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CASE REPORT

DeBakey type 1 aortic dissection presenting as abdominal pain with paraplegia: case report and literature review

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ABSTRACT

Background: Aortic dissection (AD) is a catastrophic condition that represents a challenging diagnosis for emergency physicians. Its presentation can vary widely and may present with non-specific symptoms. A delay in the diagnosis of AD is associated with a high rate of morbidity and mortality.

Case Presentation: A 77-year-old man presented to the emergency department with a history of acute epigastric abdominal pain and bilateral lower limb weakness. A suspected AD was confirmed by urgent computed tomography angiography in less than 1 hour; which showed extensive dissection of his aorta from ascending part up to the abdominal aorta; "DeBakey1." The emergency complex aortic repair was then performed by the cardiothoracic surgical team that had resulted in a successful fortunate recovery.

Conclusion: DeBakey type1 AD is a serious cardiovascular surgical emergency, with a dismal prognosis if missed in the emergency room. Our prompt diagnosis and expedited approach in handling this case led to successful recovery.

Keywords: Aortic dissection, DeBakey type1, computed tomography angiography, acute paraplegia, ischemic neuropathy, bentall pProcedure.

Introduction

Although the prevalence of aortic aneurysm and dissection is increasing [1], the condition remains relatively uncommon. Improvements have been made in diagnostic strategies and imaging, but a significant number of aortic dissection (AD) cases are still missed in the emergency department. Physicians make a correct diagnosis in as few as 15%-43% of confirmed AD cases [2], and yet if the condition is left untreated, mortality is as high as 50% in the first 48 hours after onset [2].

Two main systems are used to classify AD: the Stanford and the DeBakey. The Stanford system is more frequently used. It classifies dissections based on whether an ascending or descending part of the aorta is involved. Type A involves the ascending aorta, and type B originates distal to the left subclavian artery and involves only the descending aorta. The DeBakey classification is based on the dissection's site of origin. Type 1 involves the ascending aorta, the arch, and the descending aorta. Type 2 originates in and is limited to the ascending aorta. Type 3 begins in the descending aorta and extends distally above the diaphragm (type 3a) or below the diaphragm (type 3b) [2].

Neurologic symptoms associated with AD are often severe and can completely mask other symptoms, posing significant diagnostic challenges [3]. However, many potential difficulties attend the diagnosis of any AD, because a dissection can mimic other common conditions, and not all affected patients present with the typical sudden and severe tearing chest pain [4].

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Case Description

A 77-year-old Saudi man, with known hypertension being treated with medication, presented to the emergency department, Al Habib Hospital, Riyadh, Saudi Arabia. He complained of severe epigastric abdominal pain accompanied by lower limb pain and weakness persisting for the preceding hour. The patient was agitated and refused to provide a history, which was challenging. When his family was contacted, they reported that he had developed chest pain 3 hours earlier at home and that his blood pressure reading at that time was high, although the episode had resolved and progressed to the current symptoms. The patient's bilateral lower extremity weakness continued to progress rapidly, to the point where he experienced a complete loss of sensation and motor control of his lower extremities. No further history could be obtained because the family was not aware of any other event.

Given the patient's agitation, the Glasgow Coma Scale was administered, and he scored 15/15. On clinical examination, the patient was noted to look acutely ill, with a heart rate of 52 bpm, blood pressure of 169/66 mmHg, a temperature of 36.5°C, respirations of 29/ minutes, and oxygen saturation of 94% in room air. On physical examination, decreased breath sounds were noted bilaterally in the lower lobes. Distal pulses were symmetric. The abdomen was soft, non-tender, and mildly distended, with no bruits or palpable pulsatile masses. Although agitated, the patient was alert and oriented to person, place, and time.

Muscle group testing revealed 5/5 power for all groups in both arms, and upper limb sensation was intact. Power was 0/5 for all muscle groups in the legs, with the patient being unable to move his legs at all. Sensation to light touch and pinprick was absent from *T*6 through *S*5. Pulses in the lower limbs were undetectable manually and with Doppler ultrasonography.

Electrocardiography showed sinus bradycardia with non-specific ST changes. Bedside echocardiography showed normal contractility and left ventricle function, minimal pericardial effusion, and a mural hematoma at the aortic root.

The foregoing data raised suspicion of an AD. Urgent computed tomography angiography (CTA) was therefore arranged. The procedure was explained to the patient and his family. All were hesitant, with the patient refusing all investigations. The risk of death associated with delay of management was then explained, and the family consented to the angiographic imaging.

Fortunately, on urgent bases, he underwent CTA under minimal sedation. The results showed extensive AD starting from ascending aorta and including the aortic arch, descending aorta, and abdominal aorta. Additional dissections were also evident in the brachiocephalic, right subclavian, and right common carotid arteries. Furthermore, thrombosis with various degrees was found in common iliac and external iliac arteries bilaterally (Figure 1A,B).

Cardiothoracic surgery was then performed, with an initial Bentall procedure (Figure 2) that included aortic root replacement, resuspension of the three aortic commissures, aortic valve repair, and total ascending aorta replacement. That complex procedure was then followed by Aortic hemiarch reconstructive replacement. Fortunately, the patient survived the complex surgical procedure and was sent to the cardiac surgical intensive care unit for recovery.

Discussion

AD is reported to present with neurologic symptoms in one-third of cases [3], but it can manifest in various ways depending on the site. A dissection can mimic other disorders, and the diagnosis is therefore often delayed or missed. Few are diagnosed in the emergency

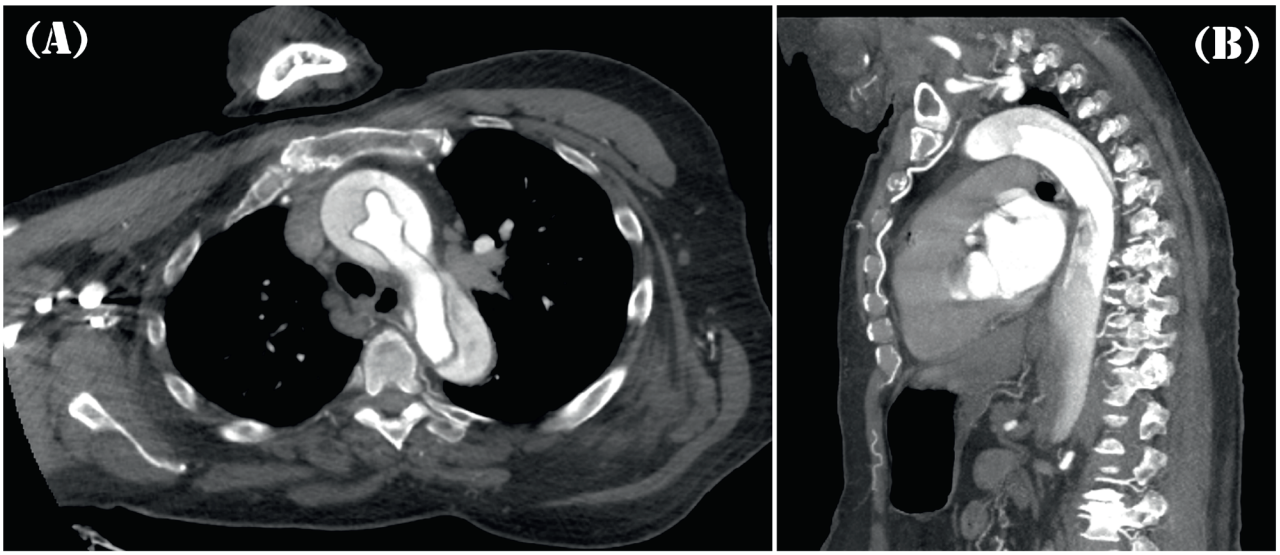

Figure 1. *(A) Aneurysm with a large dissection starting from the ascending aorta and (B) including the aortic arch, descending, and abdominal aorta.*

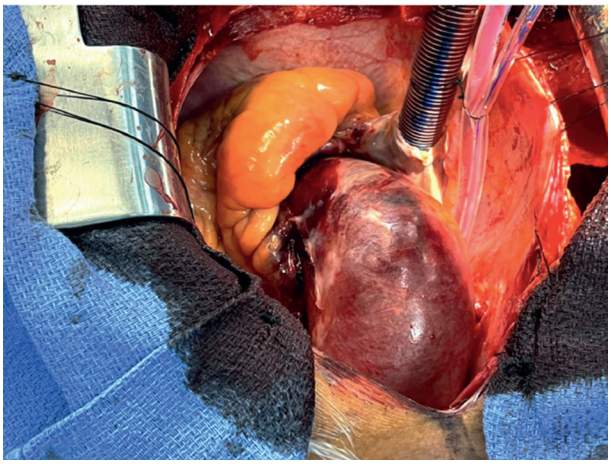

Figure 2. *Intraoperative image of the ascending aorta Bentall procedure.*

department, and most are confirmed after admission for other differentials [5]. In a comprehensive chart review of AD cases, Chua et al. [5] in the Emergency Medicine Department of the National University Health System (Singapore, PRC) analyzed 68 patients who presented during the study period, finding a missed diagnosis in 38.2%.

Donovan et al. [6] reported a case of painless AD presenting as high paraplegia in 2000 at the Rehabilitation Institute of Michigan (Detroit, MI). The patient was admitted with a diagnosis of pneumonia and paraplegia. On hospital day 4, chest radiography repeated to monitor pneumonia showed increased density in the left retrocardiac region. CTA of the chest was ordered to rule out an AD but detected a Type A dissecting aneurysm. Improvement of symptoms after management for another condition cannot rule out AD, as shown by a case of acute AD presenting with chest pain relieved by sublingual nitroglycerin as reported by Kim at Seoul National University (Seoul, ROK): a 77-year-old lady with chest pain was worked up as a case of an acute coronary syndrome, only to later find that she had AD Type A, as confirmed by CTA [4].

In our reported case, ischemia to the spinal cord "i.e., ischemic neuropathy" was very likely the cause of his acute paraplegia [6,7].

CTA is the imaging modality of choice for the diagnosis of AD [8]. CTA can reliably identify a false lumen and might provide additional details such as the anatomy of the dissection, placement of the dissection flap, extension of the flap into the great vessels, signs of aortic rupture, and signs of end-organ damage. Protocols should include images with and without IV contrast [9].

Although AD does not usually have a clear presentation and can mimic other common conditions such as myocardial ischemia, pneumonia, heart failure, stroke, and acute abdomen, chest pain is the most frequently reported symptom [4]. The pain is typically substernal, interscapular, or in the mid-back. In 5%-10% of patients, the dissection can be painless or the pain might be intermittent. Atypical symptoms of AD should therefore always be considered [6]. Vascular insufficiency can occur in any branch vessel and can lead to clinical syndromes that include (but are not limited to) acute myocardial infarction, stroke, paraplegia, paraparesis, mesenteric ischemia, and limb ischemia [10], similar to what occurred in our case.

Although AD is relatively uncommon, clinicians must always be aware of the possibility and should consider it in the differential diagnosis for any atypical complaints or the sudden onset of neurologic symptoms.

Conclusion:

To our knowledge, no case of DeBakey Type1 AD with acute paraplegia has been previously reported in our region. The present case emphasizes the importance of prompt clinical suspicion for AD in the emergency department. Moreover, it highlighted the various potential clinical presentations and the best diagnostic method.

Given the published reports of missed cases, clinicians must always be aware of the potential for AD in patients who visit the emergency department with atypical complaints or the sudden onset of neurologic manifestations. Emergency physicians can face many challenges in diagnosing AD, but they should be alert to its wide range of presentations.

List of Abbreviations

AD	Aortic dissection
CTA	Computed tomography angiography

Conflicts of interest

The authors declare no conflicts of interest related to this report.

Consent for publication

Written consent was obtained from the patient's guardians to publish this case report and accompanying images.

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Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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