

Thoracic aortic aneurysms

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The aorta, acting as the main conduit through which cardiac output is delivered to the systemic arterial bed, is continuously exposed to high pulsatile pressure and shear stress, making it prone to mechanical injury. It is also more prone to rupture than other vessels, particularly with the development of aneurysmal dilation, since its wall tension (governed by the law of Laplace, i.e. proportional to pressure x radius) is intrinsically high. The following article is an outline of thoracic aortic aneurysms and the sometimes catastrophic manifestations of diseases of the aorta above the diaphragm.

Thoracic aortic aneurysms

A thoracic aortic aneurysm is the term used to describe when a segment of the artery is dilated by more than 50% of its original diameter. Aneurysms of the thoracic aorta may be divided into those involving the ascending thoracic aorta, the aortic arch and the descending aorta. The location of the aneurysm affects clinical manifestations, natural history and treatment options, as well as offering clues as to the aetiology.

Inherited forms of aortic disease

The degenerative, sometimes cystic, change in the elastic tissue and smooth muscle of the aorta associated with aortic aneurysms has been dubbed cystic medial necrosis. These pathological findings are associated with aneurysm formation in Marfan syndrome, although they may be found in isolation, without other features of the syndrome. This degenerative change in the aortic media predisposes to dilation and dissection.

Marfan syndrome is one of the most common inherited disorders of connective tissue, with a prevalence of about 1 in 10,000. It is inherited in an autosomal dominant fashion. Recently described genetic abnormalities associated with Marfan syndrome include mutations in the fibrillin-1 gene on chromosome 15, which encodes a protein in the extracellular matrix. A second chromosomal locus on 3p 24-25 has been identified for a Marfan-like condition with thoracic aortic aneurysms.

Familial clustering of thoracic aneurysms may occur in the absence of Marfan syndrome, but there appears to be considerable genetic heterogeneity within this group. Ehlers Danlos syndrome and osteogenesis imperfecta are disorders of collagen synthesis also associated with medial degenera-

tion, aneurysm formation and dissection. However, because of their comparative rarity, these are seen less frequently than Marfan-associated, familial non-syndromic or sporadic cases.

Natural history

Aetiology, size and location of thoracic aortic aneurysms all contribute to natural history. Symptomatic patients have a much poorer prognosis than asymptomatic patients. Syphilitic aneurysms tend to expand and rupture more frequently than atherosclerotic aneurysms. Cardiovascular mortality unrelated to rupture or dissection in patients with atherosclerotic aneurysms is high, related to the high incidence of extensive atherosclerotic disease elsewhere, particularly in the coronary artery tree. These subjects present at an older age than those with Marfan syndrome or other forms of cystic medial necrosis. Aneurysmal dilation of the aorta is increasingly screened for, and monitored on a regular basis in Marfan syndrome.

In series which examined the natural history of thoracic aneurysms, rupture of the aneurysm was the most common cause of death, with rates ranging from 42 to 70%. Size is an important factor in risk of rupture, which escalates rapidly once the diameter of the aorta exceeds 5-6cm. The five year risk of rupture increases up to five-fold for aneurysms 6cm or greater. Thoracic aneurysms enlarge at an average rate of 0.2-0.4cm per year.

Despite these figures, the prognosis of thoracic aneurysms has improved in recent decades, one population-based study reports a five year survival of 56% from 1980-1994 vs 19% from 1951-1980.

Clinical features

Many patients with thoracic aortic aneurysms are asymptomatic at the time of presentation and detection of the aneurysm occurs during investigation for unrelated reasons, or as part of screening in certain familial cases (e.g. Marfan syndrome).

Symptoms tend to develop late in the course of aneurysmal dilation and result from local effects on surrounding structures. Ascending aortic aneurysms may cause aortic valvular regurgitation, giving rise to symptoms of congestive cardiac failure. Enlargement of the sinuses of Valsalva may cause myocardial ischaemia, or infarction due to arteries, or thromboembolism. Alternatively, these sinuses may rupture directly into the right ventricular cavity, right atri-



Figure 1 (called a): PA chest x-ray demonstrating an ascending aortic aneurysm

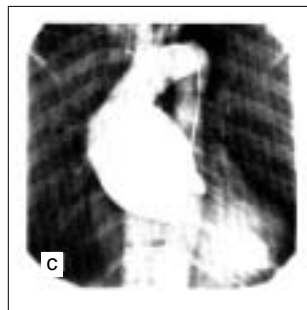


Figure 2 (called c): Aortogram of an ascending aortic aneurysm

um or pulmonary artery, causing heart failure associated with a continuous murmur.

Compression of the superior vena cava (SVC) may produce venous congestion in its distribution. Dyspnoea or cough may be caused by compression of the trachea or bronchi, dysphagia by compression of the oesophagus, or hoarseness by compression of the left recurrent laryngeal nerve. Chest pain related to compression of adjacent structures, or the erosion of ribs or vertebrae, is typically positional.

Rupture may occur into the pericardium, causing tamponade, or into the left pleural space, tracheobronchial tree oesophagus, SVC or pulmonary artery, giving rise to pleuritic pain, dyspnoea, haemoptysis, haematemesis or heart failure.

Diagnosis

The chest x-ray is frequently the first test to suggest the diagnosis in asymptomatic patients. A lateral projection will add to the diagnostic information. However, approximately 17% of patients with aneurysms or dissections will have no abnormalities on chest radiography. Computer tomography (CT) is the most widely used non-invasive technique for the diagnosis of thoracic aneurysms, revealing size, location and presence of thrombus or chronic dissection. It is a useful method of assessing the rate of growth and determining the timing of surgery. However, it involves

Table 1: Aetiology of thoracic aortic aneurysms

- Atherosclerosis - cause of the majority of descending aortic aneurysms
- Cystic medial necrosis - cause of the majority of ascending aortic aneurysms
- Syphilis - usually affects ascending the aorta and arch rather than the descending aorta
- Mycotic aneurysms - site depends on source of infection, e.g. endocarditis related, infected angiography catheters or direct infection from mediastinal lymph nodes (e.g. TB)
- Rheumatoid disorders - e.g. ankylosing spondylitis, which primarily affects the ascending aorta
- Trauma
- Other - e.g. associated with coarctation.

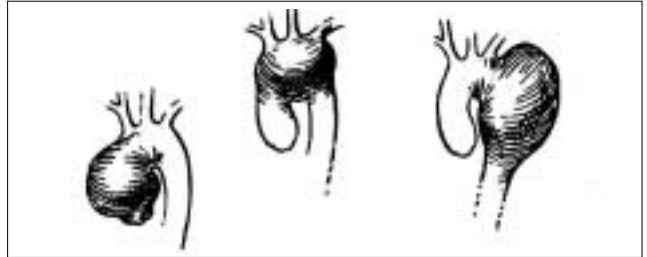


Figure 3: Types of thoracic aortic aneurysms

administration of contrast medium, which is associated with allergic reactions. Recently, magnetic resonance imaging (MRI) has come to the fore as an extremely useful imaging technique in the diagnosis of thoracic aortic aneurysms, yielding precise information about adjacent structures as well as the aneurysm itself.

Transoesophageal echocardiography (TOE) with colour Doppler imaging is vastly superior to transthoracic echocardiography for the diagnosis of thoracic aneurysms. It provides reliable information in the assessment of aortic disease, the presence and degree of aortic regurgitation and the results of surgery. Intra-operative TOE may be used to assess the presence of atherosclerosis and the adequacy of reparative procedures on the aortic valve.

Aortography, traditionally the gold standard procedure in the diagnosis of thoracic aortic disease, is losing ground to the newer, less invasive diagnostic procedures above. However, it may provide important information, particularly regarding the presence of coronary artery, brachiocephalic or renal artery disease to aid in accurate pre-operative risk assessment.

Management

The care of the patient with a thoracic aneurysm is quite complicated. The decision to intervene surgically must be based on the risk of rupture, the patient's life expectancy and detailed pre-operative assessment of operative risk. The modern surgical treatment of disorders of the thoracic aorta began in the 1950s when Gross, Swan, Lam, De Bakey and their associates reported successful treatment of coarctation and aneurysms of the descending aorta with segmental resection and graft replacement.

Successful procedures for resection and graft replacement of the ascending aorta and aortic arch, using cardiopulmonary bypass, were reported in 1956 and 1957 by Cooley and De Bakey.

These advances in surgical technique have revolutionised the management of the condition, as it is now possible, in selected cases, to electively repair the aneurysm to reduce the risk of rupture and other complications.

Patients with Marfan syndrome and cystic medial degenerative disease should undergo elective replacement of the ascending aorta and aortic root when the aortic diameter exceeds 5-5.5cm. Medical treatment with beta-blockers has been shown to slow the progression of aortic dilation and reduce the risk of dissection in patients with Marfan syndrome, probably by reducing the systolic ejection impulses. The surgical procedure of choice is aortic root replacement with a composite valve and conduct coronary reimplantation. Intervention at an earlier stage may be nec-

essary if there is significant aortic regurgitation.

Degenerative aneurysms confined to the ascending aorta are treated by graft replacement when the diameter of the aorta exceeds 5-5.5cm, or if symptoms are present. Surgery on the aortic arch usually requires the use of hypothermic cardiopulmonary bypass and a period of circulatory arrest. Focal or diffuse neurological complications occur frequently in 3-18% of patients, but permanent disability is less frequent. However, because of this risk, elective surgery is generally advised for aneurysms >5.5-6cm, or if the aneurysm exceeds 5-6cm. Thirty day operative survival for repair of descending thoracic aneurysms is greater than

90%, but the risk of paraplegia is significant, estimated at around 5% for isolated descending thoracic aneurysms. In all cases, a rapid increase in the size of the aneurysm may be taken as an indication for surgical repair. Organ compression, aortic insufficiency or ascending aortic dissection are well accepted indications for surgical intervention, regardless of aortic size.

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HEARTWISE

The Palpitating Quiz



Nora keeps coming in and going on about this sweating; she says it comes on suddenly. She was always a bit nervy you know. I got a couple of quite high blood pressures, although most are pretty normal, and I'm sure the high ones were white coat hypertension.

Is it nerves? Is it white coat? What might be going on and what tests would you do?

The prize is a £400 travel bursary sponsored by



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Entries to the Editor, HeartWise, Eireann Publications, 25 Windsor Place, Dublin 2. Fax: 4753311.

The closing date for entries is November 24th 2000. The Editor's decision is final.

Answer to the Summer edition palpitating quiz:

The correct diagnosis is peripheral embolus with atrial fibrillation.

Summer 2000 winner

Congratulations to Dr Brendan Daly, Turloughmore Health Centre, Athenry, for correctly answering the summer issue quiz.

