ACUTE AORTIC SYNDROME

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Case 1

An 83-year-old lady presented with mild, non-radiating, central chest pain. Her physical examination was unremarkable, and her pulses were symmetrical and bilaterally palpable. Her electrocardiogram (ECG) revealed normal sinus rhythm with no acute ST/T wave changes.

Her transthoracic echocardiogram showed good left ventricular systolic function, with mild dilatation of the ascending aorta (4.5 cm) (Figure 1a), without evidence of pericardial effusion.

Two hours later, she developed severe chest pain and collapsed. She was cold, clammy, and her blood pressure dropped to 60/30 mmHg. Her jugular venous pressure (JVP) was elevated. An urgent bedside transthoracic echocardiogram revealed cardiac tamponade. (Figure 1b & 1c) Urgent computed tomograph (CT) demonstrated a penetrating atheromatous ulcer (Figure 1d) with a large intramural hematoma.
Background

Acute Aortic Syndrome (AAS) refers to the acute presentation of closely related life-threatening entities having similar clinical features and challenges, including the aortic dissection (AD), intramural hematoma (IMH) and penetrating atheromatous ulcer (PAU) (Figure 2).

It has been suggested that aortic dissection, intramural hematoma and penetrating atheromatous ulcer are variations on a continuum of aortic disease (Figure 3). Coady et al² suggested that in patients suspected of aortic dissection, 5 – 17% actually have either intramural hematoma or penetrating atheromatous ulcer. The International Registry for Acute Aortic Dissection (IRAD), which reviewed 464 patients, revealed that two-third of patients were male, with a mean age of 63 years.³ Women were found to be affected less often and their mean age was 67 years.⁴

The classic aortic dissection is initiated by an intimal tear, with resultant propagation within the middle-third of the medial layer of the aorta. Intramural hematoma originates due to a rupture of vasa vasorum or hemorrhage within an atherosclerotic plaque, followed by aortic wall infarction, which in turn, weakens and may rupture, sometimes resulting in aortic dissection.⁷ Penetrating atheromatous ulcer is thought to be caused by rupture of atheromatous plaque through the internal elastic lamina, with subsequent localized medial disruption and potential dissection, pseudoaneurysm formation or free rupture.

In the IRAD registry, hypertension was the most common predisposing factor (72%) for aortic dissection, followed by atherosclerosis (31%), history of cardiac surgery (18%), Marfan’s syndrome (5%) and iatrogenic causes (4%).³

Clinical manifestations

A high index of suspicion is required to establish the diagnosis of this life-threatening entity promptly, since the symptoms can be variable and may mimic those of more common conditions. The pain caused by intramural hematoma and penetrating atheromatous ulcer is similar to aortic dissection.

Other clinical features at initial evaluation, occurring with or without associated chest pain, may include congestive cardiac failure, syncope, acute stroke, acute myocardial infarction, ischemic peripheral neuropathy, paraplegia, and cardiac arrest or sudden death.⁸

Classification systems

The most commonly used classification schemes for AAS are the De Bakey and the Stanford systems (Figure 4). For classification purposes, the ascending
aorta refers to the part of the aorta proximal to the brachiocephalic artery and the descending aorta refers to the aorta distal to the left subclavian artery.

Acute dissection is defined as occurring within 2 weeks from the onset of initial symptoms to the time of first presentation, subacute between 2 and 8 weeks from the onset of symptoms and chronic more than 8 weeks from the onset of symptoms.

**Diagnostic workup**

In the setting of suspected aortic disease, the diagnostic work-up may include ECG, CXR, myocardial markers, transthoracic echocardiogram, transesophageal echocardiogram, CT and magnetic resonance imaging (MRI).

**Electrocardiography**

The electrocardiographic findings in patients with aortic dissection are non-specific. Three-quarters of ECGs with dissection are normal or demonstrate non-specific ST segment or T-wave changes and 25% have left ventricular hypertrophy.³

**Chest X-ray**

A Chest X-ray may show widening of the aortic contour; other features may include displaced calcification, aortic kinking or opacification of the aorto-pulmonary window.⁹

**Biomarkers**

Currently, there are no reliable biomarkers diagnostic of aortic dissection, although a number of markers are under investigation. Release of smooth muscle proteins, soluble elastin fragments, and myosin heavy chain and creatine kinase BB isoforms have been reported after aortic dissection.¹⁰

D-dimers, when elevated above 500 µg/L, appear to correlate with the extent and severity of acute aortic dissection, but fail to distinguish AAS from pulmonary embolism (PE). Elevated D-dimers in the clinical context of suspected aortic dissection should prompt an urgent CT or transesophageal echocardiography for detection of either life-threatening entity.¹⁰
Diagnostic Imaging

In the acute setting, the primary goals of diagnostic imaging in patients with suspected AAS are confirmation of clinical suspicion, classification of dissection, localization of tear, assessment of the extent of dissection and identification of signs indicating the need for an emergency intervention (pericardial, mediastinal of pleural hemorrhage).

Computed Tomography

Contrast-enhanced CT scanning has become the most commonly used modality in evaluating aortic dissection. With the advent of spiral CT, studies may be performed more rapidly, with less patient discomfort, greater accuracy, and lower iodine load. Spiral CT has been proposed as the diagnostic test of choice, as it has a sensitivity of 94% and specificity of 87% in diagnosing aortic dissection.11

Echocardiography

For stable patients, any of the imaging modalities may be useful depending upon the local expertise. Transthoracic echocardiography is less sensitive (59% - 83%) and less specific (63% - 93%) for the diagnosis of aortic dissection than other modalities.

In hemodynamically unstable patients, transesophageal echocardiography may be the first diagnostic modality but may not be able to provide any details of the abdominal segments. Willens et al12 reported a high accuracy of transesophageal echocardiography for diagnosing aortic disease with a sensitivity of 97 – 100% and a specificity of 77 – 100%.

Magnetic Resonance Imaging

MRI is a high precision diagnostic modality for the diagnosis of aortic dissection. The sensitivity and specificity of MRI for the diagnosis of aortic dissection has been reported between 95 and 100%.13

Aortography

Aortography is now rarely used for the diagnosis of acute aortic dissection. Comparative studies found the sensitivity of aortography to be 88% and the specificity 94%.14
**Intravascular Ultrasound**

Intravascular ultrasonography is a promising tool for accurate determination of the location and extent of dissection and assessment of branch vessels. It may be particularly helpful to differentiate aortic dissection from penetrating atheromatous ulcer, but it has not been widely employed.

**Coronary Angiography**

Routine coronary angiography is not recommended before surgery for type A aortic dissection because of the concern about delay in emergency surgery.\(^\text{15}\)

**Management Issues**

The therapy for aortic dissection aims to halt progression of the dissecting hematoma. **Lethal complications** arise not from the intimal tear itself but rather from the subsequent course taken by the dissecting aorta, such as vascular compromise or aortic rupture.

**Initial Medical Management**

The initial management of patients with AAS involves adequate pain relief and aggressive control of blood pressure. This limits the propagation of dissected wall components and reduce dP/dt. Reduction in pulse pressure with a target systolic pressure of 100–120 mm Hg and a heart rate of 60-80 bpm, to maintain adequate cerebral, coronary and renal perfusion, is a priority. Use intravenous beta–blockade as first line of therapy.

Vasodilators may be used to control the blood pressure, but they should never be used as an initial form of therapy before commencing β – blockers because of the reflex tachycardia and increase in the force of left ventricular ejection leading to increased aortic wall stress. Opiate analgesia should be used, as it helps attenuate the release of catecholamines in response to pain, with resultant tachycardia and hypertension.

**Type A Aortic Dissection**

The main aim behind any surgical intervention in type A aortic dissection is prevention of life-threatening complications including aortic rupture and pericardial effusion, which may lead to cardiac tamponade. Acute type A dissection has a mortality of 1–2% per hour during the first 24–48 hours of the onset of symptoms.
Without surgical treatment, the mortality rate is 20% by 24 hours, 30% by 48 hours, 40% at one week and 50% at one month.\textsuperscript{3} The operative mortality for ascending aortic dissections at experienced centers varies widely, between 10 and 35% but is less than the 50% mortality with medical therapy.\textsuperscript{16}

**Uncomplicated Type B Aortic Dissection**

Medical management remains the mainstay of treatment for patients with uncomplicated type B disease. In 384 patients with type B dissection, 73% were treated medically with 10% in-hospital mortality. Long-term survival was 80% at 5 years.\textsuperscript{3}

**Complicated Type B Aortic Dissection**

Complicated type B aortic disease is differentiated from uncomplicated by the presence of a distal malperfusion syndrome or rapid disease progression. Indications for emergency intervention include organ or limb ischemia, aneurysm expansion and risk of rupture, peri-aortic blood collection, intractable pain or uncontrolled hypertension.

**Intramural Hematoma**

Surgery is advocated in patients with type A intramural hematoma, and a trial of medical therapy is advocated in patients with type B intramural hematoma, similar to type A and B aortic dissection. Indications for surgical intervention on type B intramural hematoma include recurring, refractory chest pain and increasing extent or diameter of the hematoma.

Complete resolution of type B intramural hematoma has been documented in 50 – 80% of patients.\textsuperscript{17} However, these patients can also progress to frank dissection or late aneurysm.\textsuperscript{18} Resolution is more likely in young patients,\textsuperscript{17} aortic diameter <4.0 – 4.5 cm,\textsuperscript{7} thickness of the hematoma <1 cm\textsuperscript{7} and beta-blocker usage.\textsuperscript{18}

**Penetrating Atheromatous Ulcer**

Penetrating atheromatous ulcer involving the ascending aorta is uncommon; however, the ulcer usually ruptures and is commonly lethal. Patients with penetrating atheromatous ulcer involving the descending aorta can initially be treated
conservatively with aggressive medical therapy and with close observation, similar to descending aortic dissection.

Like intramural hematoma, penetrating atheromatous ulcer is more frequent in the descending thoracic aorta. Stanson et al\(^6\) advocate an aggressive surgical approach. These ulcers often arise in the mid-descending thoracic aorta and, therefore, any surgical resection should include this portion of the aorta. Hussain and associates\(^19\) challenge this approach and present evidence that non-operative management was successful in many cases. Our algorithm for the management of AAS is shown in Figure 5.

**Prognosis**

Prompt diagnosis and appropriate and timely management reduce the frequency of fatal outcomes.

Coady et al\(^2\) demonstrated that the rate of aortic rupture to be much higher for intramural hematoma (35%) and penetrating atheromatous ulcer (42%) compared to aortic dissection (type A 7.5%, type B 4.1%).

**Long-term Follow-up**

All patients with known aortic disease require close surveillance after discharge and annually thereafter. The 10-year actuarial survival rate in this group of patients has ranged between 30 and 60%.\(^{16,20}\)

Lifelong treatment of hypertension is required and regular assessments of the aorta should be performed at 1,3,6,9 and 12 months as well as every 6-12 months thereafter, depending on the aortic size.

After medically or surgically treated acute aortic dissection, MRI appears to be the imaging modality of choice. The most important findings on imaging are aortic diameter, signs of aneurysm formation, and hemorrhage at surgical anastomosis or stent-graft sites.

**Conclusions**

Acute aortic syndrome should be considered in all patients presenting with chest pain. The etiologies include aortic dissection with a false lumen and intimal flap, intramural hematoma and penetrating atheromatous ulcer. The surgical and percutaneous strategies to treat AAS continue to improve and evolve. These disorders can be assessed by MRI, CT or transesophageal echocardiography, with
high diagnostic accuracy. There is a higher risk of rupture with intramural hematoma and penetrating atheromatous ulcer, especially when located in the ascending aorta. Currently, the recommendations are to treat type A lesions surgically, while type B lesions are initially managed medically. All patients with AAS require close surveillance after discharge.

Case Resolution

She was immediately transferred to a Cardiothoracic Surgical Center where she underwent emergency surgery. She had cardiac tamponade with significant bloody pericardial effusion and an extensive intramural hematoma of the ascending aorta. The ascending aorta was replaced with a Vacutek® Vascular graft, reinforced with bovine pericardium.
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Conflict of Interest Disclosures

None

Authors’ contributions

All the authors have read and approved the manuscript, and have contributed significantly. KA wrote the case report. ASS did the literature search and wrote the initial and the final manuscript. SM thoroughly reviewed the final draft. SM, ASS and KA reviewed and approved the final manuscript.
References


**Figure Legends**

**Figure 1a:** Transthoracic echocardiogram (parasternal long axis-view) showing mildly dilated proximal aorta at 4.5 cm (Ao: Proximal Aorta, LA: Left atrium, LV: Left ventricle, RV: Right ventricle).

**Figure 1b:** Transthoracic echocardiogram (parasternal short axis view at aortic valve level) showing large pericardial effusion with collapse of right atrium in diastole (thick arrow) suggesting cardiac tamponade. (PEff: Pericardial effusion, RV: Right ventricle, RA: Right atrium, AoV: Aortic valve).

**Figure 1c:** Transthoracic echocardiogram (subcostal view) showing a large pericardial effusion (thick arrow).

**Figure 1d:** Axial contrast-enhanced CT scan shows penetrating atheromatous ulcer of the aorta (thick arrow) filling with contrast material.

**Figure 2:** Classification of Acute Aortic Syndrome (Reprinted with permission from ref. 1)

A: Aortic Dissection  
B: Intramural Hematoma  
C: Penetrating Atheromatous Ulcer

**Figure 3:** Acute Aortic Syndrome; progression of one type to another is shown by arrows.

**Figure 4:** Classification of Aortic Disease.

**Figure 5:** The algorithm for the management of AAS.

**Key to Figure 5**

AD: Aortic Dissection  
IMH: Intramural Hematoma  
PAU: Penetrating atheromatous ulcer  
TEE: Transesophageal echocardiogram  
CT: Computed tomography  
MRI: Magnetic Resonance Imaging  
IV: Intravenous