Difficulties in the diagnosis of acute aortic dissection

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IINTRODUCTION

The annual incidence of acute aortic dissection (AAD) is approximately 5–10 per million population and of these 80–90% of patients are over 60 years old. AAD has varied clinical presentations which may make prompt diagnosis difficult. Delayed or missed diagnosis can have fatal consequences with a mortality of 1% per hour if left untreated. The following case reports illustrate the delays both in diagnosis and initiation of definitive care. They also highlight how few patients are treated appropriately for associated hypertension once the diagnosis is confirmed. This article will suggest how improvements could be made in the diagnosis and management of AAD with reference to a review of the International Registry of AAD (Hagan et al, 2000).

DISCUSSION

A review of 464 cases of AAD from the International Registry of AAD (Hagan et al, 2000) concluded that the incidence of the classical findings in the history and examination of patients with AAD were not as frequent as previously thought.

Sudden onset of pain was present in 85% of patients, differing from the gradual onset of chest pain commonly associated with myocardial ischaemia or infarction. In addition, the pain was described as sharp in nature in 65% and 'tearing' in only 50%, contrasting with these four patients whose pain was abrupt with no preceding symptoms except syncope. As many as 72% of patients in the registry had known hypertension and examination revealed only 15% had a pulse deficit and 32% had aortic regurgitation. Only 31% of patients had normal 12-lead electrocardiograms, with non-specific ST segment changes being the most common abnormal finding. A widened mediastinum was present in 62% of patients on chest radiography.

In practice AAD is often initially excluded because patients do not present with the classical signs and symptomatology. A significant number of those studied in the registry would have remained undiagnosed if this had been the case.

Imaging

Once AAD is suspected urgent imaging of the thoracic aorta should be arranged. If clinical suspicion remains high despite a normal scan, a second imaging modality should be considered (Sarasin et al, 1996). This is borne out in two of these patients in whom computed tomography (CT) was negative but further imaging or review at a tertiary centre ultimately lead to the correct diagnosis. The ideal imaging tool is governed by its sensitivity, 24-hour availability and

CASE REPORT 1

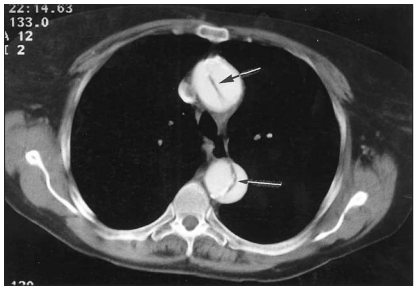
A 69-year-old woman with known hypertension presented to hospital with sudden onset severe central chest pain radiating between her shoulder blades and into her abdomen. This was associated with loss of power and sensation in her left leg lasting for 30 minutes. She complained of residual discomfort between her scapulae. Examination revealed a well-looking woman with unequal blood pressures in both arms: right 140/70 mmHg, left 100/80 mmHg, a pulse rate of 84/min and no audible murmurs. There was no vascular or neurological deficit in either leg. 12-lead electrocardiogram showed left ventricular hypertrophy by voltage criteria and chest radiograph was normal. Urgent spiral computed tomography of the chest and abdomen showed an extensive type A dissection of her aorta from her aortic valve to the abdominal aortic bifurcation (*Figure 1*). Despite attempts at haemodynamic resuscitation, she died in the ambulance en route to the regional cardiothoracic centre. Post mortem revealed a tense haemopericardium.

CASE REPORT 2

A 74-year-old hypertensive man presented with severe sudden onset sharp chest pain. He appeared pale and clammy with unequal blood pressures in the arms 130/90 mmHg right and 99/50 mmHg left but no pulse deficit and normal heart sounds. Chest radiography and 12-lead electrocardiogram were normal. Although aortic dissection was considered, he was initially treated as having unstable angina and heparinized. Following review by a consultant physician the next day computed tomography (CT) of the chest was performed which was reported to be normal.

As clinical suspicion of aortic dissection remained high he was transferred the same day to the regional cardiothoracic centre for aortography. The original CT was reviewed by a cardiothoracic radiologist at the regional centre and type B aortic dissection was diagnosed,therefore aortography was not required. He was managed conservatively on the intensive care unit with hypotensive agents and made an uneventful recovery.

Figure 1. Arrows demonstrate intimal flaps in both the ascending and descending aorta.



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the preferred investigation of the regional cardiac surgeons.

CT scanning is used most frequently in the UK, with a sensitivity ranging from 83 to 96% and specificity of 90–100% (Thorsen et al, 1986; Nienaber et al, 1993; Sarasin et al, 1996; Small et al, 1996). In addition, films can usually be faxed to the local cardiothoracic centre with relative ease. In the authors' hospital it is also the preferred imaging of the regional cardiac surgeons. One drawback is the need to move an unstable patient from the resuscitation area for a considerable period of time.

While transthoracic echocardiography can be performed at the bedside, it has a low sensitivity, 60–80% for type A dissections and 50% for type B (Nienaber et al, 1993; Sarasin et al, 1996) and hence relatively poor diagnostic value. On the contrary, transoesophageal echocardiography (TOE) has a sensitivity of 95–100% (Erbel et al, 1989; Adachi et al, 1991; Ballal et al, 1991; Chan, 1992; Armstrong et al, 1998) and has in the past been advocated as the single tool required to exclude the diagnosis (Banning et al, 1994). It also has the added advantage of being able to be done in the resuscitation room. Unfortunately, it has limited availability in most UK hospitals as it requires a skilled and experienced operator. In inadequately sedated patients and/or cases of poor operator technique, the procedure may precipitate a hypertensive surge and extend the aortic dissection. Moreover, not all cardiac surgeons are happy to operate on the findings of TOE alone and may demand additional imaging before transfer (Chu et al, 1998).

Although magnetic resonance imaging has equally high sensitivity (95–100%; Nienaber et al 1992, 1993), it introduces the potential for even greater delays and restricts access to a seriously ill patient. The choice of imaging tool will thus be governed by several factors. It is recommended that each hospital devises a protocol for investigation of AAD with the agreement of the relevant specialties to ensure that these patients are managed rapidly and effectively.

CASE REPORT 3

A 71-year-old woman with known ischaemic heart disease presented with sudden 'searing' pain in her epigastrium which subsequently radiated to her chest, back and neck. This was quickly followed by a very brief syncopal episode. On examination she appeared distressed and very unwell with a pulse of 80/min and a blood pressure of 80/65 mmHg left arm and 102/68 mmHg right arm. Cardiorespiratory examination revealed an elevated jugular venous pressure and bibasal crepitations. The epigastrium was tender with no palpable abdominal aneurysm. Chest X-ray revealed marked cardiomegaly and electrocardiogram was normal.

She was given intravenous opiates and fluids were witheld. Urgent computed tomography (CT) demonstrated a large pericardial effusion (*Figure 2*) but no dissection was noted at this time. A pericardial catheter was inserted which drained 50 ml of frank blood and immediately her blood pressure rose to 180/100 mmHg. On further review of the CT it was felt that there was a very short type A dissection of the ascending aorta and plans were made to transfer her to the regional cardiothoracic centre. Unfortunately, while awaiting departure she died following an electromechanical dissociation arrest. Post mortem confirmed a short dissection through the aortic root and into the pericardium.

CASE REPORT 4

A 55-year-old, previously fit man developed sudden onset chest tightness associated with nausea, vomiting and dyspnoea. The pain was severe and partially relieved with sublingual glyceryl trinitrate and intravenous morphine. Examination revealed an aortic regurgitant murmur; with normal blood pressures in both arms (117/80 mmHg). 12-lead electrocardiogram showed ST segment depression in leads V5 and V6 and the mediastinum appeared to be slightly widened on chest X-ray. Aortic dissection was considered and confirmed on bedside transthoracic echocardiography. Computed tomography demonstrated a type A dissection of the ascending aorta extending through the aortic valve. A glyceryl trinitrate infusion was administered and the patient was rapidly transferred to the regional cardiothoracic unit where he underwent emergency aortic valve replacement and dissection repair. He was discharged after 6 days.

Treatment

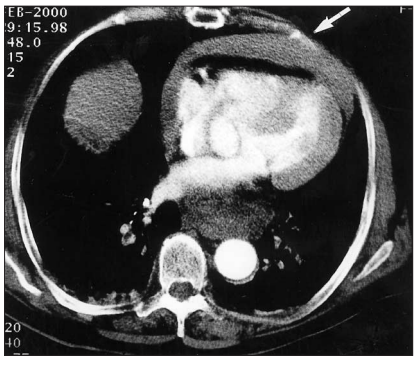
Once the diagnosis is considered and imaging arranged it is essential that hypertensive patients are aggressively treated with hypotensive agents. Untreated hypertension can result in extension of the dissection while awaiting further investigation or transfer. Patients require invasive blood pressure monitoring to allow the safe use of hypotensive agents such as sodium nitroprusside and labetolol. These agents and their infusion details should be readily accessible in accident and emergency departments to prevent further delays.

CONCLUSION

The investigation and management of patients with AAD can be improved if doctors are highly suspicious of AAD in patients presenting with sudden onset thoracic pain and do not simply exclude the diagnosis on the absence of classical findings. The authors recommend a low threshold for imaging the aorta if there is a possibility of AAD with no definite alternative diagnosis.

If clinical suspicion remains high despite a negative investigation a second imaging modality should be requested. It is vital to commence intravenous hypotensive agents in

Figure 2. Arrow shows a large pericardial effusion.



those who are hypertensive while transfer is being arranged. With regards to diagnostic imaging of the aorta agreed local protocols with radiologists, cardiologists and regional cardiac surgeons should be in place. This should go some way to reducing the morbidity and mortality of this catastrophic disease which has such varied presentations. HM

The authors would like to acknowledge the permission of Dr Andrew Macleod to include one of his patients, and thank the cardiac surgeons at Southampton General Hospital for their advice in preparing this manuscript.

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In the Public's View...

Out of sight, out of mind

Some things which are not in the public's view should be. For health care the most important is rationing. I went to a recent conference about rationing, organized by a well-known and reliable body who had assembled excellent speakers at a good venue. The lively audience was drawn from all sections of the medical community.

The day started well. We were told that far from being the universally bad thing that everyone assumes, rationing saved this country during the second world war. Rationing ensures that there is enough to go round. We think that rationing is a modern feature of the NHS, but in the 1960s it was both more prevalent than now and also more explicit.

The NHS has never, from its inception, managed to stay within budget. From about 1980 onwards, the constant response of politicians to the funding difficulties has been to reorganize, which has no effect on the difficulties but gets those particular politicians off the hook until next time. By coincidence, an excellent essay about this, mentioned by one speaker, has just been published (Walshe, 2003).

During all this time, the politicians refused even to utter the word 'rationing', and that remains so. A speaker told the story of Edwina Currie, when the Tories were in power, being challenged on this point. Currie's reply – translated from the vernacular – was that she would rather be re-elected.

Which brings me to why, in the end, I was disappointed in the day. The first couple of speakers established that rationing, by which was meant some limitation of health care from the possible to the affordable, was inevitable. But after that, we slid off into how we could best determine the clinical and economic evidence on which drugs we should use. The question that bubbled under and remained unanswered was how the wider aspects of rationing could be brought into proper public debate, given that politicians will not acknowledge it, and the media use individual hard cases to sell copy.

Doctors don't help by ever-widening the aims of medicine and sometimes not knowing when to stop. The outcome after myocardial infarction in Europe is improving, but will be more than offset by the increased prevalence of smoking, obesity and diabetes – which a medical survey regarded as 'a collective failure of medical practice' (Boersma et al, 2003). Thus we take on the responsibility for running people's lives.

A book about cardiac failure declared that the only definitive treatment was heart transplantation, failed to document why this was unrealistic (or even misguided, see Anyanwu and Treasure, 2003), and did not even have a chapter on palliative care. Another book, about cardiovascular physiology, declared on its first page that the main constraint on gene therapy for cardiac failure was the lack of proper understanding of its pathophysiology, failing even to mention that gene therapy has so far failed to cure anything at all.

The final speaker at the conference was from the Multiple Sclerosis Society. He was blunt. Rationing was inevitable (despite, he said, the continuing denial of its existence by Professor Sir Michael Rawlins, chair of the National Institute of Clinical Excellence), and it was his society's job to get the most for its members. Shout, and you shall be rewarded. HM

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