

Two Rare Cases with Aortic Dissection: future prospective in imaging

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ABSTRACT

Aortic dissection (AD) is an acute emergency condition of the aorta, which often has a fatal outcome. In the acute setting, management of AD is limited to control of hypertension, early recognition of the dissection, and activation of surgical services. In this article we reported two cases of 42 and 45 y/o male patients with chest pain, no hypertension having AD. They just lifted a roughly heavy (10Kg) object. A multi-detector CT applied to perform CT-angiography. To reduce morbidity and mortality, AD should always be considered in patients who present with severe chest pain of abrupt onset. Also, aortic dissections can occur in patients without atypical signs and symptoms.

KEY WORDS: Aortic dissection, Chest pain, Diagnostic imaging, Computed tomography.

1. INTRODUCTION

Aortic dissection (AD) most common life-threatening disorder is a tear of the aorta (the largest artery of the body) which one of the conditions included in the term “acute aortic syndrome” (Auer, 2000). This tear causes blood to flow between the layers of the aortic wall and dissects the layers apart. Of these, acute dissection is the most common, affecting up to approximately 4 cases in 100000 individuals annually in the world (Baliga, 2014). AD is more common in males than in females, with a male-to-female ratio of 2:1 to 5:1 has been reported in different studies (Nikolic, 2013; Auer, 2000). The condition commonly occurs in people in the sixth and seventh decades of life while in patients with Marfan syndrome present earlier, usually in the third and fourth decades of life (Williams, 2010). The same risk factors for AD include giant cell arteritis, congenital aortic stenosis, Fallot’s tetralogy, familial thoracic aorta hereditary diseases, atherosclerosis, trauma or injury, catheterization, coarctation, Marfan syndrome, Behcet arthritis, cocaine use, pregnancy, weight lifting or Valsalva maneuver and infections affecting the aortic wall (e.g.syphilis) (Clough and Nienaber, 2015). In an AD, blood penetrates the intimal and enters the media layer, will travel through the media and creating a false lumen which is a layer of intimal tissue separating it from the true lumen (Nienaber, 2013). This tissue is known as the intimal flap (Pasta, 2013). While most classic ADs begin at 1 of 3 distinct anatomic locations, including aortic arch, approximately 2.2 cm above aortic root, or distal of left subclavian artery, but the majority of aortic dissections originates with an intimal tear in either the ascending aorta (65%), the aortic arch (10%), or just distal to the ligamentum arteriosum in the descending thoracic aorta (25%) (Patel, 2011). Ascending aortic involvement may result in death from wall rupture, hem pericardium and tamponed, occlusion of the coronary Ostia (opening) with myocardial infarction, or severe aortic insufficiency (Tran and Khoynzhad, 2009). About 96% of individuals with aortic dissection present with severe pain that had a sudden onset. 17% of individuals will feel the pain migrate as the dissection extends down the aorta (Clough and Nienaber, 2015). Anterior chest pain is associated with dissections involving the ascending aorta, while intra-scapular (back) pain is associated with descending aortic dissections (Auer, 2000). While the pain may be confused with the pain of a myocardial infarction (heart attack), aortic dissection is usually not associated with the other signs about myocardial infarction such as heart failure and EKG changes (Auer, 2000; Pasta, 2013).

The uncommon symptoms that may be seen in aortic dissection include congestive heart failure (7%), syncope (9%), cerebrovascular accident (CVA) (3-6%), ischemic peripheral neuropathy, paraplegia and cardiac arrest. Neurologic complications of aortic dissection, such as CVA and paralysis are due to the involvement of one or more arteries supplying portions of the central nervous system. The outcome of the AD is determined by the type and extent of the dissection and the presence of associated complications, cerebral sequel, and aortic branch involvement, pericardial and visceral involvement, with early diagnosis. DeBakey et al. classify ADs into 3 types, as follows. Type I: The intimal tear occurs in the ascending aorta, but the descending aorta is also involved. Type II: Only the ascending aorta is involved. Type III: Only the descending aorta is involved. Type IIIA involves the descending aorta that originates distal to the left subclavian artery and extends as far as the diaphragm. Type IIIB involves the descending aorta below the diaphragm (Figure 1).

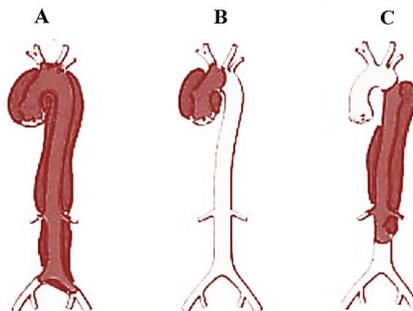


Figure.1. Aorta dissection classification according to DeBakey scale. A) Type I that originate in the ascending aorta, propagated at least to the aortic arch and often beyond it distally. B) Type II that originate in and is confined to the ascending aorta. C) Type III that originate in the descending aorta and extends distally down the aorta or rarely retrograde into the aortic arch and ascending aorta

Type A dissections usually require surgery, whereas type B dissections are managed medically under most conditions. Without surgical treatment, approximately 75% of patients die within 2 weeks of the onset of symptoms. While taking a good history from the individual may be strongly suggestive of an aortic dissection, the diagnosis cannot always be made by history and physical signs alone. Often the diagnosis is made by visualization of the intimal flap on diagnostic imaging methods. ECG and chest radiography are not sensitive enough to diagnose dissection, but will identify concomitant acute coronary syndromes or act as indicators of alternative diagnoses. Transesophageal echocardiography (TEE) is a noninvasive examination that may help detect an ascending aortic dissection flap. It has a reported sensitivity of 59%–83% and a specificity of 63%–93% for the diagnosis of aortic dissection, but have a sensitivity of 94%–100% and a specificity of 77%–100% in identifying an intimal flap. However, the distal part of the ascending aorta and the branches of the aortic arch may not be adequately evaluated with TEE. CT with intravenous (IV) contrast remains the recommended first line investigation in those suspected of having dissection which also useful for planning surgical intervention. Magnetic resonance angiography (MRA) lacks radiation exposure, and uses less nephrotoxic agent, which benefits patients with evidence of renal hypoperfusion which have a sensitivity of 95% to 98% and specificity of 94% to 98% for detecting aortic dissection. However, availability is more limited and imaging takes longer, making it more suitable in the chronic setting or for patient follow-up. Each of these tests has varying pros and cons and they do not have equal sensitivities and specificities in the diagnosis of aortic dissection. The multidetector CT scan with contrast is a fast, non-invasive test that will give an accurate three-dimensional view of the aorta. In order to delineate the aorta to the accuracy necessary to make the proper diagnosis, an iodinated contrast material is injected into a peripheral vein at a properly timed moment, so that it entered the aorta at the time that the aorta is being imaged. It has a sensitivity of 96 - 100% and a specificity of 96 to 100%.

2. CASE PRESENTATION

2.1. Case A: A 42 year old man with a 1-hour history of right-sided chest pain and with no history of hypertension came to the Namazi hospital (Shiraz, Iran), accompanied by his wife; by an ambulance, while he just lifted a heavy object, i.e. a gas cylinder (60 Kg). The patient stated that he experienced an abrupt pain that traveled from his chest to the tip of his scapula as he bent over his desk to reach for something in the wastebasket. The chest pain was relieved by sublingual nitroglycerin, which the patient took before coming to the hospital. The patient has rupture in descending and ascending aorta (DeBakey type I). Three images of the patient's aorta, with multi planner reconstruction, are presented in figure 2. As figure 2, the patient has reduction of right kidney blood perfusion in the arterial phase due to AD and involvement of right renal artery, although left renal artery involvement is more common than the right side. The wall of the false lumen is thin and as we have seen in figure B images, the expanded, false lumen cause compression of the true lumen. As we expected, the concentration of contrast material in the false lumen is lower than in true lumen due to hypoperfusion, and thus the false lumen appear hypo dens in comparison with the true lumen. Intimal flap is clearly seen in ascending and descending aorta.

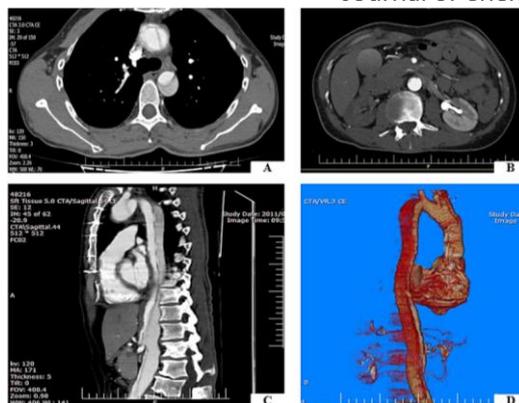


Figure.2.A: Axial view of aortic dissection (Type A aortic dissection). Intimal flap is seen clearly in ascending and descending aorta. Note the low density of the false lumen relative to the true lumen. B: Decrease of right kidney blood perfusion in arterial phase due to aortic dissection. The right renal artery is perfused via false lumen (not shown in this slice). C: Sagittal multi planner reconstruction of aortic dissection. Note that the expanded, false lumen has a pressure effect on the true lumen. D: Volume rendering of aortic dissection (3D MPR). Images A to D show that the flap rotate along the axis of the aorta.

2.2. Case B: A 45 year old man with a 2-hour history of right-sided chest pain and with no history of hypertension came to the Namazi hospital (Shiraz), accompanied by his wife; by ambulance, while he just lifted a heavy object, i.e. a bag of rice (10 kg). The patient stated that he experienced an abrupt pain that traveled from his chest to the tip of his scapula as he bent over his desk to reach for something in the wastebasket. The chest pain was relieved by sublingual nitroglycerin, which the patient took before coming to the ED. The patient has rupture just in descending aorta (DeBakey type III). Three images of the patient's aorta, with multi planner reconstruction, are presented in figure 3. As figure 3 demonstrates, the patient has an intimal flap in his descending aorta. The ascending aorta is slightly dilated, but there is no flap in it. AD has extended to the abdominal aorta. The most proximal part of the dissection is at the level of pulmonary artery bifurcation. Renal perfusion is not impaired in this case and the dissection has been ended at suprarenal region.

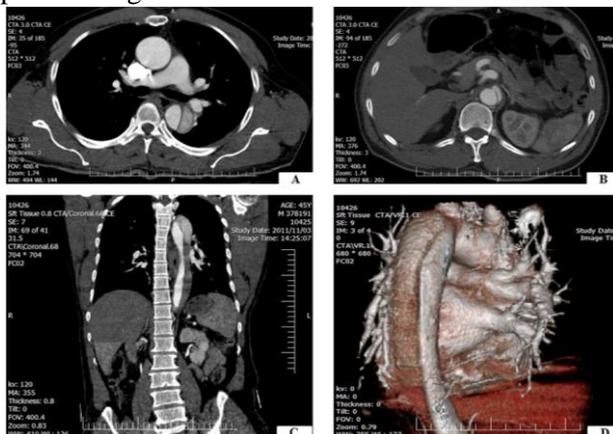


Figure 3. A: The descending aorta is involved (DeBakey type III). Note the slightly dilated ascending aorta without any flap in it, compatible with DeBakey III and Stanford B type. The level of dissection is at the pulmonary artery bifurcation. B: Axial view of aortic dissection in abdominal cavity. Note the false lumen has been extended distally, which is below the diaphragm. C: Coronal view of aortic dissection. Infradiaphragmatic extension of AD is clearly seen. D: Volume rendering of aortic dissection (3D MPR)

2.3. CT with IV contrast: A multi-detector CT-scan (Aquilion, 16 slices; Toshiba Medical Systems, Toshiba zecooperation, Tokyo, Japan) with 1-2.5 mm collimation was applied to perform the CT scan. A typical helical scanning protocol (including 5-mm collimation, 1.5 pitches, and 7.5-mm imaging increments) was used to get the images. Initially non-enhanced CT was performed for diagnosis of acute hemorrhage and aortic rupture. The process was followed by a helical CT performed approximately 25-30 seconds after injection of a nonionic contrast media. The contrast media was 120-135 ml of iodine, 350 mg/mL, iohexol (Omnipaque 350; Nycomed, Princeton, NJ) which was injected using a 20-gauge IV cannula via a peripheral IV site at a rate of 3-4 ml/s applying a power injector pump (CT injector Missouri, Ulrich Medical Systems, Germany). Adding 5 to 10 seconds to the contrast injection

duration is necessary to compensate for the difference in position between the tracking position in the ascending aorta and the top of the chest. A small field of view can be selected to optimize spatial resolution. However, a full field of view must be reconstructed to detect incidental findings. Initial examination of the thoracic aorta should include the abdominal aorta and iliac arteries, because the thoracic aortic pathology commonly involves these vessels.

3. DISCUSSION

Our cases have no risk factors except for gender and they were not in usual age range for AD. It shows that the diagnosis of AD should be considered in a wider range of age and in peoples with have no predisposing factor. Also, these cases show that relatively minor physical stress like lifting heavy objects could be cause AD.

Ninety percent of patients present with sudden onset pain in the chest. In type A dissection, it may radiate to the neck, and in type B to the inter-scapular area. Diabetes is thought to account for the remaining, asymptomatic dissections. New aortic regurgitation is picked up in 31% of patients, and a radio-radio/radio-femoral delay in 15%. Type A presents with hypotension in up to 25% of patients, whereas type B dissections tend to present with hypertension. If both true and false Lumens is perfused the aortic branches, and therefore end organs, will remain perfused. If this is not the case, dissection can present with neurological symptoms such as stroke, renal failure, bowel ischemia or limb ischemia. These are considered high-risk features, and their effect on management is discussed below. On occasion, an asymptomatic dissection can lead to aortic dilatation and rupture, either acutely, or up to three years after the initial event.

Chest pain is a vague complaint that encompasses numerous conditions, including myocardial infarction, pulmonary embolism, pericarditis, pneumothorax, pneumonia, pleurisy, acute pancreatitis, and penetrating duodenal ulcer. Also the main symptoms of AD are uncertain, the reported annual incidence is 5 to 30 cases per million populations (Gaul, 2007).

Several theories have been proposed to explain the pathophysiologic mechanism of AD, and their common feature is the creation and propagation of an intimal flap. The propagation continues until a rupture occurs either into the lumen or through the adventitia. According to one of the theories, this may be due to cystic medial necrosis (Clough and Nienaber, 2015). Degeneration of the media of the aorta weakens the cohesion inside the aortic layers, allowing the creation of an intimal flap. Cystic medial degeneration generally is more extensive in older people and in those with hypertension or Marfan syndrome. Next theory states that intimal flaps tend to form in areas of the aorta subjected to great pressure changes and flexion stress. These areas include the ascending aorta and the first part of the descending aorta. Another theory proposes that penetrating atherosclerotic aortic ulcers weaken the aortic wall and predispose to aortic vascular pathology. This tends to occur in the descending thoracic aorta in elderly people (Gaul, 2007). The pain is differentiated from the pain of myocardial infarction by the absence of radiation into the neck, shoulder, or arm unless there is associated coronary artery extension. Furthermore, more than one-third of the patients may have symptoms due to organ system involvement such as aortic regurgitation, stroke, pleural effusion, and acute gastrointestinal hemorrhage. Our patient had abrupt onset of chest pain, but he did not have any signs of organ system involvement.

The diagnosis of AD requires a high degree of clinical suspicion, since the symptoms and clues can be subtle. A missed diagnosis can be disastrous. The diagnostic work-up should begin with an ECG. However, the work-up must not stop with this test because the presence of a myocardial infarction does not rule out dissection. Multiple imaging modalities can be used if AD is suspected. These include CT, MRI, and TEE, which have the highest specificity, followed by ECG and chest radiography, which are less specific. If a dissection is found, it is important to differentiate a proximal dissection from a distal one for purposes of treatment.

Treatment depends on the status of the patient, the degree of dissection, and the progression status of the dissection. In the emergency department, the mainstay of treatment is control of hypertension. Currently, certain treatment options are available for lowering blood pressure. A β -blocker and a vasodilator are initially administered (Gaul, 2007). Stable patients, or patients who have a distal dissection, or for whom surgery is contraindicated may be managed medically. The goal is to limit the extension of the dissection, to stabilize the dissection, and to prevent complications (Pasta, 2013). If signs and symptoms of extension or worsening of the clinical condition becomes evident, surgery should be considered. Proximal dissections should be treated surgically. Numerous techniques are used, depending on the expertise of the surgeon and the circumstances of the patient. Recently, endovascular stenting has been gaining popularity and is being used to manage distal dissections, especially when symptoms of organ or lower extremity malperfusion are developed.

Previous studies didn't mention that lifting a heavy object, e.g. a gas cylinder or a bag of rice, causes AD. According to the records of the cases reported here, both of the patients have no history of hypertension and other factors that causing AD; like aortic aneurysm, cocaine use, pregnancy, Turner syndrome, Noonan syndrome, Marfan syndrome, and Ehlers-Danlos syndrome. So, what caused the rupture of the aorta? Does lifting heavy object cause AD? After referring these patients to the emergency ward, the physician examined them and ordered a CT with IV contrast from them. The CT helped that the case A has been diagnosed as a DeBakey type I aorta dissection and case

B has been found as a DeBakey type III. In conclusion aortic dissection is a potentially disastrous condition that can be missed when a patient presents with a chief complaint of chest pain. This diagnosis should be considered for patients with hypertension and other cardiovascular risk factors that are present along with chest pains. An incorrect or missed diagnosis can result in rupture and a likely fatal outcome. Based on the reported cases, one should consider lifting a heavy object as one of the causes of aortic dissection. It is clear that dissection carries significant risk of disease progression despite optimal treatment and irrespective of etiology. In those with hereditary aortic wall structure defects, mortality from rupture in an aorta measuring greater than 6 cm is 12%, with women at higher risk than men. Therefore, lifelong surveillance is mandatory, with axial imaging in the very least being used for routine imaging. Imaging at 1, 3, 6 and 12 months, followed by annual review is recommended by the European Society of Cardiology. However, with the publication of results from multi-center randomized controlled trials now becoming available, we see the potential advantages in early use of endovascular repair on both short and longer-term mortality, progressive aortic dilatation and aortic remodeling. Throughout all of this, the message persists; aortic dissection remains a disease with a high mortality and the need for lifelong follow up. CT scans of the aorta are feasible and widely available, which are important features in making an accurate diagnosis quickly in unstable patients. Multidetector CT allows imaging of the entire aorta with rapid acquisition and data reconstruction to provide prompt and accurate diagnosis and to help identify relevant complications that may have an impact on surgical planning or management.

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