JNDS Journal of the Nuffield Department of Surgical Sciences

Case Study

Aortic dissection complicated by paraplegia

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Key Learning Points

This case report illustrates the complexity and severity of acute aortic dissection. This condition has one of the highest mortality rates of any cardiovascular emergency and is often extremely challenging to treat with both open and endovascular intervention often required. This patient presented with a ruptured aortic dissection which is rare and often immediately fatal. He survived urgent extensive aortic endovascular stenting, but, despite preventative measures, developed spinal cord ischaemia post-intervention. The contemporary management of acute aortic dissection, and the pathophysiology and prevention of spinal cord ischaemia are covered in this fantastic case report.

Keywords:
Aortic dissection, paraplegia, TEVAR, spinal cord ischaemia.

Introduction

Acute aortic dissection is a medical emergency accompanied by high morbidity and mortality[1]. This is highlighted by the pioneering review of 505 patients reported by Hirst et al. in 1958[2]. Advances in surgical intervention have subsequently lowered the mortality rate[2,3]. Aortic dissection remains one of the most common aortic emergencies with an incidence of 3 per 100,000 patients, with a male predominance of 65%[4]. This condition is characterized by the separation of the layers of the aortic wall allowing entry of blood into the intima-media region; hence the dissection is further propogated[1]. The resulting compromised perfusion and systemic ischaemia is responsible for the characteristic severe pain radiating to the back[5]. Management ranges from medical analgesia, vasodilators and beta-blockers, to surgical intervention including open surgery, implementation of an endovascular stent graft, or a combination of these solutions[5]. Associated morbidities include rupture, stroke, acute renal failure, bowel ischaemia, peripheral ischemia and even paraplegia[5]. This report focuses on the rare latter complication, paraplegia, experienced by Mr X.

Case presentation

Mr X is a 67 year-old man who was admitted to the John Radcliffe with a thoracic aortic dissection. Two days prior to admission, Mr X experienced a 'sudden shooting pain' down the left-side of his back when he was getting out of his chair in order to go to bed. He continued to retire to his bed but the pain continued through the night and kept him awake. As the pain was still present, Mr X decided to drive to his local GP in the morning, but en route he experienced a very severe pain, like a 'stabbing in the back'. It was so severe that he 'couldn't breathe' and had to emergency stop the car. Mr X was able to turn the car around to return to his house and call 999 for an ambulance, before admission into ICU.

Mr X's past medical history shows recurrent pneumothoracies leading to a bilateral pleurectomy as well as a prostate removal, following the diagnosis of prostate cancer. He was not on any medication prior to the surgery and has NKDA. Mr X was an ex-smoker but there was no other significant social, or family history.

Upon admission to ICU, Mr X was conscious and the pain had marginally subsided. However, he presented with the complication of hypotension. An MRI was taken, which confirmed the diagnosis of a ruptured Type B aortic dissection from the subclavian to his common iliac artery. This case was managed surgically with a Thoracic Endovascular Repair of Aneurysm (TEVAR). Pre-operation Mr X was stable but tachycardic, and a small right-sided haemothorax was visible on CT. The operation involved placing the main body of the stent through the right groin. The deployment sequence was conducted using two proglides on the right side with manual pressure on the left side. He was under anaesthetic for 2.5 hours. CT scans were taken after the operation and Mr X remained in ICU for 4 days. Post-operation, Mr X suffered from the complication of paraplegia from level T5 below, due to spinal cord ischaemia. Many procedures were implemented to minimize the risk of this

occurring during the surgery; this includes a spinal drain, O_2-Hb transfusion, maintaining a high MAP, as well as placing Mr X in supine position in ITU. Unfortunately, these measures were not able to prevent the complication of paraplegia. The spinal ischemia has led to Mr X becoming double incontinent post-surgery. During his stay in hospital, he also developed a chest infection with crepitations in the right lower base of his lungs; this was resolved following administration of co-amoxiclav. The current plan is to move Mr X to a specialist re-habilitation centre so that he can commence physiotherapy and begin, in his words, 'a new chapter in his life' adjusting to the paraplegia. The complication of paraplegia following TEVAR to manage aortic dissection will be explored in this case report.

Discussion

Aetiology of aortic dissection

The pathophysiology of aortic dissection is not completely understood. One hypothesis proposes an initial tear in the intima of the aorta, allowing blood to surge into the media and create a false lumen[6]. Another hypothesis postulates that the outer portion of the media (vasa vasorum) haemorrhages initially, which then leads to intimal rupture[6]. Common to both theories, blood then continues to flow, extending the dissection typically in an anterograde manner[6].

The predisposition to aortic dissection has both histopathological and genetic components[4]. The most prevalent risk factor is hypertension and is present in 75% of cases[4]. Other modifiable risk factors include smoking and drug use (such as cocaine and amphetamine)[4]. Traumatic aortic dissections are most commonly caused by traffic accidents or deceleration trauma[4]. The importance of inflammation in the pathophysiology of aortic dissection is demonstrated by the increased pre-disposition in patients with inflammatory disorders[7]. This includes vascular autoimmune diseases such as Giant-cell arteritis and Takayasu's arteritis, as well as infections such as tuberculosis and syphilis[4]. There are many genetic risks linked to aortic syndromes; a large majority are connective tissue disorders including Marfan's syndrome, Turner's syndrome and Type 4 Ehlers-Danlos syndrome[4].

Classification

Classification systems are in place in order to describe the type of aortic dissection. This grouping is beneficial in deciding the course of management. The two most prevalent classification systems are the DeBakey and Stanford systems[4]. These classify the dissections in an anatomical manner, referring to the site of intimal tear[4]. As a part of the classification, the ascending aorta refers to the section of the aorta proximal to the brachiocephalic artery and the descending aorta is distal to the left subclavian artery[4]. The DeBakey system categorises the dissection type based on where the intimal tear originates whereas the Stanford system is based on whether or not the ascending aorta is involved[4,6]. This is further expanded upon in the Table 1 and illustrated in Figure 1. In the case study, the patient had a type B aortic dissection.

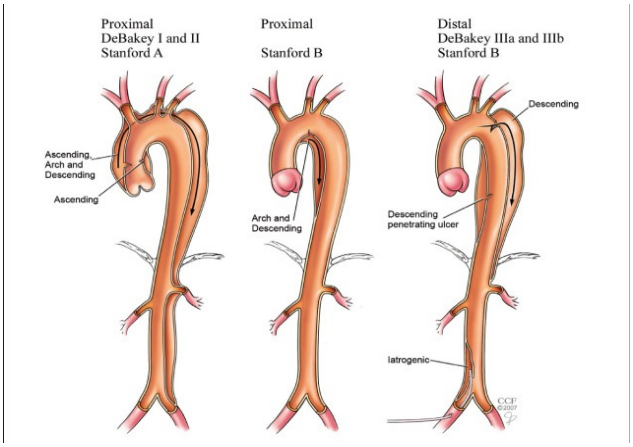


Figure 1 - Aortic dissection classification from the DeBakey and Stanford classification system. Taken from Loebe et al.[24]

DeBakey	
Category	Description
I	Tear in the ascending aorta propagating distally to include the aortic arch and also the descending aorta typically.
II	Tear is only present in the ascending aorta
III	Tear in descending aorta often propagating distally
IIIa	Tear only in the descending thoracic aorta
IIIb	Tear extends below the diaphragm

Stanford	
Category	Description
Type A	Dissections involving the ascending aorta (regardless of tear site)
Type B	All other dissections that do not involve the ascending aorta (but can involve all other parts of the aortic arch).

Table 1 - A table to show the DeBakey and Stanford classification systems.

Investigations and diagnosis

Rapid diagnosis in this potentially life-threatening medical condition is crucial in order to ascertain the correct management pathway. A review by Hagan et al. shows that 63% of patients with Type A dissection and 56% of type B dissections had mediastinal widening on the chest radiograph[1]. No abnormalities in chest radiography was reported in 12% of patients[1]. In the case of Mr X, pleural haematomas were noted in the chest radiograph, as shown below in Figure 2 by the left mid and lower zone opacification. This radiograph also demonstrated incorrect placement of an NG tube, which was later rectified.

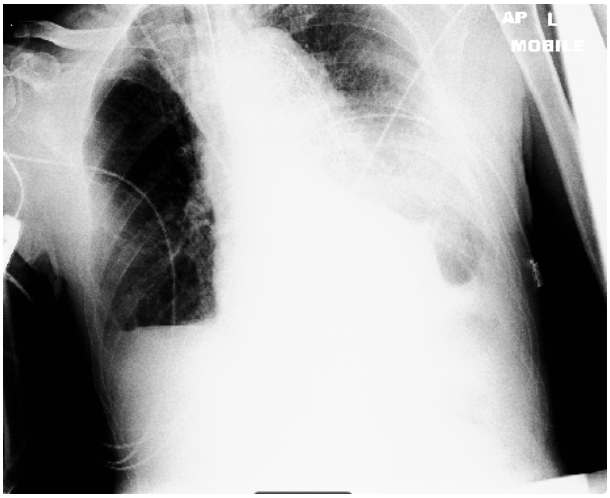
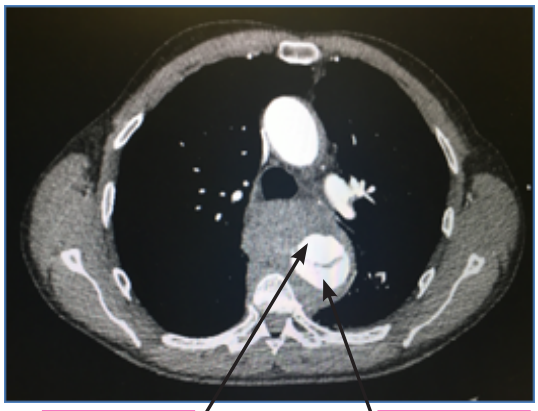


Figure 2 - Chest radiograph of Mr X taken post-operatively showing pleural haematomas and a misplaced NG tube.

Another initial investigation includes the 12 lead ECG. In the review by Hagan et al., non-specific abnormalities in the ECGs were shown, however, results were normal for 31% of patients[1]. Imaging studies are employed as a diagnostic tool including contrast-enhanced CT Angiography, particularly in type B dissection[1]. In the case of Mr X, the dissection began just after the left subclavian artery, extends to the bifurcation and into the left common iliac artery. This is seen on the CT angiogram: Figure 3a and 3b below. The false lumen can be identified by the darker shading. The image also shows a large mediastinal haematoma and bilateral haemothoracies.

(a)



(b)

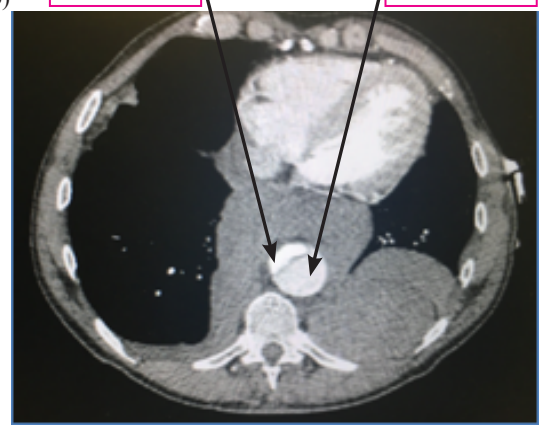


Figure 3a and 3b- CT angiogram showing a Type B aortic dissection iwth large mediastinal and pleural haemoatomas.

Other diagnostic imaging includes transthoracic echo, transoesophageal echo. MRI is also used, although rarely[1,4]. Biomarkers are another key diagnostic tool when looking to the future for diagnosis. Markers that show injury to the vascular smooth muscle, interstitium and elastic laminae can indicate dissection[4]. Currently, only D-dimer is used clinically to determine suspected aortic dissection[4]. As a future prospect, fibrin degradation products can be assayed as a marker in acute dissection[4].

Management of Aortic Dissections

Depending on the type of dissection, and the level of complication, management can be purely medical or surgical (endovascular, or open). The chosen management options for aortic dissections are shown in a flowchart below in Figure 4.

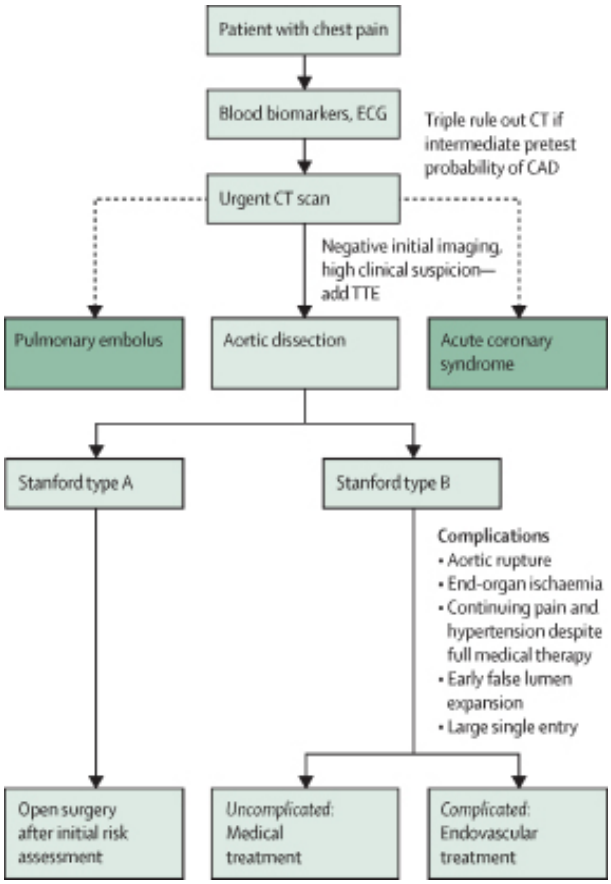


Figure 4- A flowchart demonstrating the management route depending on the classification and complication of aortic dissection. Taken from Nienaber et al.[4]

The medical management first aims to provide analgesia. The next priority is to control the blood pressure and to reduce the force of left ventricular ejection[4]. This, in turn, limits the propagation of the dissection[4,8]. The aim is to achieve a blood pressure of 100-120 mmHg[8]. Beta-blockers are used for blood pressure control, such as labetolol, as in the case of Mr X. This can be used in combination with vasodilating drugs such as ACE inhibitors, including ramipril, used in this case.

Surgical management includes both open repair as well as implementation of an endovascular stent (TEVAR). Surgical treatment, as opted for in type A dissections, aims to remove the entry into the false lumen and remodel the aortic true lumen with a graft (with or without re-implantation

of coronary arteries)[1]. 30-day mortality for ascending aortic dissection at experienced centres is between 10-35%[9]. From a propensity matched retrospective analysis, survival rates in patients with acute type A dissection were 91% after 30 days, 74% after 1 year and 63% after 5 years[9]. Therefore, early open surgery is a suitable solution. However, there has been recent movement towards endovascular repair. As following standard protocol, thoracic endovascular repair (TEVAR) was used as the management plan for Mr X's complicated type B aortic dissection; this is the first line therapeutic option[8].

TEVAR

Endovascular repair was introduced in 1999 by Dake et al. and has significantly reduced the mortality rates compared to when the only surgical solution was open repair-[10]complicated dissections. Thoracic endovascular aortic repair (TEVAR) is a minimally invasive procedure. It uses stent grafts to seal the primary tear and allow blood flow through the true lumen[10]complicated dissections. It is recommended that there is minimal aortic coverage in order to minimize spinal cord ischemia[10]complicated dissections. If there is poor perfusion of the branch vessels, endovascular revascularization may be performed by fenestration or branch vessel stenting, however this is not usually done in an emergency setting[10]complicated dissections. The stent used in the case of Mr X is shown in the CXR and CT image in Figure 5 below.

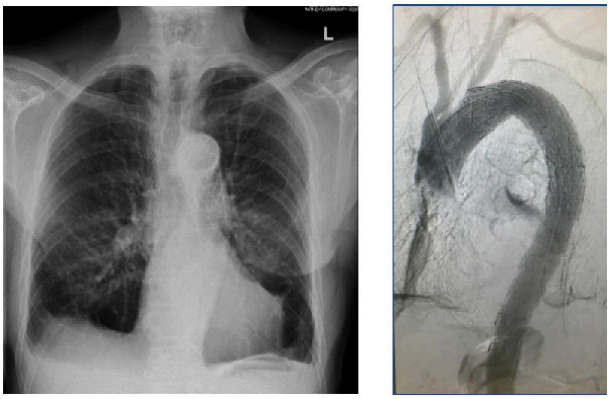


Figure 5- A CXR (left) and CTA (right) showing the stent graph thoracic aorta used to manage type B aortic dissection. The graft was placed on the left subclavian origin.

The concept of repairing type B dissections without the need for open surgery, and its associated risks, is very valuable. The stents are composed of dacron or polytetrafluoroethylene with a stainless steel or nitinol skeleton. The procedure is carried out under X-ray fluoroscopic guidance and involves passing a device through the common femoral artery to an access sheath. This sheath is removed eventually to expose the stent.

The meta-analysis for complicated type B dissections by Parker et al. compared a total of 942 patients from 29 different studies[10,11]complicated dissections. In-hospital mortality was 9% and other major complications including stroke (3.1%), paraplegia (1.9%), conversion to type A dissection (2%), bowel infarction (0.9%) and major amputation (0.2%) occurred in 8.1%[11]. Overall, technical success was achieved in 95% of the cases, with an in-hospital mortality of 9%[10]. This meta-analysis shows a promising solution to aortic dissection with endovascular repair.

Data collected from large registries show that hospital mortality is 32% for patients treated with surgery, 7% for endovascular techniques, and 10% for patients treated with only medical management[4]. Booher et al. created a Kaplan-Meier survival curve from the IRAD database for type B aortic dissections (Figure 6)[12]. This identifies the treatment option along with the time period from onset, and its effect on mortality.

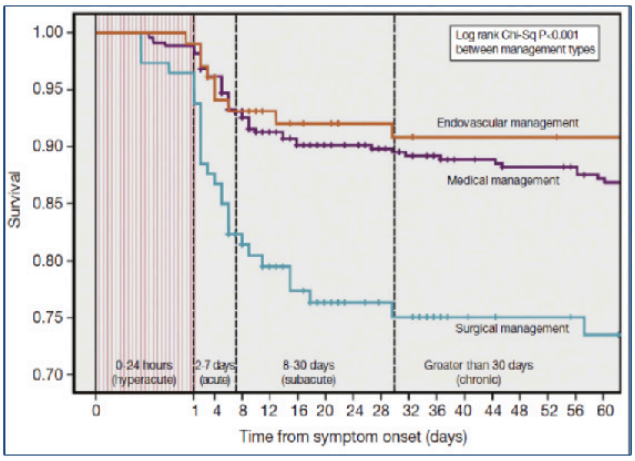


Figure 6- A Kaplan-Meier survival curve indicating the treatment option and time period from onset, and its effect on mortality. Taken from Booher et al.[11]

The question still remains as to how it is best to treat a Type B dissection, as in Mr X's case. The INSTEAD trial explored this by randomizing patients with uncomplicated type B aortic dissection between 2-52 weeks from onset into medical management or TEVAR management. 5-year mortality was 11.1% for TEVAR compared to 19.3% for purely medical management[6]. Open surgery was compared to TEVAR by Fattori et al., in the International Registry of Acute Aortic Dissection (IRAD)[13]. The study reviewed 571 patients with acute descending dissection. 10% of patients had open surgery and 12% had endovascular repair[13]. There was a much better in-hospital mortality for TEVAR (10%) than open surgery (34%)[10,13].

Spinal cord ischemia and paraplegia

Aortic dissection endovascular repair has been associated with great success, but unfortunately there are rare but disastrous complications following a dissection which cannot be prevented. This includes paraplegia as in the case of Mr X. Although there are advantages of TEVAR when compared to open repair, there is still a significant incidence of spinal cord injury; the overall incidence ranges from 2.5-8%[8,14]. Scali et al. found a 9.2% incidence in a study looking at 741 TEVAR procedures[15].

The spinal cord ischaemia was caused by the temporary obstruction for the spinal arteries, especially in critical zones such as the lower thoracic and lumbar segments[16]. This ischaemia of the spinal cord, which was found to be from the level T5, lead to the paraplegia. There are many cases that have reported this severe, although rare, complication. Weisman and Adams in 1944 described 38 cases of ischemic necrosis of the spinal cord following aortic dissection[16,17]. They proposed that paralysis occurred following occlusion of the intercostal and lumbar arteries by dissection of the aortic wall.

The development of paraplegia can be classified as immediate or delayed[18]. The former is a direct result of hyperperfusion of the spinal cord as well as secondary hypoxic

damage[18]. On the other hand, delayed complications (which can be up to 21 days following the surgery) are caused by reperfusion hyperaemia and free radical generation[18]. This then leads to oedema of the cord with hypotension in certain regions and reduced perfusion of the vasculature[18]. The latter is more associated with TEVAR with respect to open repair[14]

There many factors that contribute to the occurrence of spinal cord ischaemia during and after aortic surgery[18]. Three key aspects were identified by Svensson et al.: the duration and degree of ischemia, the failure to re-establish blood flow to spinal cord after repair, and biochemically mediated reperfusion injury[19]. When looking at the TEVAR procedure specifically, spinal cord injury has been linked to the aortic coverage levels, a history of prior aortic surgery as well as hypotension at presentation[8]. The latter was present in the case Mr X.

Distal dissections have been found to have a greater incidence of spinal cord ischaemia[20]. The spinal cord has both a complex, as well as a varied blood supply[20]. The vertebral artery and the costocervical trunk supplies the cervical and upper thoracic cord[20]. This part is less prone to vascular insult[20]. The lower half of the spinal cord is supplied by direct branches from the aorta, this includes the intercostal, lumbar, iliolumbar and sacral arteries[20]. Here the major arterial supply of the cord is from T10-L1 and is known as the artery of Adamkiewicz[20]. These arteries in particular are sheared in aortic dissection[20]. The subsequent interruption of blood flow has a maximal insult on the mid-thoracic cord, as this area is a watershed zone between blood supply of the upper and lower cord[20].

Perioperative preventative measures

Many strategies have been put into place to prevent the incidence of spinal cord injury and consequent paraplegia during the endovascular repair of the dissection. These measures are attributed to the declining incidence of paraplegia[8]. Based on the three key contributing factors, spinal cord protection methods have been implemented. In the case of Mr X, as mentioned above, CSF drainage, maintenance of MAP, and maintenance of a supine position was used.

The drainage of CSF acts to reduce the severity of ischaemia. Animal studies have shown that decreasing the spinal fluid pressure lead to a decrease in incidence of paraplegia[21]. This can be accomplished by CSF drainage and has been put into place clinically, with high risk patients being given CNS drainage and naloxone[18]. This holds a slight controversy following a study that failed to show any benefits of CNS drainage alone[22]. This study has been criticized as there was a small volume of drainage (50ml) and the drainage was not by free gravity[18]. Subsequently, more encouraging clinical results were found by Svensson et al., which allowed drainage freely by gravity [23]. This study showed CSF drainage to be protective and since, CNS drainage is used as one of the key methods for spinal cord protection. Further methods of spinal cord injury protection include avoiding perioperative hypotension and creating a temporary endoleak, both allowing for sufficient perfusion[8]. Adjunct protective methods include perioperative induction of hyperthermia and intrathecal medication[8,18]. Finally, during surgery itself, staging the procedure has shown spinal cord neuroprotection[14].

Although these measures are currently used in practice, there is no definitive recommendation for spinal cord injury prevention for TEVAR from current literature; there are no randomized controlled trials evaluating any of the preventive measures. The rationale behind the strategies used are drawn from those used in open surgery, as well as the basis of theoretical spinal cord injury pathophysiology[8].

Conclusion

The treatment of aortic dissection is still associated with significant morbidity and mortality. The progressive evolution in operative techniques, including TEVAR, has been able to improve this. The case of Mr X demonstrates a very severe, although rare complication following aortic dissection and highlights the employment of techniques used to achieve spinal cord protection. The cause of the post-operative neurological complication is now mostly understood and therefore targeted in the protective methodology. By directing our efforts towards the three major contributing factors – the duration and degree of ischemia, failure to re-establish blood flow to spinal cord after repair, and biochemically mediated reperfusion injury – we can aim to reduce the complication of paraplegia due to spinal cord ischaemia.

Conflicts of interest

None.

Funding

None.

Consent

The patient has consented for the publication of this case study.

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