Imaging findings of acute aortic syndromes

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Learning objectives

1. To review the pathophysiology and clinical significance of the acute aortic syndromes (AAS).

2. To illustrate the imaging findings and differentiating features of each AAS on ECG-gated multi-detector CT (MDCT).

3. To compare ECG-gated MDCT with other modalities (CXR, TEE, Aortography, MRI).

Background

Introduction

Acute Aortic Syndromes (AAS) comprises of four clinical entities: Aortic Dissection (AD), Intramural Hematoma (IMH), Penetrating Atherosclerotic Ulcer (PAU), and unstable thoracic aneurysm. Due to the fact that the highest mortality of AAS occurs during the first 48 hours after onset of symptoms, a good prognosis depends on prompt diagnosis and early intervention. However, several studies have shown that a delay in diagnosis of more than 24 hours after presentation occurs in up to 39% of patients with AD. This is mainly due to significant overlap of clinical symptoms between AAS, acute coronary syndromes, and pulmonary embolism, and the relative rarity of AAS compared to these other two emergencies.

The clinical presentation of AAS is often very dramatic with the patient in critical condition. The acute event is characterized by hyper- or hypotension, loss of pulses, diaphoresis, dyspnea, and weakness. The location of pain correlates well with the site of the involvement. Anterior chest pain tends to occur with involvement of the ascending thoracic aorta and arch of aorta, whereas back pain and interscapular pain occurs with involvement of the descending thoracic aorta. Syncope often suggests the development of dangerous complications such as cardiac tamponade, obstruction of cerebral vessels, and activation of cerebral vasoreceptors. Some of the most serious clinical presentations of AAS include pulmonary edema, neurological findings due to carotid artery obstruction (hemiplegia, hemianesthesia) or spinal cord ischemia (paraplegia), bowel or myocardial ischemia, hematuria, compression of adjacent structures (SVC syndrome, hoarseness, dysphagia, and dyspnea secondary to airway obstruction).

Aortic Dissection
Aortic dissection (AD) occurs when there is a tear in the intimal layer leading to blood accumulating in the medial layer of the aortic wall. This gives rise to a true lumen and a newly created false lumen, which is separated by an intimomedial flap. The more common term "intimal flap" is actually a misnomer, because the flap is actually composed of the entire intima and the inner two-thirds of the media, while the outer wall of the false lumen is composed of the outer one-third of media and adventitia. This outer wall is consequently very thin and prone to rupture. The most common sites of entry tear are the right lateral wall of the ascending aorta and the descending aorta just distal to the left subclavian artery, where the shearing stress against the aortic wall is greatest.

Risk factors for AD include hypertension, aortic disease (bicuspid aortic valve, coarctation, aneurysm), connective tissue diseases (Marfan, Ehler-Danlos), direct trauma, cocaine abuse, and pregnancy. The typical aortic dissection presents with sudden onset of severe chest or back pain with a tearing or ripping quality in an older patient who is hypertensive. However, recently it has been found that the incidence of tearing or ripping pain (51%) is actually less frequent than the more common sharp pain (64%) experienced by AD patients. The location of the pain is related to the site of AD. Patients with ascending or aortic arch AD are more likely to have anterior chest pain, while those with descending AD are more likely to have back or abdominal pain. The AD can migrate anterogradely or retrogradely from its initial tear site, which can cause a migrating nature to the pain. Focal neurological deficits, pulse deficits or pressure differences between extremities may also be found. A new diastolic murmur caused by acute aortic regurgitation can also be a severe consequence of AD. On chest radiograph, a widening of the mediastinum should make the clinician suspicious of AD in the right clinical context.

When AD is identified, it is important to classify it based on the Stanford system. Type A dissections involve the ascending aorta and/or aortic arch, with or without involvement of the descending aorta. Type B dissections involve the descending beginning distal to the left subclavian artery. Type A dissections should be surgically repaired immediately to avoid fatal complications such as hemopericardium, hemothorax, MI, or acute AR. Type A dissections can also cause stroke, visceral ischemia, or circulatory failure. Without surgery, 20% of patients with type A dissections die within 24 hours, and 50% within 1 month. Type B dissections are usually treated conservatively with the goal of controlling hypertension unless there is end-organ ischemia, persistent pain, or aneurysmal dilatation.

**Unstable Thoracic Aneurysm**
An aortic aneurysm is defined as a permanent dilatation of at least 150% of normal size, which corresponds to larger than 5 cm in thoracic aorta and larger than 3 cm in abdominal aorta. True aneurysms contain all layers of the aortic wall while false aneurysms are contained ruptures and consists of adventitia and surrounding fibrosis. An unstable thoracic aneurysms is defined as when such an aneurysm is enlarging rapidly (> 1 cm/year), show signs of imminent rupture (will discuss more in the imaging findings section), or have already ruptured (but usually contained if patient survives to be imaged). Thoracic aortic aneurysms often are asymptomatic, but may be associated with vague chest pain resulting from compression on adjacent structures. In contrast, unstable thoracic aneurysms is characterized by severe chest pain. As the diameter of thoracic aortic aneurysms increases, the risk of rupture also increases based on Laplace’s Law. Patients are typically treated when a dilatation in the ascending aorta and descending aorta reaches 5.5 and 6.5 cm, respectively. If patients have connective tissue disorders like Marfan’s, they should undergo treatment at lower thresholds.

**Intramural Hematoma**

Intramural Hematoma (IMH) is a static collection of clotted blood within the tunica media of the aortic wall. There are two main theories on the pathogenesis of IMH. The first theory is that IMH is due to spontaneous rupture of the vasa vasorum that supplies the aortic media. The second theory is that IMH results from complete thrombosis of the false lumen in an otherwise classic AD with an entry tear. IMH can also be caused by blunt trauma or penetrating atherosclerotic ulcers. IMH are classified using the same Stanford system as AD. The treatment for IMH is essentially the same as for AD. Type A IMH has traditionally been treated urgently with surgery, due to the risk of progression to AD, aortic rupture, or pericardial, pleural or mediastinal hemorrhage. Type B IMH are typically managed with medical therapy and often regress with time, although sometimes they can progress to dissection or aneurysms as well.

**Penetrating Atherosclerotic Ulcer**

Penetrating atherosclerotic ulcer (PAU) is an aortic atherosclerotic ulcer which has penetrated the internal elastic lamina into the media layer, and it can lead to dissection, IMH, aneurysm, and aortic rupture. The most common location for PAU is in the mid to lower thoracic descending aorta. The typical demographic is an elderly patient with many coexisting atherosclerotic atheromata and aneurysmal disease. Clinical presentation of PAU can range from asymptomatic to full-blown AD. Surgery to stabilize the disease is suggested for a PAU that causes acute aortic syndrome, or in patients with hemodynamic instability, aortic rupture, distal embolization, or a rapidly enlarging aorta. For a PAU found
incidentally in asymptomatic patient, medical management of cardiovascular risk factors with annual follow-up to assess interval change is the current recommended practice.\textsuperscript{3}

**Imaging findings OR Procedure details**

**Aortic Dissection**

The two most important signs in the diagnosis of AD is (i) the existence of aortic flap with its proximal extent and, (ii) presence of a double channel aorta.\textsuperscript{4} There is certain information which should be obtained once AD is identified on MDCT: extent of AD (Stanford Classification), site of entry tear, side branch involvement such as coronary, carotid, subclavian, mesenterics, renals, and iliacs, presence of aortic rupture; differentiation between true and false lumen, and size of false lumen as a predictor for rupture.\textsuperscript{1} Identification of the entry tear site is important because current endovascular stent-graft therapy targets the exclusion of the entry tear. The entry tear site is often at the most proximal location of the intimomedial flap and can be identified on contrast-enhanced MDCT in most cases. Generally, the entrance tear is perpendicular to the long axis of the aorta and therefore will be detected easier by using imaging planes perpendicular to the tear like oblique sagittal views.\textsuperscript{4}

Aortic rupture can be diagnosed by the presence of a high-density hematoma on unenhanced MDCT. Hemopericardium or right hemothorax usually is associated with rupture of the ascending aorta, while hemomediastinum and left hemothorax is more likely to be due to rupture of aortic arch and descending aorta, respectively.\textsuperscript{1} It is important to differentiate between true and false lumen because major side branches originating from the false lumen may be occluded after stent-graft insertion, especially in cases without a reentry tear. A simple way to discriminate true from false lumen is to demonstrate its communication with the uninvolved aortic segment. The false lumen usually is the larger lumen due to higher pressure in the false lumen than the true lumen. The false lumen may appear less opacified than the true lumen because of lower blood flow velocity. Intraluminal thrombus is more often seen in the false lumen owing to the slower flow velocity. Intimal calcification is more commonly seen along the walls of the true lumen or true lumen side of the intimomedial flap. The false lumen may show a Beak sign (acute angle between the outer wall of false lumen and the intimomedial flap). The Cobweb sign are slender areas of low attenuation "cobwebs" seen in the false lumen caused by debris from the dissection - although not sensitive, this sign is pathognomonic for false lumen.\textsuperscript{3}
Unstable Thoracic Aneurysm

Once again, the definition of an unstable thoracic aneurysms is one which is enlarging rapidly (this requires serial imaging), has signs of imminent rupture, or has already ruptured but now is contained. MDCT signs of imminent rupture include: high-attenuating crescent (hematoma, hemorrhage) in the wall of the aorta, discontinuous calcification in a circumferentially calcified aorta, an aorta that conforms to the neighboring vertebral body ("draped" aorta), and an eccentric nipple shape to the aorta.\(^3\) CT signs of rupture include hemothorax (usually the left hemothorax) and periaortic fat stranding. The plain film chest film is often helpful as the first modality to usually identify a rupture via the presence of a hemothorax.\(^4\)

Intramural Hematoma

The major CT finding for IMH is a crescentric or ring-shaped high attenuation of the aortic wall on an unenhanced MDCT. On unenhanced scans, this is easily recognized by the higher Hounsfield-unit value of the blood products in the wall in comparison with the flowing blood in the lumen. Aortic wall thickening on contrast-enhanced CT may be missed or alternatively may be confused with atheromatus mural thrombus. IMH is distinguished from AD by three things: (i) IMH does not enhance after contrast since there is no flow, (ii) usually do not spiral around the aorta, and (iii) there is no entrance tear and no direct communication with the aortic lumen.\(^3\) Several CT findings are associated with a worse outcome and should be actively sought and reported if found: (i) IMH with an ulcerlike projection (ULP) regardless of whether it is a true PAU is associated with increased complications of overt AD and rupture, and (ii) thickness of IMH greater than 11 mm was associated with progression of IMH to AD.\(^1\)

Penetrating Atherosclerotic Ulcer

The imaging finding for PAU on contrast-enhanced MDCT is a contrast-filled irregular outpouching of the aortic wall. They are usually focal lesions, most frequently located in the middle to lower descending thoracic aorta. Usually there is also extensive aortic atheroma and co-existent aneurysms present.\(^4\) PAU can lead to any of the previous acute aortic syndromes, so those imaging findings may also be present in acute presentations. PAU should be differentiated from atheromatous ulcers, which are confined within the intima. The location of intimal calcification can be helpful, in that atheromatous ulcers often conform to the expected aortic contour and calcified intima, whereas PAU will extend outward beyond the expected aortic margin and calcified intima.\(^1\) Whether the size of PAU has any association with its natural history and progression is controversial,
so currently the interval change on follow-up imaging is likely the more reliable indicator of disease progression.¹

Comparison of Modalities

Plain chest radiography is performed routinely in AAS but has a poor sensitivity of only 64% in the diagnosis of AAS. The sensitivity is even lower (down to 47%) for proximal aortic disease. Conversely, in the case of acute traumatic aortic injury, a normal chest radiography virtually excludes significant injury.⁴ Furthermore, chest radiograph may be helpful to exclude certain nonaortic causes of chest pain presentation like pneumothorax, pneumonia, etc. However, in the context of AAS diagnosis, chest radiography cannot be relied upon to exclude aortic disease.

Conventional angiography has been used for many years and was formerly considered the gold standard in diagnosis. However, aortography has been found to be less accurate and has the potential for false negatives due to thrombosed false lumen, simultaneous equal opacification of true and false lumens, and intramural hematoma.⁵ The imaging is limited only to one or two 2-D projections and only visualizes the vessel lumen but not the adjacent structures. Furthermore, conventional angiography is more invasive with higher complication rates, with major complication of dissection, embolization, myocardial infarction, or cerebrovascular accidents occurring at a rate of 5-6% with 0.2% mortality.⁴

Echocardiography can be divided into transthoracic (TTE) and transesophageal (TEE) modes. TTE is limited for evaluating AAS. However, TEE is about 95% sensitive and specific for diagnosing AD, IMH, and associated valvular regurgitation if the personnel who performs and interprets the test is highly experienced.⁴ The advantage of TEE is that it is a rapid and safe test. It may be performed at the bedside in a hemodynamically unstable patient and avoids unnecessary delays caused by transferring patients to the imaging department for CT or MR.⁶ However, there are several limitations to TEE. Imaging of the proximal aortic arch and descending thoracic aorta is difficult with TEE due to interference from the air-filled trachea and mainstem bronchus. TEE requires the use of conscious sedation and can cause bradycardia and elevation in blood pressure from the patient retching and gagging. Also, TEE is also contraindicated in patients with esophageal varices and strictures.⁵ The biggest limitation of all may be the lack of highly experienced personnel to perform and interpret TEE’s in the emergency setting, and given the operator-dependent nature of this test in terms of its diagnostic value, this is the major obstacle for Echo as the first line imaging modality for AAS.
MR has advantages of lack of radiation and the use of a significantly less nephrotoxic contrast agent. Metanalyses have shown that MR is the most accurate technique for diagnosis of thoracic aortic dissection, likely due to its superior tissue contrast.\textsuperscript{7} Given all these advantages, MR is the modality of choice for follow-up imaging in the chronic AD patient. However, MR is simply not practical in the acute unstable patient with AAS because of the relatively long examination times and difficult monitoring/treating unstable patients in the presence of a magnetic field.\textsuperscript{1}

With appropriately obtained MDCT data, the diagnostic sensitivity and specificity of contrast-enhanced MDCT for AAS is nearly 100\%.\textsuperscript{1} A major strength of MDCT is the ability to image the aortic branch vessels as well as the entire aorta without any of the limitations encountered with TEE. Furthermore, MDCT allows visualization of non-aortic structures and thus allows for non-aortic diagnoses such as pulmonary embolism.\textsuperscript{1} With the establishment of multidetector, helical, and dual-source technology, MDCT images can be obtained rapidly, which is critical in a hemodynamically unstable patient. The two drawbacks are ionizing radiation and contrast nephrotoxicity. MDCT for AAS should start off with an unenhanced scan to look for any IMH and high-density blood in the pericardium, pleural space, or mediastinum, indicating aortic rupture. Contrast greatly improves the accuracy of MDCT. Cardiac gating has allowed correction of cardiac motion artifacts and led to improved evaluation of the proximal aortic root and coronary arteries.\textsuperscript{3} Post-processing via multiplanar reformations, maximum intensity projections, and volume rendering have enabled improved visualization of complex anatomical relationships in AAS and facilitated communication with surgeons and emergency physicians.\textsuperscript{1} All this has led to MDCT being presently the modality of choice for imaging of suspected AAS.

Images for this section:
Fig. 1: Coronal MDCT of a man with aortic dissection involving only the descending thoracic aorta (Type B). The false lumen is the larger of the two lumens, with scattered intimal calcification seen in the smaller true lumen.
Fig. 2: Frontal CXR of a patient with unstable (leaking) descending thoracic aneurysm (white arrow) and right hemothorax (black arrow).
**Fig. 3:** Lateral CXR of a patient with unstable (leaking) descending thoracic aneurysm (white arrow) and right hemothorax (black arrow).
Fig. 4: Axial contrast-enhanced MDCT of a patient with unstable (leaking) descending thoracic aneurysm and right hemothorax. An = aneurysm. HTX = hemothorax.
Fig. 5: Patient with PAU which progressed to IMH and hemopericardium - Coronal CT showing PAU (black arrow) in descending thoracic aorta just distal to left subclavian origin.
**Fig. 6:** Patient with PAU which progressed to IMH and hemopericardium - Axial CT showing IMH in the ascending aorta at a later time.
Fig. 7: Patient with PAU which progressed to IMH and hemopericardium - Axial CT showing Hemopericardium (white arrow) in the left anterolateral aspect of the heart after further progression in the same patient.
Conclusion

1. Due to the often non-specific clinical presentation of AAS, the timely and accurate diagnosis via imaging is essential for guiding patient management.

2. The various types of AAS can be easily diagnosed and differentiated with the proper use of unenhanced and enhanced scans, cardiac gating, and image post-processing.

3. ECG-gated MDCT technology has continued to improve and has significant advantages over other modalities. It has become the modality of choice for assessment of AAS.

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