Acute aortic syndrome

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The term acute aortic syndrome (AAS) describes a spectrum of related emergency aortic conditions including aortic dissection, intramural haematoma, penetrating aortic ulcer with acute traumatic aortic injury (ATAI), all variably included in this definition due to the overlap of imaging features. Aortic dissection is the most common and immediately life-threatening, with up to 40% of patients dying before reaching hospital. The classic clinical feature of AAS is severe, sudden onset chest and/or interscapular pain. While traditional teaching describes the pain as tearing, sharp pain was found to be the most common description. There may be unexplained hypotension. Anterior chest pain is suggestive of type A dissection, whereas back pain suggests descending aortic involvement. AAS is, however, very varied in its presentation and it is important to note that the absence of ongoing clinical signs in an apparently well patient does not exclude the condition, as the patient’s symptoms may improve after the initial event. This can result in false reassurance, with rapid deterioration and mortality following discharge.

Important risk factors include hypertension (most common risk factor), connective tissue disorders (eg Marfan, Ehlers-Danlos, Loeys-Dietz syndromes), family history of AAS, aortic valve disease or recent aortic instrumentation/catheterisation. However, the absence of risk factors does not exclude the condition, and if the clinical history is suggestive of AAS this should be urgently investigated. Each hospital will have a different investigative strategy for suspected AAS. One potential strategy is to initially perform a bedside transthoracic echocardiogram (TTE).

1. Normal TTE – Confirmatory CT aortogram is then conducted which will also establish the extent of aortic pathology. It is important to note that echocardiography can demonstrate a ‘pseudo dissection flap’ (figure 1) due to reverberation artefact from the aortic wall; clinical correlation is again vital to prevent unnecessary investigations and surgery.

2. If TTE confidently diagnoses type A dissection, the patient could be considered for surgery with a transeosophageal echocardiogram (TOE) under general anaesthetic prior to operation. The rationale for general anaesthetic is that the act of swallowing the probe in an unsedated patient could produce a surge in blood pressure and worsen the condition. A normal TOE would prompt consideration of other diagnoses, CT may again be helpful.

3. If TTE is not immediately available, a CT aortogram should be conducted. While MR aortography may also be used, this is more time consuming and less readily available and as these patients often require close monitoring, is more problematic.

CT aortography

In some departments, an initial unenhanced CT is performed to detect intramural haematoma. Following iodinated intravenous contrast injection, bolus tracking or contrast test bolus with the region of interest on the ascending aorta is used to optimise opacification of the aorta. In the presence of chest/upper back pain, a typical scan region extends from above the aortic arch to at least the level of the diaphragmatic hiatus, with continuation of the scan to below the aortic bifurcation if abdominal involvement is suspected. ECG gating will remove motion artefact from the aortic root.

Aortic dissection

Aortic dissection produces a tear in the internal aortic wall (intima or intima and media). Blood tracks into the media and splits it, producing an intimo-medial flap which can extend in both antegrade and retrograde directions. Dissections may be classified as Stanford type A, which involve the ascending aorta, and type B dissections which occur distal to the ascending aorta. Because type A dissection can extend into the coronary arteries, aortic valve or pericardium there is a risk of sudden death.

In patients with aortic dissection, CT aortography demonstrates the intimal flap and allows differentiation between the true and false lumen (figure 2). ECG-gating through the aortic root may be considered to remove pulsation artefact, which can simulate a dissection flap (figure 3).

In the presence of type A dissection involving the aortic root, there may be a pericardial effusion, coronary artery dissection or areas of low density myocardium indicating infarction. Poor contrast enhancement of the abdominal organs may occur if the intimal flap occludes the artery supplying them. Over time, due to weakening of the aortic wall, aneurysmal dilatation may occur which can result in rupture.

Intramural haematoma

Intramural haematoma is a spontaneous event thought to be caused by bleeding from the vasa vasorum within the media, and represents 10-20% of cases who present with AAS. This is classified as type A or B as per aortic dissection. Expansion of the haematoma can weaken the wall, and if it ruptures into the lumen, can result in aortic dissection.

Intramural haematoma is well seen on non-contrast scan, where there may be crescentic high attenuation within the aortic wall. As there is no intimal tear this does not communicate with the lumen. No enhancement is seen on post-contrast imaging and the intramural thrombus is often still apparent, although if shallow can be incorrectly interpreted as mural thrombus, pericardial effusion or aortic wall thickening (figure 4) due to its relative low attenuation compared to intraluminal contrast.

Penetrating aortic ulcer

Penetrating aortic ulcers originate from ulcerating atherosclerotic plaques eroding through the aortic wall, which can in turn result in intramural haematoma, aortic dissection, or pseudoaneurysm/rupture.
Penetrating ulcer appears as a focal breach of the aortic wall communicating with the aortic lumen, and may have an associated intramural haematoma, aortic dissection or progressive pseudoaneurysm or complete rupture (figure 5).

Conclusion
Acute aortic syndrome is a life-threatening condition which often presents a diagnostic challenge to clinicians and imaging plays a vital role on the diagnostic pathway. It is important to be aware of the spectrum of findings that present as AAS to prevent unnecessary mortality.

References

Figure 2
Aortic dissection. Patient presented with acute central chest pain and hypotension. Images from non-gated thoracic CT aortogram. Left: Axial CT image shows a clear dissection flap (black arrow) in the ascending aorta dividing the true (T) and false (F) lumens. Middle: Sagittal CT reformat demonstrating the dissection flap extending into the left common carotid artery (arrowhead). Right: Coronal CT shows the dissection extending into the aortic root with poor filling of the false lumen (white arrow).

Figure 3
Motion artefact simulating aortic dissection flap. Patient presented with central chest pain radiating to the back. Left: Non-gated CT thorax. The white arrow indicates motion artefact in the ascending aorta and main pulmonary artery, which are commonly seen on non-gated scans and can simulate aortic dissection. Right: The artefact is no longer visible on subsequent gated cardiac CT on the same day.
**Figure 4**
Intramural haematoma.
Left: Axial CT aortogram. Cresenteric low attenuation is seen in the wall of the descending aorta (white arrowhead), consistent with intramural haematoma. Right: Sagittal CT image again demonstrates type B intramural haematoma, with low attenuation soft tissue in the aortic wall extending from the distal arch into the abdomen (white arrow).

**Figure 5**
Penetrating aortic ulcer. Coronal CT image demonstrates a small focus of intramural contrast communicating with the aortic lumen (black arrow) consistent with penetrating ulcer. There is also a low density soft tissue cuff around the ascending aorta which is in keeping with secondary intramural haematoma (arrowhead). Note the large pericardial effusion (asterisk).