Acute aortic syndromes

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Abstract

Acute aortic syndromes comprise acute aortic dissection, intramural haematoma, symptomatic penetrating aortic ulcers and traumatic aortic dissection. These conditions result primarily from disruption of the outer aortic layer, and involve thinning of the aortic wall, increased wall stress, progressive dilatation, evolution of intramural haemorrhage, and possible dissection and rupture. Chronic hypertension and connective tissue disorders are often implicated. Echocardiography, contrast-enhanced CT, dynamic MRI and aortography are currently used to confirm the diagnosis. Aortic dissection is primarily classified according to anatomical characteristics; those with and without ascending aortic involvement are distinguished for prognostic and therapeutic reasons. In general, open surgery is indicated when dissection involves the ascending aorta, whereas medical management or endovascular stent-graft implantation are reserved for cases where the ascending aorta is spared. Pathology involving the aortic arch may be treated using a hybrid approach combining debranching of the head and neck vessels and interventional stent-graft implantation. Stent-graft induced remodelling of dissected aorta seems to have long-term benefits both in complicated and so-called uncomplicated distal dissection. In addition, long-term medical therapy to control hypertension is of paramount importance in all patients who have survived aortic dissection to reduce late complications, including recurrent dissection, aneurysm formation or late extension or rupture.

Keywords Acute aortic syndrome; aortic dissection; hybrid approach; intramural haematoma; penetrating aortic ulcer; peripheral vascular disease; stent-graft

'Disease is very old, and nothing about it has changed. It is we who change as we learn to recognise what was formerly imperceptible'

Jean Martin Charcot

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Acute aortic syndrome is an established term that includes aortic dissection, intramural haematoma (IMH) and symptomatic penetrating aortic ulcer (PAU), as well as traumatic aortic lacerations. First described by Morgagni more than 200 years ago,[1] acute aortic dissection requires a tear in the aortic intima that is commonly preceded by medial wall degeneration or cystic medial necrosis. Although early studies[2] highlighted the high mortality rate and infrequency of ante mortem diagnosis,[2] knowledge regarding the incidence of aortic dissection in the general population is limited. Studies suggest an incidence of 2.6–3.5 cases per 100,000 person/year[3] and the prevalence is 0.2–0.8% in large autopsy series. Peak incidence is in the sixth and seventh decade of life and men are affected twice as commonly as women.[4] Aortic dissection in individuals under the age of 40 years is most common in those with Marfan's syndrome and during pregnancy.

Pathology

Aortic dissection is believed to begin with formation of a tear in the aortic intima, which exposes an underlying diseased medial layer directly to the driving force of intraluminal pulsatile blood. Penetration cleaves the diseased medial layers and progressively dissects the aortic wall. Driven by persistent intraluminal pressure, the dissection may progress antegradely (and sometimes retrogradely) along the aortic wall from the site of initial intimal tear, thereby forming a false lumen. Shear forces may lead to further tears in the intimal flap (the inner portion of the dissected aortic wall) to create additional entry or exit sites into the false lumen. Distension and systolic pressure within the false lumen may lead to dynamic compression of the true lumen and distal malperfusion.

Aortic IMH is considered a precursor of dissection and originates from ruptured vasa vasorum within the medial wall layers, resulting in aortic wall infarction that may provoke a secondary tear and classic aortic dissection. IMH is usually located in the descending aorta, typically associated with hypertension, and may extend, progress or resorb.[5,6]

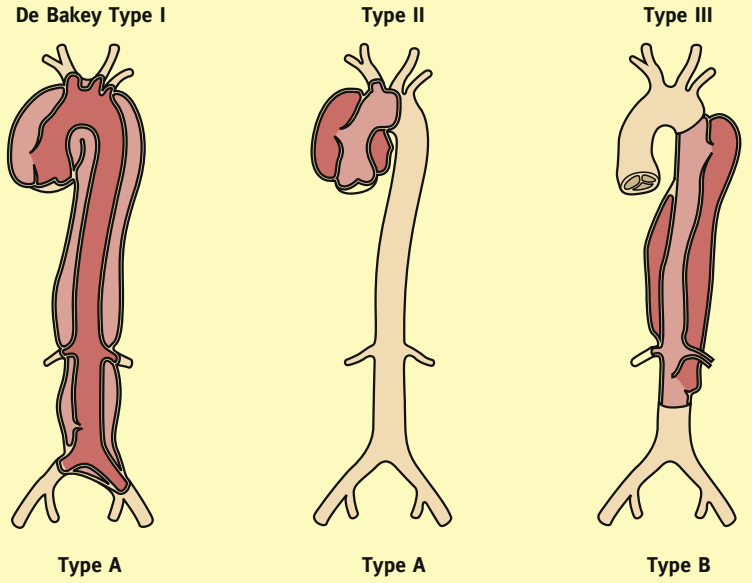
Deep penetrating aortic atherosclerotic plaques can lead to IMH, aortic dissection, or perforation. Non-invasive imaging has recently elucidated this disease process, which often further complicates IMH.

Classification

The prime classification is based on the anatomical location of the intimal tear and degree of propagation of the false lumen. Intimal tears occur at points of presumed greatest haemodynamic stress; namely, the lateral wall of the ascending aorta and just distal to the ligamentum arteriosum of the descending thoracic aorta. Overall, 65% of intimal tears occur in the ascending aorta, 20% in the descending aorta, 10% in distal aortic arch and 5% at abdominal level. Two anatomical classification systems predominate (Figure 1) – in both, dissections with and without ascending aortic involvement are distinguished for prognostic and therapeutic reasons.[7,8]

Aortic dissection may also be classified according to the timing of diagnosis relative to the onset of symptoms: acute within 2 weeks, subacute within 2–8 weeks, or chronic beyond 8 weeks. About one-third of patients with aortic dissection fall into

Commonly used classification systems in aortic dissection



Acuity:

Acute: <2 weeks after onset
Subacute: 2–8 weeks after onset
Chronic: >8 weeks after onset

Anatomic location:

Ascending aorta: Stanford Type A, De Bakey Type II
Ascending and descending aorta: Stanford Type A, De Bakey Type I
Descending aorta: Stanford Type B, De Bakey Type III

Pathophysiology:

Class 1: Classical aortic dissection with initimal flap between true and false lumen
Class 2: Aortic intramural hematoma without identifiable intimal flap
Class 3: Intimal tear without hematoma (limited dissection)
Class 4: Atherosclerotic plaque rupture with aortic penetrating ulcer
Class 5: Iatrogenic or traumatic aortic dissection (intra-aortic catheterization, high-speed deceleration injury, blunt chest trauma)

Figure 1

the latter category; both mortality and the risk of progression decrease over time, with implications for the most appropriate treatment strategy.[9,10]

Aetiology and pathogenesis

Any disease process that undermines the integrity of the elastic or muscular components of the media predisposes the aorta to dissection, and degeneration of this layer is the major predisposing factor in most non-traumatic aortic dissection (Table 1). Degeneration of the media as a result of enhanced apoptosis is a feature of several hereditary connective tissue defects, notably Marfan's and Ehlers–Danlos syndromes. In the absence of Marfan's syndrome, medial degeneration is usually minor in most cases of aortic dissection but is nevertheless qualitatively and quantitatively greater than that expected as part of the ageing process, possibly as a result of increased apoptosis secondary to hypertension.

A bicuspid aortic valve is associated with acute aortic syndrome in 7–14% of patients. Other congenital cardiovascular abnormalities may predispose to dissection, including coarctation of the aorta and giant cell arteritis. Direct trauma to the aorta may cause dissection, whereas blunt trauma tends to cause localized tears, haematomas or transection; iatrogenic trauma (e.g. during cardiac catheterization, cardiac surgery or insertion of an intra-aortic balloon pump) may induce dissection, probably as a result of direct trauma to the aortic intima.[11]

Clinical features

Diagnosis and effective clinical management of acute aortic syndromes require a high level of clinical suspicion and prompt action. The differential diagnosis in acute aortic dissection includes acute coronary syndrome, pulmonary embolism, pneumothorax, pneumonia, musculoskeletal pain, acute cholecystitis,