Picking the Hard Right Not the Easy Wrong: Acute Aortic Dissection with Catastrophic Outcome

Eussera El-Magbri¹, Awad Magbri²*, Michael G Abesamis³, Kaveh Ilkhanipour¹, Kevin Leonard¹ and David E Zimmerman⁴

¹Department of Emergency Medicine, University of Pittsburgh Medical Center, USA
²Regency Surgery and Vascular Center, University of Pittsburgh Medical Center, USA
³Department of Toxicology, University of Pittsburgh Medical Center, USA
⁴Department of Emergency Medicine, Duquesne University School of Pharmacy, USA

Abstract

The authors discuss a case of aortic dissection in a young man with hypertension. The delay in considering the diagnosis and ordering the proper investigations secondary to the unusual presentation of the case complicated by the logistics of patient’s transfer to specialized Cardiothoracic unit where surgery can be performed safely have impacted the unfortunate outcome.

Keywords: Dissecting aortic aneurysm; Computer tomographic angiography; Dissecting flap; Continuous-veno-venous hemo filtration; Hypertension

Case Presentation

A 29-year-old African American obese male with no self-reported past medical history presenting to the emergency department at UPMC Mercy Hospital, Pittsburgh with 3-days of bilateral flank pain and dark coca cola colored urine. Patient stated the pain is moderately severe, dull-aching and constant. Patient stated that he has not taken any medication for pain relief. He denies any history of similar symptoms, kidney stones, fevers, chills, nausea, vomiting, abdominal pain, diarrhea or constipation. He also denies any chest pain or shortness of breath at presentation. The patient did not recall any trauma or injury to the area, or any numbness or tingling of the lower extremities, or any generalized weakness. Patient does report that he’s been having burning sensation on urination, change in the urine color with, small amount of urine, increased frequency and urgency of urination since back pain started.

His laboratory work-up reveals high blood urea nitrogen and creatinine (24/2.16 mg/L), respectively, and the rest of the chemistry-panel were normal as was his complete blood picture. His urine analysis revealed the presence of 300 mg of protein and moderate amount of Red Blood Cells (RBC’s) but no casts and no evidence of infection. The Electro-Cardiogram (ECG) showed prolonged atrio-ventricular conduction, sinus rhythm, widespread ST and T wave inversion denoting widespread ischemia, more marked in the anterolateral wall. And prolonged QT interval (Figure 1).

His Computerized Tomographic Angiography (CTA) showed acute type-A aortic dissection with proximal extension involving the aortic root. The dissection flap continues into the great.
vessels of the arch of the aorta. The distal flap portion terminates in the right carotid artery. The left common carotid artery arises from the true lumen. Dissection flap involves the descending thoracic and abdominal aorta, celiac trunk, Superior Mesenteric Artery (SMA), and right renal artery. The Inferior Mesenteric Artery (IMA), and the left renal artery arise from the false lumen. There is fusiform enlargement of the ascending thoracic aorta (Figures 2-5).

His post-operative course was complicated by hypoxic respiratory failure requiring nitric oxide and maximal ventilator support. He developed pseudomonas pneumonia and post-operative acute renal failure requiring Continuous Veno-Venous Hemo filtration (CVVH). His blood pressure was labile and required frequent titration of multiple antihypertensive agents. On 7/19 and 7/20 his heart rhythm evolved into unstable Supra-Ventricular Tachycardia (SVT) and atrial fibrillation with Rapid Ventricular Rate (RVR) requiring numerous cardio versions attempts as well as amiodarone, diltiazem, and esmolol. Numerous attempts were made to contact the family at that time to inform them about the health status of the patient were without avail. On the morning of 07/21, the patient became severely hypoxic and arrested. Resuscitation was unsuccessful to revive him, and eventually he passed away.

Case Presentation

Aortic Dissection (AD) is relatively uncommon, (2.6 to 3.5 per 100,000 person-years) [1-4], but often presented acutely with catastrophic illness with sudden onset “tearing or ripping pain” originating in the chest and radiating to the back. Unlike acute coronary syndrome pain, the pain of AD is maximal at the onset and is not gradual in nature. Early and accurate diagnosis and treatment are crucial for survival. Other presentation of AD is organ hypoperfusion due to occlusion of arteries by the dissecting flap (e.g. coronary ischemia, stroke, intestinal ischemia, renal failure, limb ischemia).

Aortic dissection is classified; I (as proximal (type-A) aortic
dissection confined to the ascending aorta and the arch, which can present with acute inferior or right-sided Myocardial Infarction (MI) due to involvement of the right coronary artery, II) and distal AD (type-B) which involves the rest of the thoracic aorta and the descending abdominal aorta. Type-B AD is usually managed medically with aggressive blood pressure control.

Patients with AD tend to be 60 to 80-years-old-men [5-10]. In a review of 4428 patients from the International Registry of Acute Aortic Dissection (IRAD), 66% were men with mean age of 63-years [6]. Women with AD are generally older (67-years) and have a more delayed presentation [6,11]. Younger patients are often having history of hypertension, connective tissue disease, or anatomical abnormalities (Marfan syndrome, Ehlers-Danlos syndrome, Turner's syndrome with coarctation of the aorta with bicuspid aortic valves) [12-15]. Inflammatory diseases that cause vasculitis, trauma, pregnancy and delivery, and the use of Fluoroquinolones have been implicated as risk factors and causes for AD [14-21].

The three major predictors of AD diagnosis are sudden, tearing chest pain; differential pulses and blood pressures between the right and left arms; and abnormal aortic or mediastinal contour on chest-X-ray. Trans Esophageal Echocardiography (TEE) is fast and the most portable method for diagnosis of the unstable patients [22-24]. The sensitivity of the TEE has been reported to be as high as 98%, and the specificity ranges from 63% to 96% [25,26]. However, the gold-standard method for diagnosing AD is Computed Tomographic Angiography (CTA) or Magnetic Resonance Angiography (MRA) [27-30]. The sensitivity of CTA ranges from 83%-95% and the specificity 87% to 100% for the diagnosis of AD [24,31]. The diagnosis of AD by CTA requires the identification of two distinct lumens; the intimal flap may or may not be demonstrated [31]. The MRA is an alternative to CTA but is not widely available. The sensitivity and specificity of MRA is in the range of (95% to 100%) [32].

D-dimer reflects activation of the extrinsic pathway of the coagulation cascade by tissue factor exposed in the aortic media by the intimal tear. D-dimer has emerged as a potential serum marker for AD [33]. However, D-dimer is non-specific and can be elevated in many conditions. Though D-dimers appears to be a useful screening tool to identify patients who do not have AD. A cutoff value of 500 ng/ml, a level below this has a high negative predictive value for excluding AD [33]. In a recent systematic review, a D-dimer value of <500 ng/ml have a sensitivity and specificity of (97% and 56%, respectively, and a negative predictive value of 96%) [34]. This study and others have concluded that in patients with D-dimers of <500 ng/ml, aortic imaging is not going to benefit them [33-40]. A low D-dimer value with low probability of AD may be more useful for ruling out rather ruling in the diagnosis of AD.

Death from AD can be related to cardiac tamponade, acute aortic insufficiency, and acute coronary syndrome with myocardial infarction due to involvement of the right coronary artery in cases of proximal aortic dissection [41,42]. Mortality related to AD was high, up to 30%, however, advances in cardiac surgical techniques have lowered mortality related to proximal AD to 20% [5,6].

A significant contributing factor in the high mortality in AD is delayed time to diagnosis and transfer to cardiothoracic unit capable of performing surgery on AD. So, picking the right diagnosis does not only increase survival but can also save lives and keeps the river of life flowing.

The pathophysiology of AD can be due to iatrogenic, spontaneous (genetically mediated), or traumatic mechanisms [43]. These mechanisms can induce tear in the intima of the aorta. On the other hand, degenerative process involving the media, like cystic medial necrosis can also induce tear in the intima-media layers and causes AD. Blood passes through the tear into the aortic media separating the intima from the media and adventitia and creating a false lumen [7]. Fifty to 65% of tears originated in the ascending aorta and extends to remaining portion of thoraco-abdominal aorta [3]. Approximately, 20% to 30% of intimal tear originates in the vicinity of the left subclavian artery and extends into the descending thoracic and thoraco-abdominal aorta [5].

Adequate blood pressure control in the management of type-A AD is paramount. Treatment of hypertension to systolic of 110 to 120 mmHg using intravenous (IV) β-blockers (esmolol, labetalol) and then IV nitroprusside are indicated. Avoidance of anticoagulation and management of risk factors of AD cannot be overemphasized. Surgery is indicated for complications of dissection, end-organ damage, and contained rupture, penetrating ulceration of significant size or extension despite medical therapy.

In conclusion missing the diagnosis of AD or delay in the optimal treatment and management of these unfortunate patients are often escorted with catastrophic outcome threatening the lives of these patients. Losing these patients because of our shortcomings in diagnosis and management can be devastating to the medical community and the society alike. So, listening carefully to the patient’s history, performing a detailed physical examination, considering the odd causes of patient’s symptoms along with ordering the proper investigations, and evacuating patients in a judicious manner might save lives and keeps the river of life flowing.

References


