Patient

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Aortic Dissection

Aortic dissection is defined as disruption of the medial layer of the wall of the aorta provoked by intramural bleeding, resulting in separation of the aortic wall layers and subsequent formation of a true lumen and a false lumen with or without communication. Acute aortic dissection (less than 14 days since onset) is distinct from subacute (15-90 days) and chronic aortic dissection (more than 90 days)[1].

The presentation of aortic dissection can be quite variable. It is not an easy diagnosis to make and an index of suspicion is required.

Epidemiology

The incidence is approximately 3-4 per 100,000 per year[2].

- Aortic dissection is the most common emergency affecting the aorta.
- The male:female ratio varies - in one study, almost 70% of cases were in males.
- It is most common between the ages of 50-70, being rare below the age of 40.
- Aortic dissection is very rare in children but it has been reported in association with coarctation of the aorta.

Risk factors

Approximately 50-75% of patients with dissection will have evidence of hypertension or a previous diagnosis[2]. Other risk factors include smoking and raised cholesterol. The most common risk factor is hypertension. Other risk factors include pre-existing aortic diseases or aortic valve disease, family history of aortic diseases, history of cardiac surgery, cigarette smoking, direct blunt chest trauma and use of intravenous drugs (eg, cocaine and amfetamines)[1].

Inherited risks include Marfan's syndrome, Ehlers-Danlos syndrome and familial thoracic aortic aneurysm type 1 and type 2[3, 4]. Congenital bicuspid aortic valve is also a risk.

Classification[5]

The Stanford Classification in common use classifies the dissections into type A and type B:

- Type A involves ascending aorta (DeBakey types I and II).
- Type B does not involve the ascending aorta (DeBakey type III).

The DeBakey Classification classifies the dissections into:

- Type I: aorta, aortic arch, and descending aorta.
- Type II: ascending aorta only.
- Type III: descending aorta distal to left subclavian.

Presentation

Aortic dissection may present with chest pain, aortic regurgitation, myocardial ischaemia, congestive heart failure, pleural effusions, syncope, neurological symptoms (eg, acute paraplegia, upper or lower limb ischaemic neuropathy), mesenteric ischaemia and acute kidney injury[1].

Aortic dissection often presents in two phases:

- After a first event with severe pain and pulse loss, the bleeding stops.
- The second event starts when the pressure exceeds a critical limit and rupture occurs, either into the pericardium with cardiac tamponade or into the pleural space or mediastinum.

Fluid extravasation into the pericardium, pleural space and/or mediastinum demands urgency. Sudden death is not uncommon.

Signs and symptoms

- The typical patient is a man in his 60s with hypertension and sudden onset of chest pain.
- The majority of patients with aortic dissection, present with a sudden severe pain of the chest or back, classically described as 'ripping'.
- However, some may describe mild pain in the chest, back or groin, and it is easy to dismiss such cases as musculoskeletal.
- Hypertension.

There are a number of important features about the pain:

- In aortic dissection, pain is abrupt in onset and maximal at the time of onset. In contrast, the pain associated with acute myocardial infarction starts slowly and gains in intensity with time. It is usually more oppressive and dull.
- The pain migrates as the dissection progresses. This is also very important.
- In proximal dissections, the pain is usually retrosternal; however, with distal dissections the location is between the scapulae and in the back. Hypertension is typically associated with distal aortic dissection.
- Although *tearing* is the classical description, the pain is described as *sharp* more often than tearing, ripping, or stabbing.

Other symptoms which may occur are a direct result of occlusion of smaller arteries by the dissecting process:

- Angina due to involvement of the coronary arteries.
- Paraplegia due to involvement of the spinal arteries.
- Limb ischaemia due to distal aortic involvement.
- Neurological deficit due to carotid artery involvement.
- Pulse deficit may be present initially, may develop, or may occur transiently. There is a difference in blood pressure in limbs on the right and left side of the body.

Differential diagnosis

- Acute coronary syndrome with and without ST elevation.
- Aortic regurgitation without dissection.
- Aortic aneurysms without dissection.
- Musculoskeletal pain.
- Pericarditis.
- Mediastinal tumours.
- Pleuritis.
- Pulmonary embolism.
- Cholecystitis.
- Atherosclerotic or cholesterol embolism.

The most important differential diagnosis is that of myocardial infarction which may co-exist with aortic dissection. Thrombolysis may well prove fatal if aortic dissection is present.

Aortic intramural haematoma is pathologically distinct with no intimal tear. However, the clinical course and mortality rates are similar in the two conditions. The haematoma may rupture through the intima, to evolve into the typical dissection.

Investigations

Often the first problem is to distinguish aortic dissection from myocardial infarction. Both conditions may exist if the dissection involves the coronary ostium. For this reason, the ECG is very important.

ECG

- There will be ECG signs of acute myocardial infarction if this is present and this could lead to thrombolytic therapy.
- A normal ECG is present in one third of patients with coronary involvement and most of these patients have nonspecific ST-T segment changes.
- About 20% of patients with type A dissection have ECG evidence of acute ischaemia or acute myocardial infarction.

Imaging

Anyone with suspected aortic disease and ECG evidence of ischaemia must have diagnostic imaging before thrombolytic therapy is started, although if there is just myocardial infarction, the sooner thrombolysis is started the better. Diagnostic imaging cannot be performed in all patients presenting with myocardial ischaemia, and erroneous administration of thrombolytic therapy may be unavoidable in the occasional patient who is subsequently found to have aortic dissection.

- CXR alone is inadequate to exclude aortic dissection.
- Transthoracic/transoesophageal ultrasound will give an indication of site and extent of dissection.
- CT scanning may be used to confirm the diagnosis but is less helpful in complications and prognosis.
- MRI scanning will confirm diagnosis, will identify involvement of other vessels and will be increasingly useful as scanning times decrease, and with better access. Of all of the imaging modalities it has the best sensitivity (98%) and specificity (98%) for aortic dissection[2].
- Colour flow Doppler is useful for assessing aortic regurgitation.

Multiorgan failure is a major cause of death and results from impaired flow in abdominal arteries. Ultrasound is of limited value because of poor-quality images. Aortography and intravascular ultrasound may reveal:

- Static narrowing of a branch artery.
- Dynamic narrowing of a branch artery.

Both mechanisms may be present simultaneously at different levels. Angiography may be necessary to identify these mechanisms, as interpretation of MRI or CT images may be difficult. Involvement of side branches of the aorta such as the coeliac artery, mesenteric arteries or renal arteries can be detected with intravascular ultrasound.

In aortic dissection without a distal tear (re-entry) the aorta may become totally obstructed as the true lumen is compressed by the false lumen. This will show on CT or MRI. Angiography is necessary to demonstrate the full anatomical situation including collateral flow. In the future, MRI angiography may replace conventional angiography.

Management

General measures

- Intravenous access.
- Adequate analgesia - eg, morphine.
- Transfer to an intensive care unit or high dependency unit.
- Hypertension must be managed aggressively in all cases to reduce further damage. The aim is a systolic pressure of between 100 and 120 mm Hg.
- Intravenous beta-blockers are usually employed, as they also reduce the force of ventricular contraction. If vasodilators are used they should be combined with beta-blockers.
- There may be evidence of blood loss due to sequestration of blood. Separate lines are required for administration of blood and drugs.

Surgery

Surgical intervention may involve the placing of stents or grafts to the aorta but accurate assessment is essential first, as there may be entry, re-entry and multiple tears.

For acute type A (types I and II) dissection, surgery aims to prevent aortic rupture and pericardial tamponade and to relieve aortic regurgitation [6]. Implantation of a composite graft in the ascending aorta, with or without re-implantation of coronary arteries, is performed. Either total aortic arch replacement or partial or hemiarch replacement may be considered [7, 8].

For type B aortic dissection, thoracic endovascular aortic repair (TEVAR) in addition to optimal medical treatment is associated with improved five-year aorta-specific survival and delayed disease progression. However, open repair still has a significant role, as endovascular repair is not applicable in all patients and further research is required regarding long-term benefits [9]. TEVAR seems to have a more favorable outcome regarding aortic remodelling and aortic-specific survival rate when compared with medical therapy alone [10].

Chronic aortic dissections [1]

Patients with involvement solely of the descending aorta may be treated conservatively with medical therapy and regular clinical and ultrasound follow-up. Surgery is associated with a significant risk of paraplegia but should be considered if there is evidence of progression of the dissection. Repair can be either by open surgical techniques or increasingly by endovascular approach. If the aortic valve is involved it may need resuspension or replacement.

Complications

Problems arising from aortic dissection will depend on the site and extent of the involvement and if and to where the false lumen ruptures. Dissection can result in occlusion of aortic branches, with resulting damage to supplied organs. There may be occlusion of the renal, iliac, spinal or coronary arteries. Rupture of the false lumen may occur back into the aorta, or externally into the mediastinum or pericardium.

- Hypotension indicates a poor prognosis, as it may be due to cardiac tamponade or myocardial infarction.
- An aortic diastolic murmur occurs in 50% due to aortic regurgitation. Wide pulse pressure may be absent.
- After a period of pain, cardiac failure may result from gross aortic regurgitation.
- Cardiac tamponade can cause hypotension and syncope. Signs of pericardial involvement such as a pericardial friction rub, jugular venous distension or a paradoxical pulse suggest that surgical intervention may be required.
- Sometimes pyrogens can be released from the aortic wall and cause pyrexia that will be misleading.
- Mesenteric malperfusion is associated with a worse prognosis [11].
- Involvement of the coeliac artery can produce persistent abdominal pain, elevation of acute phase proteins and increase of lactate dehydrogenase.
- Transient pulse differences suggest involvement of the brachiocephalic or subclavian arteries.
- A superior vena cava obstruction syndrome may be seen.
- When dissection involves the distal aorta, it often involves the renal arteries. Urgent repair is required. Urine output must be closely monitored and this requires catheterisation. Monitor fluid balance and beware of hypotension with angiotensin-converting enzyme (ACE) inhibitors.

Prognosis

- Diagnosis is difficult but mortality is high and increases by the hour.
- Rupture is catastrophic and aortic rupture has an 80% mortality.
- Up to 20% die before reaching hospital [2].
- Early intervention and control of hypertension dramatically improve the prognosis.

- Despite adequate antihypertensive therapy, the long-term prognosis for patients with stable type B (not involving the ascending aorta) dissections is characterised by a significant aortic aneurysm formation in 25-30% within four years and survival rates from 50-80% at five years and 30-60% at 10 years [12].
- Survival has improved greatly over the years [13]. Without surgery the prognosis for patients with involvement of the ascending aorta is poor. Although successful surgery greatly increases the chances of survival, there remains significant operative mortality.
- Late deaths are usually due to aortic rupture.

Prevention

The management of patients with predisposing inherited diseases such as Marfan's syndrome and Ehlers-Danlos syndrome should include:

- Periodic aortic diameter screening.
- Lifelong beta-blockade.
- Consideration of prophylactic replacement of the aortic root if dilated.
- Moderate restriction of physical activity.

Further reading & references

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