

## Beware of the migrating chest pain and widened mediastinum: case series on aortic dissection

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**Objective:** Aortic dissection is not a rare life-threatening emergency. Undiagnosed and untreated aortic dissection is associated with a high mortality. A review of cases in the hospital may provide a baseline picture to guide clinical decisions. **Design:** Retrospective case review for a period of 3 years and 4 months. **Setting:** District general hospital near the Hong Kong – Shenzhen 'border' with 24-hour Accident & Emergency service but without cardiothoracic surgical capability. **Population:** All cases coded as 'aortic dissection' in the computerized 'Clinical Management System' of the hospital. **Main outcome measures:** Date, sex, age, history of hypertension, presenting symptoms, pulse deficit, chest X-ray findings, pericardial effusion, A&E diagnosis, type of dissection and patient outcome. **Results:** From August 1998 to November 2001, 26 cases of aortic dissection were identified. There were 19 males and 7 females. Age range was 26 to 90 years (mean  $65.04 \pm SD 15.04$ , median 66.50, mode 65). In the plain chest X-rays, widened superior mediastinum (>8 cm) was present in 19 patients (73.1%) and pleural effusion in three (11.5%). Pericardial effusion was found in six patients (23.1%). As regard to outcome, 11 were discharged home (42.3%), 14 were transferred to cardiothoracic surgical unit (where two subsequently died) and one died in the hospital. **Conclusion:** The prevalence of aortic dissection may be more common than is generally appreciated by emergency physicians. Owing to its variable clinical presentations mimicking other diseases, the diagnosis of aortic dissection is easily missed. Higher clinical vigilance should be exercised for this potentially deadly condition. (*Hong Kong j.emerg.med.* 2002;9:95-101)

**Keywords:** Aneurysm, aortic rupture, chest pain, dissecting, mediastinum

### Introduction

Seven non-traumatic life-threatening conditions may present with chest pain. Their differential diagnoses include acute myocardial infarction, unstable angina, pulmonary embolism, aortic dissection, tension pneumothorax, pericarditis and the Boerhaave's syndrome.

Aortic dissection is not a rare life-threatening emergency. However, the variable clinical presentations of aortic dissection, mimicking other diseases, make its diagnosis difficult. If not recognised early and treated promptly, it is often rapidly fatal.<sup>1,2</sup> A retrospective hospital case review may provide a baseline picture to guide future clinical decisions.

### Materials and methods

North District Hospital is a district general hospital situated near the Hong Kong – Shenzhen 'border' with about 600 beds, serving a population around 300,000. It had four in-patient units of general medicine, general surgery, orthopaedics and paediatrics, but no cardiothoracic surgery. Its Accident & Emergency

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(A&E) department had an annual attendance of around 110,000.

Cases of 'aortic dissection' in a period of 3 years and 4 months from August 1998 (the month of opening of the A&E department of the hospital) to November 2001 were traced from the hospital computer 'Clinical Management System'. The A&E and hospital medical records were retrieved and reviewed. The following data were extracted: date, sex, age, triage category, general condition, systolic blood pressure, diastolic blood pressure, history of hypertension, presenting symptoms, pulse deficit, chest X-ray findings, pericardial effusion, A&E diagnosis, type of dissection, treatment and patient outcome. Descriptive statistics were used for data analysis.

## Results

Twenty-six cases were retrieved (0 in 1998, 4 in 1999, 9 in 2000 and 13 in 2001). Two were known chronic dissecting aneurysms (one with Ehler-Danlos syndrome). The rest were acute aortic dissections (less than two weeks) presenting to the A&E department. There were 19 males (73.1%) and 7 females. Age ranged from 26 to 90 years (mean  $65.04 \pm SD 15.04$ , median 66.50, mode 65). Of the four patients below age 50, one had Marfan's syndrome, one had Ehler-Danlos syndrome and the other two had chronic hypertension. The majority (73.1%) were triaged as category 3 (four in category 2, 19 in category 3 and three in category 4). The overwhelming majority (24 or 93.3%) was regarded as clinically stable. Fifteen patients (57.7%) had history of hypertension. Systolic Blood pressure at presentation to the A&E department ranged from 96-230 mmHg (mean  $158.15 \pm SD 31.99$ , median 158.50). Diastolic blood pressure ranged from 20-140 mmHg (mean  $83.15 \pm 22.77$ , median 84.50). The presenting symptoms are shown in Table 1. Pulse check was recorded in only six patients in the A&E department, and only two had pulse deficit and one had asymmetric blood pressure detected. Widened superior mediastinum ( $>8 \text{ cm}$ )<sup>3</sup> was found in the plain chest X-rays in 19 patients (73.1%). Pleural effusion (Figure 1) was detected in the chest X-ray in only three (11.5%). Pericardial

effusion was detected in six. Provisional diagnoses at the A&E department were shown in Table 2. There were 18 Stanford Type A (69.2%) and eight type B aortic dissections. Thirteen patients had medical treatment, twelve had surgical treatment and one patient discharged against medical advice. As regard to outcome, 11 were discharged home after medical therapy (42.3%), 14 were transferred to cardiothoracic units (one known chronic dissection of another hospital) and one died in our hospital. Two patients were in critical conditions at the time of transfer, and they subsequently died. Hence, the overall mortality rate was 11.5%. All the three patients who died suffered from acute type A aortic dissection. Notably, four patients were initially discharged from the hospital after the acute episode, with an appointment for computed tomography (CT). The diagnosis was made only after the CT scan.

The followings are five interesting illustrative cases:

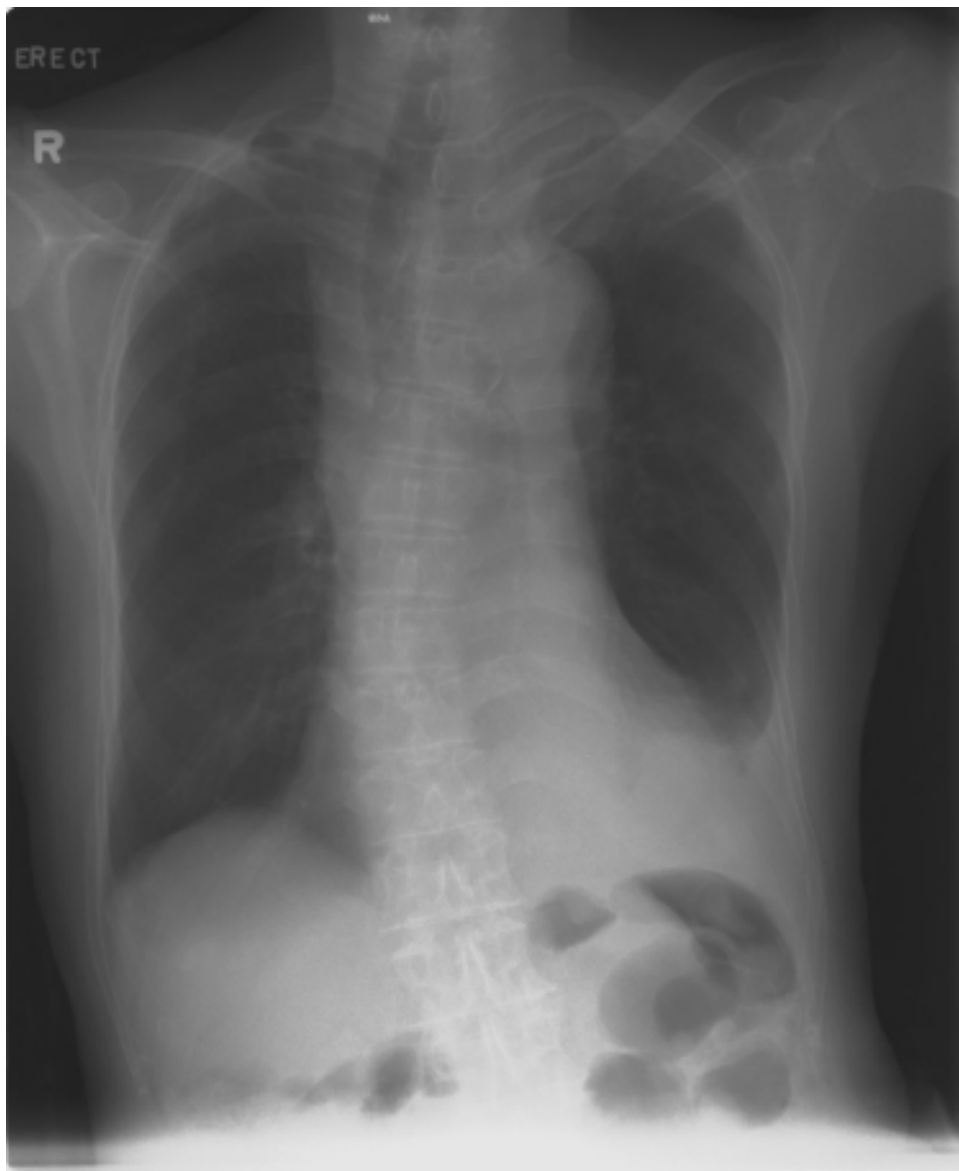
### Case 1

A 58-year-old man with history of hypertension followed up in China. He had an episode of loss of consciousness and was admitted into hospital in China. He developed chest pain, shortness of breath and dizziness. Deep breathing aggravated the chest pain. He returned to Hong Kong three days later for medical care. At the time of presentation at the A&E department, his blood pressure was 138/97 mmHg, pulse rate 135/minute, respiratory rate 32/minute, oral temperature 36°C, and SpO<sub>2</sub> 95% in room air. Erect chest X-ray showed widened mediastinum (10.7 cm)

**Table 1.** Presenting symptoms\* of aortic dissection.

Symptoms	Frequency
Chest pain	12
Back pain	10
Abdominal pain	7
Neck pain	1
Syncope/Dizziness	2
Shock	1
Shortness of breath	2
Paraparesis	1
Hemiparesis ± other neurological deficits	3

\* Patients may have more than one symptom



**Figure 1.** Widened mediastinum (12.3 cm), cardiomegaly, and left pleural effusion in CXR.

**Table 2.** Provisional A&E diagnosis.

A&E diagnosis	Frequency	Percentage	Remark
Chest pain/Acute coronary syndrome	11	42.3	
Suspected aortic dissection	7	26.9	
Acute pulmonary oedema	2	7.7	
Syncope	1	3.8	
Perforated peptic ulcer	1	3.8	
Chronic obstructive airway disease	1	3.8	Dissection incidental
Chest infection	1	3.8	Dissection incidental
Stroke	1	3.8	Dissection incidental
Anemia	1	3.8	Anemia incidental

and cardiomegaly. Electrocardiogram showed sinus tachycardia, left ventricular hypertrophy and diffuse ST elevation with upward concavity. (Figure 2) He was admitted into the medical unit and later transferred to the intensive care unit. Transthoracic echocardiogram showed small pericardial effusion. CT showed aortic dissection from ascending to descending aorta, extending into the pericardium, but the aortic valve was intact. He suddenly developed cardiac arrest and finally succumbed.

### Case 2

A 52-year-old man, mainly living in China, had history of hypertension. He presented with epigastric and back pain, with weakness and numbness of both lower limbs. He was admitted into hospital in China, but discharged himself and returned to Hong Kong for further medical care. At the time of presentation at the A&E department, his blood pressure was 169/70 mmHg, pulse rate 112/minute and tympanic temperature 36.7°C. The femoral pulses were weak. Dorsalis pedis pulses were impalpable. The lower limb perfusion was poor. There was paraplegia of the lower limbs and numbness below the umbilicus. Chest X-ray showed widened mediastinum (sitting, 9.5 cm).

Electrocardiogram showed sinus tachycardia only. Aortic dissection with thrombosis was suspected. He was admitted into the surgical unit. Computed tomography showed aortic dissection from the arch down into the descending aorta. He was later transferred to cardiothoracic surgeons of another hospital for further management. However, he died on the next day.

### Case 3

A 58-year-old man, mainly living in China, complained of back pain after sex with his wife. He also had epigastric pain. He returned to Hong Kong for medical care. At the time of presentation at the A&E department, his blood pressure was 168/92 mmHg, pulse rate 96/min, and oral temperature 37.2°C. The right femoral pulse was weak. There was tenderness and guarding over the epigastrium. There was no widened mediastinum (sitting, 7.6 cm) or free gas under the diaphragm in the chest X-ray. He was admitted into the surgical unit, with a provisional diagnosis of perforated peptic ulcer. CT scan and transesophageal echocardiography (TEE) confirmed Stanford type B aortic dissection. He was put on antihypertensive medication and later discharged from hospital.

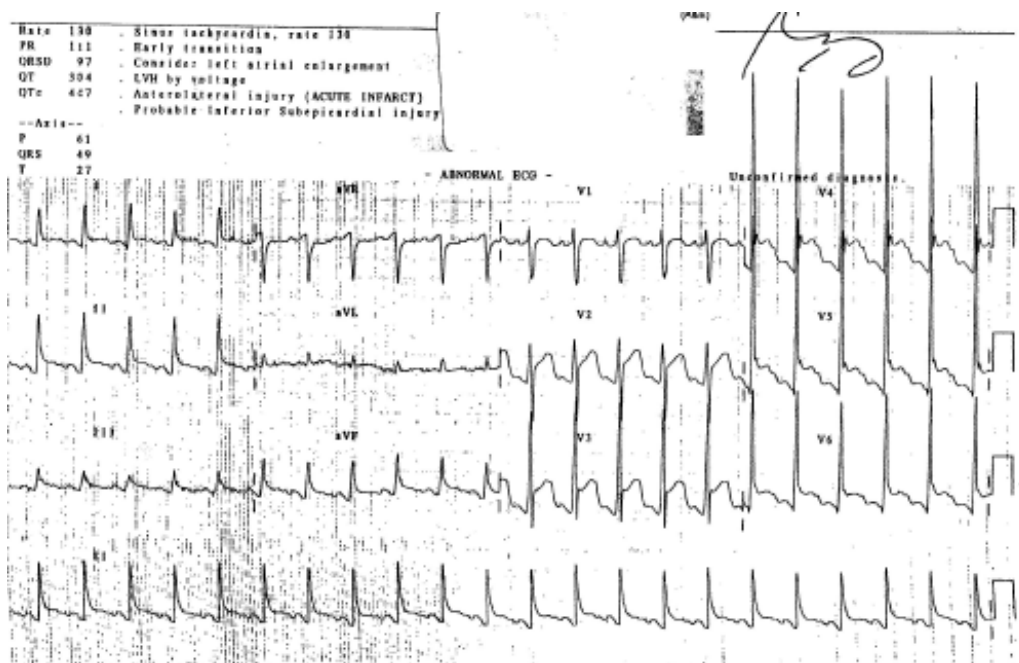


Figure 2. Diffuse ST elevation in ECG (Case 1).

#### **Case 4**

A 26-year-old woman, suffering from Marfan's syndrome with known aortic root aneurysm, first presented to the A&E department complaining of severe low back pain, chest pain and epigastric pain. Blood pressure was 133/43 mmHg, pulse rate 81/minute, oral temperature 36.2°C and SpO<sub>2</sub> 100% in room air. There was tenderness at the epigastrium. There was no pulse deficit. The mediastinum was not widened in the chest X-ray (erect, 6.9 cm). Electrocardiogram was normal. She was discharged after an analgesic injection. She re-attended the next day because of persisting epigastric pain. The physical examination was essentially unchanged. She was admitted into the medical unit. CT scan revealed aortic dissection from the aortic root down to the renal arteries. She was transferred to the cardiothoracic surgeons for surgical management. Operation was successful.

#### **Case 5**

A 73-year-old man, with history of hypertension and ischaemic heart disease, presented with severe chest pain for half-an-hour. He also had dyspnoea and sweating. There was weakness of the right side of the body. At the time of presentation at the A&E department, the blood pressure was 196/140 mmHg, pulse rate 97/min, and tympanic temperature 35.6°C. Erect chest X-ray showed widened mediastinum (10.4 cm). Electrocardiogram showed left ventricular hypertrophy only. He suddenly developed hypotension in the A&E department, with blood pressure 50/30 mmHg. Transthoracic ultrasound revealed pericardial fluid. He was admitted into the intensive care unit. Transesophageal echocardiogram and CT scan revealed aortic dissection from the root of ascending aorta to the descending aorta with pericardial and right pleural fluid. He responded to resuscitation and was transferred to a cardiothoracic hospital. However, he died four days later.

## **Discussion**

Aortic dissection is the most common catastrophe involving the aorta. It has been estimated to be two to three times commoner than rupture of abdominal

aortic aneurysm.<sup>2,4</sup> The incidence in our hospital was similar to other reported series.<sup>5,6</sup>

Aortic dissection is an expanding intramural haematoma resulting from the separation of the layers of the media wall of the aorta by blood, usually with a tear in the intima, and producing a false channel with variable proximal and distal extension. Rarely, aortic dissection may demonstrate medial haematoma and no intimal tear.<sup>7</sup> The dissection may extend into and interrupt the blood supply to branch arteries. The thin outer wall may rupture into the pericardial or pleural space. The term 'dissecting aortic aneurysm' has been inaccurately applied to this entity, because the affected aorta is uncommonly aneurysmal.<sup>7</sup> Owing to its variable clinical presentations mimicking other diseases, the diagnosis of aortic dissection is easily missed. If not recognised early and treated promptly, death will ensue. The mortality in the first 24 to 48 hours of aortic dissection is between 1-2% per hour.<sup>7,8</sup> Approximately 75-80% untreated aortic dissection die within two weeks of the onset of symptoms, rupture being the commonest cause of death.<sup>4,7</sup> As a result, aortic dissection is classified as acute if it is less than two weeks and as chronic if more than two weeks. The two common anatomical classifications are shown in Table 3. The ratio of Stanford type A to type B is about 2:1,<sup>7</sup> as in our cases.

Medial degeneration appears to be a prerequisite for the ultimate propagation of a dissection. A variety of congenital and acquired conditions lead to medial degeneration, with aging and hypertension being the two most common factors.<sup>6,9</sup> From 70% to 95% of patients with aortic dissection have hypertension and hypertension accelerates the natural medial degenerative changes that occur with aging. Inherited connective tissue disorders, especially Marfan's and Ehlers-Danlos's syndromes, are characterized by medial degeneration predisposing to dissection. Iatrogenic vascular trauma such as cardiac catheterization and cardiac surgery, may occasionally lead to aortic dissection. Other predisposing factors include coarctation of the aorta, bicuspid aortic valve, aortic stenosis, pregnancy, and cocaine use. There is a male predominance of about 3:1, with usual ages between 50-70 years<sup>5,6,10</sup> – as in our cases.

**Table 3.** Anatomical classifications of aortic dissection and their treatment.

Site	DeBakey	Stanford	Treatment of acute dissection
Ascending + arch ± descending	Type I	Type A	Surgical
Ascending only	Type II	Type A	Surgical
Descending: Thoracic only	Type IIIa	Type B	Uncomplicated – Medical Complicated/Treatment failure – Surgical
Descending: Thoracic + abdominal	Type IIIb	Type B	Uncomplicated – Medical Complicated/Treatment failure – Surgical

Sudden severe chest pain radiating to the back is the most common presenting symptom in aortic dissection,<sup>6</sup> so common (>90%), in fact, that one should be careful of making the diagnosis of aortic dissection when these symptoms are not present, unless the patient is unconscious or neurologically impaired.<sup>7</sup> The onset is abrupt, dramatic and often described as 'tearing' in character. The pain is excruciating and most severe at onset, in contrast to the crescendo-type of intensity in myocardial infarction.<sup>7</sup> One special characteristic is that the pain may migrate as the dissection propagates along the aorta. Pain confined solely to the precordial region usually signifies dissection of the ascending aorta. Pain radiating to the neck and jaw implies involvement of the aortic arch, while that in the interscapular area and back often signifies distal dissection. Pain in the lumbar area and abdomen implies dissection below the diaphragm.

As the dissection propagates, blood supply to major branch arteries may be interrupted. This results in its variable clinical presentations mimicking other diseases, and adds to the difficulty in its diagnosis. Occlusion of coronary branches may result in myocardial infarction, arch branches in stroke or syncope, intercostals in paraplegia, renal branches in anuria and iliac branches in lower limb ischaemia. Unequal pulses or asymmetric blood pressure had been claimed to be present in 50% of cases,<sup>7,10,11</sup> but these were poorly recorded in our A&E records. Aortic dissection is an important differential of myocardial infarction to be excluded since the administration of thrombolytics in this setting will have catastrophic consequences.<sup>4</sup> Blood may also rupture into the periaortic tissue, pericardial, pleural or peritoneal space, resulting in acute aortic valve insufficiency, acute heart failure, cardiac tamponade, shock or even

sudden death. The above findings were well illustrated in our cases, such as the diffuse ST elevation in case 1 simulating pericarditis. As can be seen, only about a quarter of the cases was suspected at the A&E department. In addition, clinical symptoms and signs alone correlate poorly with the site of origin and extent of the dissection.<sup>2,6</sup>

Chest X-ray is usually the first investigation performed.<sup>1,8</sup> It may be the first hint suggestive of aortic dissection.<sup>5,6</sup> A common but not pathognomonic finding is a widened superior mediastinum and aorta (>8 cm)<sup>3</sup> which occurs in 75%, as in our series. Widening of the superior mediastinum is difficult to evaluate, as most patients will be too ill to hold their breath in full inspiration or to go through the ordeal for an erect postero-anterior exposure.<sup>2</sup> It was shown that magnification on supine chest films was minimal at the level of the tracheal bifurcation.<sup>12</sup> Comparison with previous chest X-rays is often helpful in evaluating changes in the configuration of the aorta.<sup>2,5,6</sup> Cardiomegaly is also common but non-specific.<sup>2</sup> Rare chest X-ray findings such as pleural effusion (usually on the left) and intimal calcification separated more than 6 mm from the edge of the aorta (calcium sign) may be helpful but their low frequencies make them less useful. Up to 20% of patients with dissection may not have abnormal chest X-ray findings, and so a negative X-ray does not exclude the presence of dissection.<sup>1,2,5</sup> Plain chest radiographic features alone has a sensitivity of 81%, a specificity of 75%, and an accuracy of 78%.<sup>13</sup> Similar plain film findings in patients without a history suggestive of aortic dissection are less reliable because these radiological signs can be simulated by other disease processes such as elongation and tortuosity of the aorta, especially in elderly hypertensive patients.<sup>5</sup> In summary, chest X-ray is a valuable, though non-specific, first

investigation.<sup>1</sup> The superior mediastinum should always be assessed in patients with chest pain. Transthoracic ultrasound examination can assess for haemopericardium, aortic valvular and ventricular function,<sup>1</sup> and may be helpful at the A&E department. Evidence of pericardial or pleural fluid implies poor prognosis as it suggests rupture into the pericardial or pleural spaces.<sup>2</sup> Other more definitive investigations include aortography, spiral CT scan, trans-oesophageal echocardiography and magnetic resonance imaging.

The most important prognostic factor is involvement of the ascending aorta, irrespective of the site of the primary intimal tear or the distal extent of the dissection. As a result, most patients with acute type A dissections are treated surgically. The surgical risk has decreased considerably in recent years, with surgical mortality below 10%.<sup>2</sup> As less than 10% of untreated patients survive more than two weeks, chronic type A dissections are uncommon. Patients with chronic aortic dissection have already survived their period of highest mortality risk and are usually treated medically.<sup>7</sup> As a result, acute type B and all chronic dissections are treated medically unless there is evidence of treatment failure or major complications.<sup>2</sup>

In conclusion, the prevalence of aortic dissection may be more common than is generally appreciated by emergency physicians. Its incidence seemed to be rising, but it might only be the result of the better diagnosis coding by clinicians of our hospital. Increased diagnosis might also be the result of our aging population, higher clinical vigilance and better medical technology especially with the widespread use of Spiral CT and TEE.<sup>2</sup> Several clinical syndromes should alert emergency physicians on the diagnosis of aortic dissection:<sup>7</sup> -

1. Migrating pain that progresses over hours or days from chest to neck, to back or to the abdomen.
2. Chest pain with concomitant neurological deficits.
3. Chest pain with unequal pulses or asymmetric blood pressure.
4. Chest pain with widened superior mediastinum in the chest X-ray.

## Limitation

As this was a retrospective review, there was always a chance of bias due to missing information. Cases might not have been retrieved if the diagnosis coding had not been entered by the clinicians concerned. Clinical data might be inadequately recorded, especially with history of hypertension and differential blood pressure or pulse recording.

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