. In fact, the sudden onset of severe chest and/or back pain is a hallmark of AAD, and more than 90% of patients with AAD complain of severe pain on presentation in the emergency room (ER).[1] Nevertheless, there is a subgroup of patients who have little pain or even none at all.[1,2] The authors report a rare case of AAD in which the patient presented with painless, rapidly progressive paraplegia. Although paraplegia is a well-known manifestation of AAD, reportedly experienced by 2% to 8% of all AAD patients,[3] most of these patients experience severe pain prior to the development of paraplegia.[4] In the case reported here, AAD was detected by MRI, which is considered a vital tool for making a definitive diagnosis in patients presenting with sudden onset, painless paraplegia.

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Case report

A 50-year-old man with a prior medical history of a small lacunar stroke and arterial hypertension suddenly noticed numbness in his lower extremities upon standing up. The patient was not diabetic. His medication included aspirin and an anti-hypertensive agent. Weakness of the lower extremities developed within 1 h, and the patient could no longer stand in an upright position. When he arrived in the ER by ambulance 2 h after the onset of numbness, he was alert but paraplegic. His blood pressure in the right arm was 111/68 mmHg, his pulse was 54 beats/min, and his respiration rate was 18/min. The patient did not complain of any pain at any time. An ECG showed sinus bradycardia, but no ischemic changes were observed. Manual muscle testing revealed the strength of the muscle groups in the lower extremities to be 1/5. Sensory tests showed a decrease in pain and temperature sensation below the L1 level. Position and vibration senses at the same level were only slightly impaired. The deep tendon reflexes were intact in the upper extremities, but were diminished in the lower extremities. Pulsation of the dorsalis pedis artery was equal to palpation bilaterally. An ischemic lesion of the spinal cord in the form of the anterior spinal artery syndrome was suspected, and the patient underwent an emergency MRI of the thoraco-lumbar spine. Unexpectedly, the MRI revealed a dissection of

the aorta extending from the ascending aorta proximally to the orifice of the renal artery distally. The patient was diagnosed with AAD of Stanford type A, which has been defined as dissections involving the ascending aorta (regardless of the site of the primary intimal tear and extent of distal propagation of the dissecting hematoma)[1] (Figure 1). The false lumen of the dissected aorta from the ascending to the descending thoracic aorta was obliterated by a thrombus, indicating the presence of AAD with an intramural hematoma (IMH).[5] Occlusion of the artery of Adamkiewicz was suspected to have caused the clinical manifestations. The patient was referred to cardiovascular surgeons. Since the patient had AAD with IMH, he was treated conservatively by strictly controlling his blood pressure. The patient was referred to the Department of Rehabilitation Medicine 70 days after admission, with paraplegia and dulled sensation to pain below the L1 level.

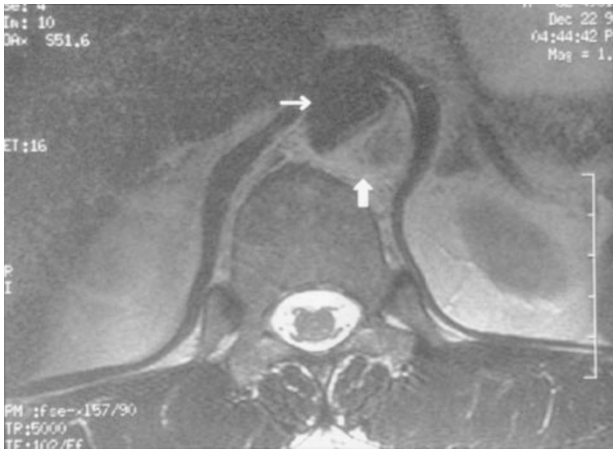

Figure 1 MRI of the lumbar spine, at the L1 level. T2 weighted-image revealed the presence of aortic dissection. Note the presence of an intramural thrombus in the false lumen (thin arrow) together with the patent true lumen (large arrow)

Discussion

Patients with AAD almost invariably present with sudden onset of severe pain. Although the severity, duration, and distribution of the pain may vary from patient to patient, more than 90% of all AAD patients complain of pain when they present in the ER.[1] The severe pain often serves as an alarm to physicians, enabling them to make the correct diagnosis quickly and easily. However, a certain proportion of AAD patients experience little or no pain at the time of presentation[1,2] and pose a significant diagnostic challenge to physicians. Most patients with painless AAD present with an episode of syncope resulting from cardiac tamponade.[6–8] Less frequently, involvement of cerebral vessels causes neurological manifestations represented by a progressive disturbance of consciousness that may dull the patients' perception of pain.[9] However, although 'painful' paraplegia as a result of AAD is present in 2% to 8% of all AAD cases,[3] 'painless' paraplegia caused by AAD is very rare. Most AAD patients with paraplegia experience severe pain prior to presentation.[4] Only eight such cases have been reported in the literature (Table 1),[3,9–16] and five of the patients had type A dissections and the other three patients had type B. Unlike acute myocardial infarction, in which the incidence of painless infarct is greater in elderly and diabetic patients,[17] few common clinical characteristics can be derived from the constellation of patients with painless AAD.

The reason some patients with AAD do not experience any pain remains unknown. Thus far, three possible explanations have been proposed. 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.[18] Second, the involvement of cerebral vessels may dull the patients' perception of pain.[9] Third, loss

Table 1 Clinical characteristics of the nine patients with AAD who presented with paraplegia that have ever reported in the English literature

No.	*References*	*Patients' age, sex*	*Underlying disease*	*Initial diagnosis*	*Type of dissection*	*Treatment*	*Outcome (GOS)*
1	Waltimo *et al*, 1980[11]	52, M	HBP	Medullary infarction	Type A	Medical	Death
2	Gerber *et al*, 1986[9]	78, M	n.d.	Embolism of both legs	Type A	Medical	Severe disability
3	Rosen, 1988[10]	67, F	none	AAD	Type A	Refused surgery	Death
4	Zull and Cydulka, 1988[3]	67, M	HBP	Deteriorated in ER	Type A	CPR in ER	Death
5	Holloway *et al*, 1993[15]	92, F	HBP	Undetermined	Type A	Medical	Death
6,7	Spittel *et al*, 1993[13]	n.d. two cases	n.d.	n.d.	Type B (two cases)	Medical	Moderate disability
8	Elefteriades *et al*, 1999[14]	n.d.	n.d.	n.d.	Type B	Medical	n.d.
9	Present case	50, M	Lacunar stroke, HBP	Ant. spinal cord synd	Type A	Medical	Moderate disability

AAD: acute aortic dissection; Ant: anterior; CPR: cardiopulmonary rescucitation; ER: emergency room; GOS: Glasgow Outcome Scale; HBP: high blood pressure; n.d.: not described; synd: syndrome

of visceral and spinothalamic perception of pain caused by the preceding severe spinal ischemia may dull the patients' perception of pain.[19] None of these hypotheses has been proved conclusive. Most patients with painless AAD have been found to harbor localized dissections that only involve the proximal aorta, and thus, they often present with syncope.[6–8,20] This finding reciprocally supports previous reports that the clinical presentation of AAD in the form of painless paraplegia is very rare,[3,9–16] since the dissection in these cases always involves the descending thoraco-abdominal aorta, where the anterior spinal artery of Adamkiewicz originates.

Among several non-traumatic spinal disorders in which acute paraplegia without pain is the presenting symptom, spinal ischemia is a rare but essential category in the differential diagnosis.[19] In our patient, the rapid progression of the paraplegia was completed in 1 h, precluding other demyelinating, tumorous, or degenerative diseases of the spinal cord. The presence of a sensory disturbance also made Guillain-Barré syndrome unlikely.

Among the various etiologies of spinal ischemia, aortic diseases including AAD are not extremely rare.[19,21] Cheshire *et al*[19] reported that out of 44 patients with spinal cord infarction, two (4.5%) had AAD as the cause of their spinal ischemia. In our case, spinal MRI revealed the presence of AAD and enabled differential diagnosis of the suspected spinal cord infarction. Although T2-weighted images occasionally fail to demonstrate the presence of acute ischemic changes in the spinal cord, they can delineate related pathoanatomy around the spinal cord and contribute to an accurate diagnosis. Interestingly, the correct diagnosis was not made while the patient was in the ER in four of the five previous case reports listed in Table 1. These patients had a very poor outcome and for that reason, MRI may be essential for the diagnosis of patients who present with sudden onset, painless paraplegia.[22]

In summary, a rare case of painless AAD in which the patient presented with sudden onset of paraplegia is reported. AAD should be considered in the differential diagnosis of patients who present with paraplegia of acute onset, regardless of whether they complain of pain. Spinal MRI is the examination of choice for such patients, as was demonstrated by this report.

References

1 DeSanctis RW, Doroghazi RM, Austin WG, Buckley MJ. Aortic dissection. *New Eng J Med* 1987; **317:** 1060–1066.
2 Greenwood WR, Robinson MD. Painless dissection of the thoracic aorta. *Am J Emerg Med* 1986; **4:** 330–333.
3 Zull DN, Cydulka R. Acute paraplegia: a presenting manifestation of aortic dissection. *Am J Med* 1988; **84:** 765–770.
4 Tanaka T *et al*. Transient paraplegia caused by acute aortic dissection. Case report. *Neurol Med Chir (Tokyo)* 1990; **30:** 54–58.
5 Morioka Y *et al*. Intramural hematoma of the thoracic aorta. *Eur J Cardiothorac Surg* 1998; **13:** 230–239.
6 Cooke JP, Safford RE. Progress in the diagnosis and management of aortic dissection. *Mayo Clin Proc* 1986; **61:** 147–153.
7 Kuhlmann TP, Powers RP. Painless aortic dissection: an unusual cause of syncope. *Ann Emerg Med* 1984; **13:** 549–551.
8 Rahmatullah SI *et al*. Painless limited dissection of the ascending aorta presenting with aortic valve regurgitation. *Am J Emerg Med* 1999; **17:** 700–701.
9 Gerber O, Heyer EJ, Vieux U. Painless dissection of the aorta presenting as acute neurologic syndromes. *Stroke* 1986; **17:** 644–647.
10 Rosen SA. Painless aortic dissection presenting as spinal cord ischemia. *Ann Emerg Med* 1988; **17:** 840–842.
11 Waltimo O, Karli P. Aortic dissection and paraparesis. *Eur Neurol* 1980; **19:** 254–257.
12 Beach C, Manthey D. Painless acute aortic dissection presenting as left lower extremity numbness. *Am J Emerg Med* 1998; **16:** 49–51.
13 Spittell PC *et al*. Clinical features and differential diagnosis of aortic dissection: experience with 236 cases. *Mayo Clin Proc* 1993; **68:** 642–651.
14 Elefteriades JA *et al*. Management of descending aortic dissection. *Ann Thorac Surg* 1999; **67:** 2002–2005.
15 Holloway SF *et al*. Painless aortic dissection presenting as a progressive myelopathy. *J Neurol Sci* 1993; **120:** 141–144.
16 Cambria RP *et al*. Vascular complications associated with spontaneous aortic disection. *J Vasc Surg* 1988; **7:** 199–209.
17 Antman EM, Brunwald E. Acute myocardial infarction. In: Fauci A *et al*. (ed). *Harrison's Principles of Internal Medicine*. 14th edn. McGraw-Hill: New York, 1998, pp 1352–1365.
18 O'Donovan TPB, Osmundson PJ, Payne WS. Painless dissecting aneurysm of the aorta. *Circulation* 1964; **24:** 782–786.
19 Cheshire WP, Santos CC, Massey EW, Howard Jr JF. Spinal cord infarction: etiology and outcome. *Neurology* 1996; **47:** 321–330.
20 Borst HG, Heinemann MK, Stone CD. Clinical presentation of aortic dissection. In: Borst HG, Heinemann MK, Stone CD (ed). *Surgical management of aortic dissection*. Churchill Livingstone: Edinburgh, 1996, pp 55–67.
21 Howell JM, Mcfarling DA, Chisholm CD. Ischemic injury to the spinal cord as a cause of transient paraplegia. *Am J Emerg Med* 1987; **5:** 217–219.
22 Friese KK, Steffens JC, Caputo GR, Higgins CB. Evaluation of painless aortic dissection with MR imaging. *Am Heart J* 1991; **122:** 1169–1173.