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CASE REPORT

Multiple Acute Aortic Dissection in a Young Adult

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ABSTRAK

Diagnosa pembelahan aorta secara spontan jarang dilaporkan berlaku pada golongan dewasa muda, terutamanya tanpa aterosklerosis atau trauma. Lebih jarang lagi adalah kehadiran serentak pembelahan aorta dan aneurisma aorta dalam ketgori umur dewasa muda. Kami mengetengahkan suatu kes yang melibatkan pembelahan aorta dan aneurisma aorta yang dilihat berlaku pada seorang lelaki berusia 35 tahun. Computed tomographic angiography (CTA) menunjukkan pembelahan aorta jenis Stanford B dan pembelahan aorta daripada bifurkasi arteri iliac kanan sehingga arteri femoral. Aneurisma aorta juga dilihat melibatkan asal arteri subklavian. Prosedur hibrid melibatkan gabungan pembedahan terbuka dan endovaskular telah dijalankan.

Kata kunci: aneusima aorta, pembelahan aorta, prosedur endovaskular, mortaliti

ABSTRACT

The diagnosis of aortic dissection in a young adult in the absence of atherosclerosis or prior history of trauma is extremely rare. The presence of more than one arterial dissection site in such a patient is even more unheard of. We highlight a case of spontaneous multiple acute arterial dissections occurring in a 32-year-old male. Stanford B aortic dissection and a separate dissection extending from the bifurcation of the right common iliac artery to the right common femoral artery was noted on computed tomographic angiography (CTA). A small aneurysm of the right subclavian artery was also noted. A two-stage hybrid procedure involving a combination of open and endovascular surgery was employed. The rarity and lethality of this condition warrants a high index of suspicion for early diagnosis and prompt intervention.

Keywords: aortic aneurysm, aortic dissection, endovascular procedure, mortality

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INTRODUCTION

First described by Dr. Nicholls almost 200 years ago, acute aortic dissection has become a feared cause of acute severe inter-scapular or chest pain radiating to the back (Hirst et al. 1958). Acute aortic dissection refers to a sudden split of the intimal layer occurring within two weeks of onset of symptoms, while the term chronic aortic dissection is reserved for dissection that persist for more than two weeks. The speed, lethality and rarity of this clinical entity warrants high index of suspicion in order to execute prompt and definitive treatment. Seen largely in men above the age of 60 years old (Hagan et al. 2000), aortic dissection can be divided into Stanford A and B depending on region of dissection. Stanford A dissection involves the ascending aorta presents more commonly with a higher mortality rate, while Stanford B involves dissection distal to the left subclavian artery (Golledge & Eagle 2008). Unlike in the older age group where trauma, atherosclerosis and previous history of arterial dissection or aneurysm have been implicated as prominent risk factors, the rare occurrence of aortic dissection in the younger age group has been associated with trauma, Marfan's syndrome, uncontrolled essential hypertension, connective tissue disease, bicuspid aortic valve disease, cocaine abuse and during peri-partum period (Januzzi et al. 2004). Recent advances in medical technology have paved the way for less invasive methods of treatment such as endovascular aortic repair. The choice between endovascular and open repair would involve a broad spectrum of criteria extending from clinical to radiological. We aim to highlight an interesting case of multiple synchronous acute arterial dissections occurring in a 32-year-old male.

CASE REPORT

A 32-year-old male presented to the Emergency Department with sudden onset of excruciating pain of the inter-scapular region for the past 12 hours. He described the nature of pain as tearing and stabbing. The initial onset of pain occurred while he was resting in bed. The pain was not relieved by oral analgesics and had increased in severity over the past two hours. He denied prior medical illness, recent trauma or cocaine abuse. At the triage, he was noted to be hypertensive with blood pressure of 217/116mm Hg. His resting heart rate was around 90 beats per minute with oxygen saturation of 98% under room air. Clinical examination of his cardiovascular, pulmonary and gastrointestinal systems was otherwise unremarkable. Echocardiogram revealed no evidence of cardiac ischemia, however chest radiograph showed significant mediastinal widening. On further questioning, he denied experiencing weakness of limbs, claudication or rest pain. Detailed examination of the peripheral pulses revealed an audible bruit over the right femoral artery with monophasic Doppler signals heard over the right femoral artery and arteries distal to it. Capillary refill time, motor and sensory examination of all limbs was satisfactory.

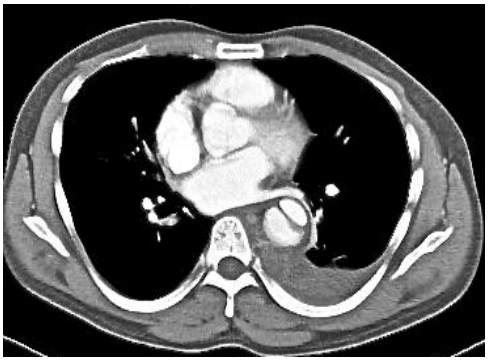

Figure 1a: Contrast enhanced CT angiogram of the thorax reveals an intimal flap within the descending aorta giving rise to a true and false lumen.

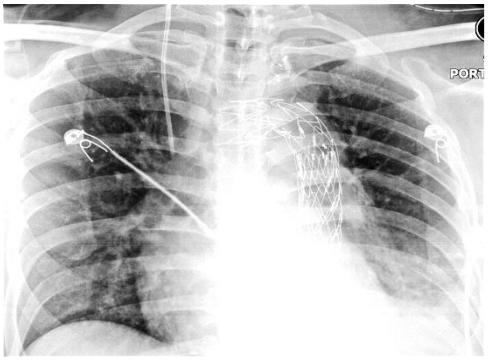

Figure 1b: Chest radiograph day 2 post TEVAR with endovascular stent in situ.

Intravenous infusion of antihypertensive agents was commenced with close cardiac monitoring. An urgent computed tomographic angiography (CTA) of the thorax, abdomen and lower limbs was requested (Figure 1a). CTA revealed multiple areas of arterial dissection. A 9.4 cm Stanford B dissection and another separate arterial dissection extending from the bifurcation of the right common iliac artery to the right superficial femoral artery were noted. An isolated small fusiform aneurysm was noted close to the origin of the left subclavian artery measuring approximately 1 cm in diameter.

A staged procedure involving open surgery followed by endovascular repair of the aortic dissection was performed. The first stage involved sternotomy and placement of interposition bypass graft between the left common carotid and second part of the left subclavian artery. This was followed by plugging of the origin of the left subclavian artery by interventional radiology. During surgery, the wall of the left subclavian artery was noted to be friable. Biopsy specimen of the left subclavian artery wall was taken to rule out connective tissue disease. Two days later, thoracic endovascular aortic repair of the Stanford B dissection was carried out as part of stage 2 management of this patient. Chest radiograph done following TEVAR confimed position of the endovascular stent (Figure 1b). Immidiate post-operative recovery was unremarkable. Patient however seccumbed on day 5 of TEVAR following cardiopulmonary arrest secondary to acute myocardial infarction. Biopsy result from left subclavian arterial wall was negative for connective tissue disease.

DISCUSSION

Hallmark of aortic dissection is intimal injury. Intimal injury permits pulsatile high-pressured blood flow to track between the tunica intima and tunica media. The passage of blood between these two layers raises an intima-medial flap by splitting the tunica media into an inner layer that is also known as the intima-medial layer and an outer

layer. In some cases, this process is augmented by pre-existing medial cystic degeneration. High-pressured passage of blood beneath between the two layers of tunica media gives rise to a false lumen, which is nothing more than a blind track. A large intimal tear leads to a larger false lumen. Secondary tears in the tunica intima can occur due to re-entry of blood into the true lumen from the false lumen (Roberts & Roberts 1991).

Exact pathophysiology of intimal tear has been a topic of much debate. Rupture of atherosclerotic plaque or plaque ulceration, intimal injury due to uncontrolled hypertension, connective tissue disorder and direct iatrogenic injury have all been implicated as causative factor for that matter. However, aortic dissection secondary to intramural hematoma (IMH) has a slightly different IMH occurs secondary to rupture of vasa vasorum. The blood within the media, splits the tunica media layer and give rise to a false lumen. Although there is no direct communication between the true lumen and false lumen in the case of aortic dissection secondary to intramural hematoma, the mainstay of treatment remains the same (Roberts & Roberts 1991; Svensson et al. 2002).

The rarity, varying as well as non-specific clinical presentation of aortic dissection often leads to a missed diagnosis (Khan & Nair 2002). Left untreated, one in for four will die within the first 24 hours and mortality rates could reach up to 90% within two weeks from onset of symptoms (Sommer et al. 1996). The most common presenting symptom is excruciating back pain. Other symptoms include abdominal pain, syncope and weakness or numbness of extremities. The pain is most severe at the beginning and is often localized to the site of intimal tear. Intimal tear that is distal to the left subclavian will manifest as chest pain radiating to the back where else a tear above the aortic valve will present as anterior chest pain. Dissection of thoracoabdominal aorta often presents as lower back or left flank pain. Spread of pain is indicative of progressive dissection (Ledbetter et al. 1999; von Kodolitsch et al. 2000). Painless dissection has been documented to occur in a small percentage of patients. Such asymptomatic dissections tend to involve the elderly, individuals on long-term steroids and those diagnosed with Marfan's Syndrome (Svensson & Crawford 1992). Focal neurological deficits are seen with involvement of the great vessels Hemiparesis occurs following poor perfusion to the spinal vasculature. Sycope is seen following hemorrhage into the pericardial space in proximal dissection. Hemopericardium causes pericardial tamponade that may manifest as syncope or even sudden death. Dissection of the thoracoabdominal aorta is often implicated in causing malperfusion of the kidneys, mesentery and lower limbs. In such instance, chief complaint would be severe abdominal pain and symptoms suggestive of lower limb ischemia such as claudication and rest pain. Perfusion deficit in these organs can result as a consequence of obstruction due to the dissection flap either by dynamic obstruction or static obstruction. Dynamic obstruction is

due to prolapse of dissection flap across a visceral artery. Static obstruction occurs when there is dissection extending into the visceral vasculature. Post obstructive arterial thrombosis, embolism, aortic valve distortion leading to severe cardiac failure are other causes that have been identified as cause for end organ damage (Williams et al. 1997). Hypertension is seen as a common feature of distal dissection, occurring in almost 70% of cases. On the contrary, only 36% of proximal dissection presents with hypertension. Approximately 20% of Standford A dissection is associated with hypotension secondary to severe aortic insufficiency, cardiac tamponade and myocardial infarction (Klompas 2002). Pseudohypotension is another entity that one should be aware of. Pseudohypotension occurs when there is compromise to branchial artery circulation following proximal dissection (Sheikh et al. 2013).

Echocardiograph is often the initlal investigative tool. Presence of an elevated ST wave or inverted T wave denotes acute myocardial ischemia (Klompas 2002). Widened mediastinum on chest radiograph suggests aortic dissection. Such a finding however warrants a more definitive imaging modality with higher sensitivity and specificity. CTA has sensitivity of 99% and specificity of almost 98% in detecting aortic dissection. Magnetic resonance Imaging (MRI) is more specific than CTA with ability to differentiate between penetrating atherosclerotic ulcers from intramural hematoma. However, the long duration required for image acquisition renders it less favourable (Zeman et al. 1995; Shiga et al. 2006). Negative finding on radiological imaging in the presence of high clinical suspicion of an acute aortic dissection warrants application of a second imaging modality (Svensson et al. 1999). Raised serum urea, creatinine and lactate suggest ongoing visceral hypoperfusion and end organ damage.

Treatment of acute aortic dissection would depend on site of dissection, severity of presentation and concomitant existence of life threatening complications (Hiratzka et al. 2010). Stanford A acute aortic dissections requires urgent surgical evaluation for emergency surgical repair in view of it's high risk of rupture. Open surgery is preferred over endovascular repair in the setting of acute proximal aortic dissection, requiring excision of proximal extent of the dissection and replacement with Dacron graft (Hagan et al. 2000). Stanford B acute aortic dissection on the other hand can be managed conservatively through medical management. Due to a lower pressure of blood flow through the false lumen in distal dissection as compared to proximal aortic dissection, further progression of the dissection often comes to a halt due to thrombosis of the false lumen. Use of beta-blockers and angiotensin converting enzymes has been proven to control hypertension and limit progression of distal aortic dissection. However, progression of dissection, malperfusion syndrome, enlargement of co-existing aortic aneurysm warrants prompt surgical intervention (Estera et al. 2006). Transthoracic endovascular aortic repair (TEVAR) is the preferred surgical

treatment option in complicated Stanford B aortic dissection. Studies show a reduction in mortality and paraplegia rates with TEVAR as compared to conventional open surgery (Leurs et al. 2004; Eggebrecht et al. 2006; Chen et al. 2006).

CONCLUSION

Our patient was diagnosed with acute Stanford B aortic dissection. Decision for surgical intervention was carried out due to the close proximity of the dissection with the origin of the left subclavian artery. Left untreated, it could have led to impaired perfusion of the entire left upper limb. Decision for TEVAR was in accordance to recommended guidelines, however the risk of iatrogenic occlusion of the left subclavian artery outweighed the benefit in that setting and prompted us to proceed with an interposition bypass graft between the left common carotid and left subclavian prior to embarking on endovascular repair. The non-complicated distal aortic dissection in our patient was managed conservatively. The cause of dissection in our patient remains unknown. Acute aortic dissection is a life threatening condition and failure to identify and administer appropriate treatment can be fatal. Thus, it is imperative that surgeons know of this rare clinical entity. The non–specific and broad spectrum of clinical presentation poses a diagnostic challenge. A combination of atypical chest pain, ischemic pattern on ECG, mediastinal widening on chest radiograph and deranged renal profile as well as elevated serum lactate should increase the likelihood of aortic dissection. Decision to intervene surgically or continue with medical management should depend on CTA finding, progression of dissection and evidence of life threatening complications.

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