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Progressive Ischemic Myelopathy due to Painless Aortic Dissection: A Case Report

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The clinical appearance of aortic dissection is characterized by the sudden, acute chest or abdominal pain, accompanied by coldness in the limbs and sweating-like symptoms of shock. Absent pulse or asymmetric pulse was observed in some patients. If the feeding arteries of the specific organs are located in the false lumen, this may lead to sudden loss of blood supply to the tissue e.g. spinal cord, resulting in cord damage. In the previous reports, approximately 20% of the individuals with aortic dissection might have neurological deficit resulting from insufficiency of blood supply. Most of the patients had suffered from severe chest or abdominal pain with sudden onset. Here we report a case with progressive neurological impairment because of long-term insufficient blood supply to the spinal cord. It is probably due to a painless aortic dissection. The patient was a 66-year-old male who presented with paraparesis. On examination, strength was 5/5 in both upper extremities and 3-4/5 in both lower extremities. Pricking sensation was relatively retarded below the sixth thoracic vertebra on both sides. Fine-touch sensation was limited below the ninth thoracic vertebra on the right side and below the eleventh thoracic vertebra on the left side. Responses to vibration and joint position sense were well preserved. Deep tendon reflexes were increased in his legs. He had voluntary anal contraction and preserved anal and bulbocavernous reflexes. MRI revealed dissection of the descending aorta from T4 to T10 levels with thrombus present in the false lumen, and the thoracic cord was marked atrophy, especially from T6 to T8 levels. To the best of our knowledge, no such case had been reported so far. (J Rehab Med Assoc ROC 2002; 30(4): 251 - 258)

Key words: aortic dissection, myelopathy, spinal cord injury

INTRODUCTION

Aortic dissection is due to weakening of the middle muscular layer of the blood vessel, accompanied by lesions on the inner layer, through which blood passes and accumulates between the inner and middle layers. It occurs most frequently in patients of 50 to 70 years old with a history of high blood pressure, and more frequently among males than females. [1] Patients generally...
present with sudden excruciating precordial or abdominal pain, diaphoresis, and coldness of limbs, symptoms characteristic of shock.\cite{12} If at the time of onset, a vascular false lumen ruptures into the pericardium or retroperitoneum, life threatening conditions such as cardiac tamponade or massive blood loss may arise.\cite{12} In the process of expanding, if vascular false lumens extend to the degree that the orifices of the segmental arteries are occluded or blocked, many different clinical manifestations, such as stroke, renal failure, or spinal cord ischemia, may occur.\cite{12,4}

In previous reports, 8% to 33% of aortic dissection cases involve neurological damage and 4.2% present with spinal cord damage.\cite{11} Moreover, all the cases indicate intense chest or abdominal pain of sudden onset. Here, we present a case of aortic dissection manifested with painless, gradual onset and symptoms of progressive neurological impairment due to long-term insufficient blood supply to the spinal cord. To the best of our knowledge, no such case had been reported earlier.

**CASE HISTORY**

This 66-year-old male was a patient of hypertension and bronchial asthma under irregular control in our hospital. He had been unconscious for five days once in 1981 as a result of a head injury, but which seemed not to leave any sequela. He first experienced numbness and weakness of the legs progressively extending upward from both feet in 1987 after a prostate surgery. At that time, he underwent traditional Chinese medical treatment, but symptoms did not improve, and indeed spread to his upper body was reported. In 1991, he was diagnosed as having hydrocephalus and then received a ventricle-peritoneum shunt surgery, but his symptoms were progressively worsened. Herniated intervertebral disc was once suspected in other hospital but the correspondence treatment was in vain.

By 1993, he needed the support of a walker to ambulate, and was suffering from repeated urinary tract infections. He visited our hospital and was admitted for further examination and physical therapy.

Upon the patient's admission, physical examination revealed situs inversus, regular heart beat without murmur, and normal blood pressure. The neurological status showed good consciousness and intact cranial nerves. There was severe extensor spasticity with ankle clonus in the right leg, and only moderate in the left. The muscle tone of upper extremities was normal. The strength of the upper limbs was within normal limit, but that of the lower limbs had only 3-4/5 (Table 1) according to manual muscle tests. The patient's response to pricking was relatively retarded below the sixth thoracic vertebra on both sides. Fine-touch sensation was diminished below the ninth thoracic vertebra on the right side and below the eleventh thoracic vertebra on the left side. Responses to vibration and joint position sense were well preserved.

Deep tendon reflexes in the upper extremities were normal. The Baojun’s sign of the abdomen was negative. The knee and ankle jerks were three to four pulses. The Babinski’s sign of both legs was positive.

Regarding the function of the sphincter, the patient was able to contract his anus. The sensation and muscle tone of the anus were normal with preservation of the anal and bulbocavernosus reflexes.

Most of the laboratory data were normal, with the exceptions of ALT (alanine transaminase) at 49 IU/L (normal range: 2-32 IU/L), and AST (aspartate transaminase) at 46 IU/L (normal range: 10-30 IU/L). The VDRL (venereal disease research laboratory) slide test revealed negative. Cystometry revealed hyperreflexia of the detrusor muscle and low bladder capacity (290 cc), which was compatible with upper motor neurogenic pathology.

The image evaluation of the cardiovascular system was as follows. The cardiac echo revealed hyperensive cardiovascular disease. MRI revealed dissection of the descending aorta from T4 to T10 levels with thrombus present in the false lumen (Figure 1), and thoracic cord was marked atrophy, especially from T6 to T8 levels, and a spot of myelomalacia in the center of the sixth thoracic spinal cord.

**Table 1. Result of manual muscle test**

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Right</th>
<th>Left</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hip flexors (L2)</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Knee extensors (L3)</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Ankle dorsiflexors (L4)</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Big toe extensors (L5)</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Ankle plantar flexors (S1)</td>
<td>3</td>
<td>4</td>
</tr>
</tbody>
</table>
Figure 1.  (A)(B) Sagittal view of T1WI and T2WI revealed marked spinal cord atrophy at the thoracic level, especially from T6 to T8 levels (as arrows)  
(C)(D) Coronal view of T1WI and T2WI over T6 level revealed dissection of the descending aorta with thrombus present in the false lumen (as arrow a) and marked spinal cord atrophy with a spot of myelomalacia in the center of spinal cord present as hypointense on T1WI and hyperintense on T2WI (as arrow b).
According to the results from these examinations, Type 3 aortic dissection was imposed for the patient. We prescribed the anticoagulant (warfarin sodium) and the antiplatelet agent (dipyridamole) in an attempt to prevent the progression of thrombosis.

A comprehensive rehabilitation program, including the intermittent catheterization program to train the bladder, the strengthening program of the lower limbs, the ambulation training, and the education and training of the activities of daily living was instituted. The patient was trained to become ambulatory with bilateral axillary crutches.

**DISCUSSION**

Acute aortic dissection is a serious, life-threatening condition with many different clinical manifestations depending on the severity and distribution of the dissection. The acute onset of severe chest pain was the most common initial complaint (74%) of acute aortic dissection. Other manifestations included congestive heart failure, syncope, cerebrovascular accident, shock, paraplegia, and lower extremity ischemia. Risk factors include atherosclerosis, high blood pressure, Marfan's syndrome, pregnancy, bicuspid aortic valve, coarctation of aorta, prior aortic catheterization or trauma. Hypertension was the most common predisposing factor (78%) among these factors. DeBakey identifies three classifications. Type 1: originates at the ascending aorta and extends downward beyond the aortic arch. Type 2: originates at the ascending aorta and extends downward not beyond the brachiocephalic artery. Type 3: originates at the left subclavian artery and extends downward.

In addition, the Stanford system classifies dissections as type A (proximal) and B (distal). Stanford Type A is equivalent to DeBakey Types 1 and 2 and Stanford Type B is equivalent to DeBakey Type 3. Most of the Type A cases require surgical intervention, but many Type B cases are given conservative treatment.

The pain associated with aortic dissection is generally very acute, and the patient usually describes the feeling as one of "tearing" or "ripping." Pain occurs most frequently in the substernal, interscapular or mid-back areas, and may sometimes extend to the abdomen, lower back, waist, neck and the involved extremities. If the pain area extend above and below the diaphragm and is migratory, it is very likely to have been caused by aortic dissection. However, some of the aortic dissection was painless. In 1948, Baer and Goldburgh reported that 55% of 44 patients with aortic dissection did not have a history of pain. In 1958, according to the elaborate review by Hirst, 14% of 409 patients whose symptoms were recorded, did not mention pain. More recent series compiled by clinicians and radiologists including more acute cases showed an incidence of painless aortic dissection of only about 10%. Donovan et al. reported a case of painless aortic dissection that presented as new-onset paraplegia in 2000. Although this phenomenon is rare, it should be considered in the differential diagnosis of new-onset paraplegia, particularly in a patient with predisposing risk factors. Several explanations for the absence of pain in patients with aortic dissection were speculatively categorized as: 1. Aortic dissection diagnosis was established weeks to years after actual dissection, and pain may not have been remembered at the time of diagnosis. 2. There existed changes in consciousness, mentation, memory, or speech such that these patients could not describe a pain syndrome. 3. Pain was present but not in the classically recognized locations (such as hip or groin pain). 4. Diverse complaints other than pain (such as epigastric discomfort) were described. 5. Slow dissection may not result in pain. 6. Sparing of adventitia may obviate pain. 7. Circumventing and thus sparing intercostal, lumbar, and splanchic vessels may result in painless dissection.

Tracing back the patient's history, he also searched for management in our medical department due to chest discomfort; however, the time of chest discomfort was much later than the time of muscle weakness and voiding disorders. Moreover, the patient did not show any characteristic feature of the aortic dissection syndrome when he presented his paraparesis. We, therefore, classify it as painless aortic dissection, possibly because the infarction was not acute, or the decrease in the blood supply to the spinal cord was gradual.

In many clinical manifestations of aortic dissection, neurological impairment is about 20%, generally ranging from 8 to 33%. Neurological impairment can be categorized as: (a) Cerebrovascular accident, due to insufficient blood supply to the brain due to carotid artery occlusion. (b) Spinal cord ischemia, due to blockage of
blood supply to the spinal cord and especially to the artery of Adamkiewicz. (c) Peripheral nerve ischemia, due to decreased blood flow to the limbs, e.g., blockage of the subclavian artery or iliac artery. (d) Aneurysm pressure phenomena, for example, compression of the superior cervical ganglion causing Horner's syndrome.

Zull and Cybulka\(^1\) reviewed 1805 cases reported with aortic dissection, and found that 4.2% of cases manifest acute paraplegia, and a higher incidence of distal than proximal dissection. The system of blood supply to the spinal cord is very complicated (Figure 2).\(^{1,13-17}\) The cervicothoracic territory of the spinal cord, richly supplied by the vertebral and the costocervical arteries, is not susceptible to damage. The midthoracic and thoracolumbar territory is supplied by direct branches of the aorta, the most important of which being the Adamkiewicz artery from T8 to L1, and these direct branches of the aorta are most susceptible to aortic dissection. The Adamkiewicz artery originates from the intercostal artery and/or lumbar artery and supplies most of the blood to the anterior spinal artery, which perfuse the anterior two third of the spinal cord.\(^{15}\) The Adamkiewicz artery is relatively vulnerable because it is the last major vessel feeding the spinal cord and it makes an acute angle of entry, which may contribute to its vulnerability.\(^{16}\) There is no functioning anastomosis between the anterior and posterior circulation even though communications may be demonstrated.\(^{16}\) The lower thoracic spinal cord is most vulnerable to ischemia because of the relative paucity of blood vessel anastomoses in that region.\(^{17}\) Once the Adamkiewicz artery is blocked, there will be severe damage to the thoracolumbar territory of the spinal cord.

![Diagram of blood supply to the spinal cord]

Figure 2. Blood supply to the spinal cord. (A) The midthoracic and thoracolumbar territory is supplied by direct branches of the aorta, the most important of which being the Adamkiewicz artery from T8 to L1, and these direct branches of the aorta are most susceptible to aortic dissection. (B) a: Aortic dissection from T4 to T10 level in this case; b: Blood supply of midthoracic territory affected by aortic dissection that caused thoracic cord atrophy, especially from T6 to T8 level, in this case. (Redrawn from Reference 14)
Spinal cord injury caused by aortic dissection is usually incomplete \[^{[109]}\] with the manifestation of anterior spinal artery syndrome, in which the joint position sense, sense of vibration and sacral sensation were preserved. This resultant anterior spinal artery syndrome is due to the fact that the anterior spinal artery supplies blood to the anterior two thirds of the spinal cord and its collateral circulation is significantly less than that of the posterior spinal artery. According to Waters et al., \[^{[110]}\] recovery from ischemic myelopathy was poor, especially for the motor component. The patient presented with bilateral lower limb weakness, loss of pinprick and fine touch sensation, but well preserved proprioception sensation. Voluntary contraction of the anal sphincter, anal tone and anal sensation were normal, compatible with the clinical manifestations of the anterior spinal artery syndrome. A clinical diagnosis of anterior spinal artery syndrome has been attributed to various etiologies including: \[^{[111]}\] syphilitic arteritis, hypotension and cardiac arrest, intravenous neurosphenamine, angiography, sympathectomy, atheromatous emboli, aortic surgery, and intervertebral disc herniation. Anterior spinal artery occlusion demonstrated at autopsy is reported in cases of syphilis, cervical spondylosis, arteriosclerosis, and vertebral subluxation. Tracking the patient’s history, there nothing listed above was found. There was no previous history of acute precordial, abdominal or back pain. There was gradual progressive lower limb weakness. MRI revealed marked thoracic cord atrophy and myelomalacia, indicating long-term insufficiency in the blood supply to the thoracic cord, \[^{[112,113]}\] and consequent gradual necrosis might not be caused by an episode of acute infarction.

To our knowledge, painful aortic dissection presenting as progressive ischemic myelopathy for years has had not been reported so far. We propose that the hemodynamic of the segmental arteries was influenced by Type 3 aortic dissection coupled with poor collateral circulation, which leads to long-term insufficient blood supply to the thoracic cord causing thoracic cord atrophy and neurological impairment.

**CONCLUSION**

According to reports in the literature, aortic dissection always has acute onset, and in a small percentage of cases, some degree of recovery can be achieved over a period of time. \[^{[122]}\] Among 23 papers reviewed, only one disclosed that a patient suffered from a painless aortic dissection and progressive neurologic symptom over 5 days until a complete transverse myelopathy. \[^{[21]}\] But in the present case we reported, onset was gradual over years, and the neurological deficit (i.e. degree of paraplegia) was progressive. With the passage of time, symptoms became more marked and severe, as a result of long-term insufficiency of blood supply. We have presented an unusual case hoping that it will be of interest and use to the medical community.

**REFERENCES**

無痛性主動脈剝離引發進行性缺血性脊髄病變：
病例報告

闕弘昌 林樺貞 黃力升
行政院衛生署新竹醫院復健科

主動脈剝離之臨床症狀包括突發性的胸痛或腹痛，肢體發冷及肢體出汗，出現如同休克般的症狀，有些病人會出現肢端消失或不對稱的情形。假如某些特定器官的支配血管正位於剝離段前造成血腫的位置，就會產生組織突發性的缺血性傷害，如脊髄的缺血，進而造成脊髄的傷害。在過去的文獻中顯示大約有 20%主動脈剝離的病人會因血液供應的問題而合併神經學的症狀，而大部分主動脈剝離的病人都會有嚴重的急性胸痛或腹痛之症狀。在本篇文章中，我們要報告一個脊髄長期缺血並以進行性神經缺損來表現的病例，而長期缺血的原因可能來自於無痛性的主動脈剝離。這是一位 66 歲的男性病患，到院時以下半身輕微癱瘓表現，檢查後發現其雙上肢肌力為 5/5 而雙下肢肌力則為 3-4/5，刺痛覺在兩側第六胸椎以下異常，輕觸覺在右側第九胸椎與左側第十一胸椎以下異常，震動覺與本體感覺則皆屬正常，深肌腱反射在兩下肢均有異常增強的情形，肛門的主動收縮正常且肛門反射與球體海綿體肌反射均正常，核磁共振影像顯示降主動脈在 T4 到 T10 處有剝離的情形並伴於椎體內發現血腫，此外，病患之胸部脊髄則有明顯萎縮的現象，以 T6 到 T8 處最為明顯，從目前可見的文獻資料，並無類似的報告被提出。（中華復健醫誌 2002; 30(4): 251 - 258）

關鍵詞：主動脈剝離(aortic dissection)，脊髄病變(myelopathy)，脊髄損傷(spinal cord injury)