

## Common Presentation of an Uncommon Disease: A Fatal Acute Aortic Dissection in a Young Male

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### ABSTRAK

*Diseksi aorta jarang berlaku di kalangan kanak-kanak dan pemuda. Kami ingin membentangkan kes Diseksi Aorta Stanford B yang terjadi terhadap seorang pesakit lelaki berusia 30 tahun, yang telah membawa maut. Pesakit ini datang dengan aduan sakit perut akut yang teruk dan didapati bacaan tekanan darahnya tinggi. Walaupun tanpa tanda-tanda tipikal diseksi aorta, namun kadar kesakitannya yang teramat kuat disertai dengan tekanan darah yang amat tinggi menyebabkan kami mengesyaki diseksi aorta dalam kes ini. Abdomen ultrasound ada dilakukan di zon Kecemasan namun tanda khusus diseksi aorta iaitu "intimal flap" tidak dapat dikesan awal kerana bukan semua pegawai perubatan mahir dalam ultrasound. Dalam kes ini, komputer tomografi (CT) Abdomen dilakukan juga untuk membuktikan diagnosis ini. Walaubagaimanapun, keadaan klinikalnya semakin merosot dan pembedahan yang dilakukan tidak berjaya menyelamatkannya, lalu beliau meninggal dunia selepas 36 jam rawatan. Kami menekankan kepentingan untuk mengesyaki penyakit ini pada peringkat awal bersama bantuan kaedah ultrasonografi untuk mengesan ciri-cirinya dengan segera untuk membantu menangani pesakit yang serius ini.*

*Kata Kunci: diseksi aorta, flap intimal, muda, ultrasonografi*

### ABSTRACT

Acute aortic dissection (AAD) is rare in the paediatric and young adult population. We present a fatal case of acute aortic dissection Stanford B in a young male diagnosed with hypertension. He presented with severe acute abdominal pain with malignant hypertension. He did not have any trauma to the chest or did not have history of an illicit drug abuse. He had no features suggestive of connective tissue

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disease as well as other typical signs of aortic dissection. The complain of acute, severe abdominal pain which was out of proportion and required multiple doses of intravenous opioid, raised the suspicion of aortic dissection in this case. Point of care sonography (POC) was done in Emergency Department (ED). However, due to its highly operator dependability, the intimal flap was missed. Computed tomography (CT) scan of abdomen was done and confirmed the diagnosis of AAD. Unfortunately, his clinical condition rapidly deteriorated few hours later with no response to surgical intervention and succumbed within 36 hours of admission. We highlighted the importance of the early recognition of this disease as well as the point of care sonography in ED as a diagnostic tool to tackle this time-sensitive disease.

Keywords: aortic dissection, bedside ultrasound, intimal flap, young

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## INTRODUCTION

Acute aortic dissection (AAD) is a clinical emergency with high morbidity and mortality. AAD commonly occur in males above 60 years. This diagnosis is uncommon in a young adult with absence of atherosclerosis, trauma or connective tissue disease (Kosai et al. 2014). Essential feature of AAD is a split of the intimal layer with formation and propagation of the subintimal haematoma. Most common risk factor contributing to the intimal injury in the AAD include hypertension, rupture of atherosclerotic plaque, connective tissue disease or traumatic. High-pressured blood flow track between the tunica intima and tunica media creating an intima-medial flap which gives rise to a false lumen. Re-entry of blood into the true lumen from the false lumen are also possible due to secondary tears in the tunica intima (Roberts & Roberts et al. 1991).

Excruciating back pain is the most common symptom of AAD, followed

by chest pain, abdominal pain, syncope, hemiparesis and claudication leg pain. Some patients may have painless dissection. Hemopericardium causes pericardial tamponade that may manifest as syncope or even sudden death (Kosai et al. 2014). Non-specific symptom of this disease which simulates other common diseases often leads to a missed diagnosis.

Abdominal pain can be a challenging complaint in patient with AAD because abdominal pain is commonly associated with benign condition. Therefore, high index of clinical suspicion, followed by appropriate action taken is important in managing acute aortic dissection (Nesser et al. 2002). Misdiagnosis can have fatal consequences as untreated patients have an early mortality rate as high as 25% in 24 hours and 75% by 2 weeks (Moore et al. 2002; Lauterbach et al. 2001). AAD associated with a high mortality, affecting 3 to 4/100,000 people/year represents the most common aortic emergency.

Furthermore, patients with AAD have 20% out of hospital mortality and 30% of in hospital death (Karthikesalingam et al. 2010). Point-of-care sonography (POC ultrasound) involves bedside ultrasound assessment performed by the emergency medical officer is an integral part of a physical examination. It is important to have the skills and knowledge to gather information with the use of POC ultrasound to prompt early diagnosis for early surgical consultation thus to reduce mortality.

### CASE REPORT

A 30-year-old male with underlying chronic hypertension on tablet Amlodipine 5mg daily presented to our Emergency Department (ED) at 7.50 pm with complain of severe, acute-onset, sharp burning epigastric pain which radiates to the back, started at 3 pm on the same day. He denies any history of trauma, illicit drug use or usage of traditional medication. The pain had no aggravating cause or relieving factor and increased in intensity throughout the day. He denied having chest pain,

shortness of breath, vomiting, fever or weakness of the limbs.

On arrival, he was conscious and alert, but in severe pain with the pain score of 10/10. His blood pressure was 211/132 mmHg, heart rate of 96 beats/minute and pulse oxymeter of 100% on room air. He was afebrile with temperature of 37°C and not tachypnoeic (respiratory rate of 18/min) on arrival. The radial, dorsalis pedis, and posterior tibialis pulses were full and equal bilaterally. No radio-radial delay or radio-femoral delay. There were no features suggestive of Marfan's Syndrome or any other connective tissue disorder. The abdominal examination revealed tenderness and localised guarding at the epigastric and umbilicus region with no pulsatile mass palpable. No heart murmur was detected on heart auscultation. Lung auscultation was clear.

The electrocardiogram (ECG) suggests of left ventricular hypertrophy. Initial bedside echocardiogram showed hypertrophic ventricles with dilated chambers, aortic root measured 3.0cm with no pericardial



Figure 1: Bedside USG Abdomen in ED shows visualization of an intimal flap (red arrow)

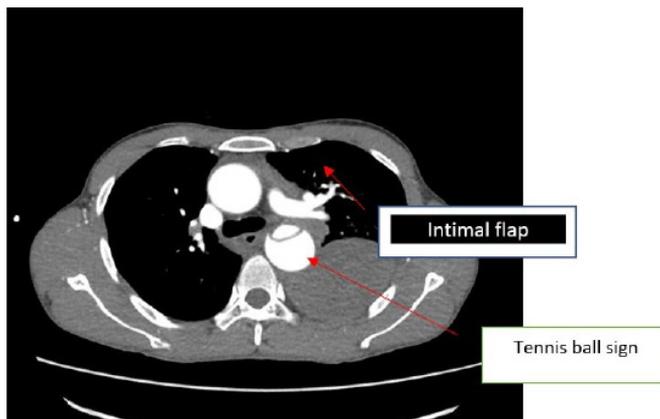


Figure 2: CTA abdomen-axial view shows ‘tennis ball ‘sign

effusion. Bedside ultrasound abdomen in ED showed 2.8cm abdominal aorta diameter. Patient was sent for formal ultrasound abdomen based on high index of suspicion of aortic dissection. Formal ultrasound abdomen showed evidence of intimal flap along the coeliac trunk up to the renal artery (Figure 1). The abdominal aorta was normal measuring 2.8cm with no aortic dilatation. The liver, pancreas and spleen were normal. There were gall bladder polyps, bilateral renal parenchymal disease as well as minimal right pleural effusion.

Chest X-ray anteroposterior view showed widened mediastinum 9.0cm with prominent aortic knuckle and absence of air under diaphragm. Computer Tomographic Angiogram (CTA) abdomen showed extensive aortic dissection (Stanford B) involving proximal and mid descending thoracic aorta, down to proximal coeliac trunk and proximal left common iliac artery, with active leaks at proximal and mid descending thoracic aorta into the left hemithorax with mediastinal shift (Figure 2&3).

On arrival, he was given a total

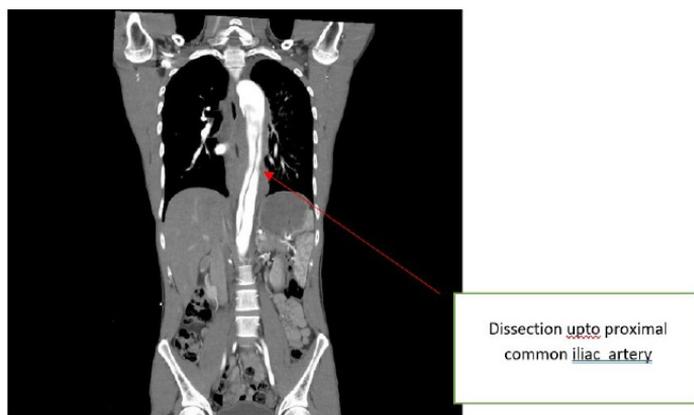


Figure 3 : CT Abdomen coronal view

of 6mg of intravenous Morphine in titrating dose for the pain management. He received an intravenous Labetalol bolus followed by continuous infusion of Labetalol to treat the hypertensive emergency with the aim of SBP between 100-120 mmHg. Six hours post labetalol he developed hypotension with cold peripheries and pallor. He became more tachypnoeic requiring 5L of facemask oxygen. Lung auscultation revealed increasing dullness over the left side of the chest up to the middle zone. The laboratory results showed reduction of haemoglobin from 12.1g/dl to 7.1g/dl thus 2 pints pack cells were transfused. A diagnosis of hypovolemic shock with left haemothorax was made. Other relevant investigations were normal.

Following multidisciplinary evaluation, consensus was reached to treat him aggressively and he was transferred to cardiothoracic team in Institut Jantung Negara (IJN) for surgical intervention due to unavailability of cardiothoracic specialty in this hospital. He was then transferred to intensive care unit (ICU) in IJN. Prior to the transfer, his saturation was 100% under nasal prong 3L/min, haemodynamically stable with MAP<65 (on IVI Labetalol 5mg/hr) and pulse rate 70-80bpm. His urine output was 300cc/hr. He underwent thoracic endovascular aortic repair (TEVAR) transferred to coronary care unit (CCU) in IJN after the procedure. Unfortunately, he succumbed one hour, post-operation.

## DISCUSSION

Acute aortic dissection in younger individuals is very rare. AAD mainly affect the elderly group with the mean onset of age affected by aortic dissection is 56.5 years (Fikar & Koch 2000) and hypertension is the leading risk factor in this group (Landenhed et al. 2015). The disease has been associated with generalized connective tissue disorders in younger patients, such as Marfan syndrome (MS), Ehler-Danlos syndrome (EDS), and the Loeys-Dietz syndrome (LDS) (Aalberts et al. 2008). However, spontaneous abdominal aorta dissection is rare and accounts for less than 2% of all aortic dissections (Roberts & Roberts 1991)

In our case, he had spontaneous abdominal aortic dissection with underlying chronic hypertension as the risk factor. He had no signs suggestive of any connective tissue disorders. Diagnosing abdominal aortic dissection is often challenging and it requires a high index of suspicion due to its various presentations which frequently absence of classic findings.

It is challenging to manage a patient with complaint of abdominal pain as it is frequently associated with a benign disease but it may also mimic other common conditions which are more severe and serious. The frequently reported symptoms in thoraco-abdominal aortic dissections are sudden onset of severe back pain (64%) and/or chest pain (63%) and sudden abdominal pain (43%). Majority of patients with Stanford type A AAD presented with chest pain, while abdominal pain and leg pain occurred more often in those with Stanford type B AAD (Zhao et al. 2017).

The presentation of epigastric pain in our case was acute, severe and out of proportion which requires multiple doses of intravenous morphine and associated with morbid hypertension, hence the clinical suspicion of acute abdominal aortic dissection was made. Usage of bedside ultrasound in emergency medicine in diagnosing AAD were shown to have a sensitivity of 94-99% (Nakashima 2010). It is safe, effective, can be performed in less than 5 minutes but it is highly operator dependant. Rapid identification of AAD via POC ultrasound reduced the mortality by 20-60%. Visualisation of an intimal flap is the most common sonographic finding associated with an abdominal aortic dissection thus it is a hallmark for an aortic dissection (Touati 2003) regardless of the diameter. It has a sensitivity of 67-80% and a specificity of 99-100% (Colla et al. 2017). A combination of a bedside transthoracic and transabdominal ultrasound provides a better and comprehensive bedside evaluation for aortic dissection.

Rapid identification should prompt early surgical consultation before computed tomography (CT) to expedite further surgical management in comparison to CT which took an average of 83 minutes/study. The normal diameter of the abdominal aorta via ultrasound is less than 3.0cm. The size of the aorta normally increases about 1-2 mm/year, therefore the normal range has to be corrected by age and sex. The aortic size is also related to exercise and workload in adult (Raimund & Holger 2006). In young and healthy patients,

weightlifting is recently a recognized risk factor for AAD (Gwan-Nulla 2000). In our case, bedside ultrasound in ED revealed the abdominal aorta diameter of 2.8cm but the intimal flap was not identified due to inexperience, thus causing delay in acquiring CT abdomen and surgical referral.

## CONCLUSION

Acute aortic dissection is more common among the older age group with history of chronic hypertension and often misdiagnosed in young adult without any connective tissue disorders. The non-specific and diffuse clinical presentation of AAD possesses a diagnostic challenge. The emergency doctors should have high index of suspicion for aortic dissection especially if the presentation is severe abdominal pain associated with morbid hypertension. POC ultrasound in ED with pathognomonic finding of intimal flap is important for rapid diagnosis of aortic dissection hence will reduce the mortality by expedite the surgical intervention. Despite advances in diagnostic and therapeutic techniques, morbidity and mortality remains high.

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