Unusual association of diseases/symptoms

Chest pain, dyspnoea and elevated D-dimer in a recent air traveller

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Summary

A previously asymptomatic 69-year-old lady, who recently travelled on a 4 h flight, presented with acute left-sided pleuritic pain, dyspnoea and calf pain. Blood gases revealed hypoxaemia and D-dimer was significantly elevated. She also had low-grade fever, leukocytosis and a small left-sided pleural effusion on chest x-ray. The working diagnosis was pulmonary embolism and chest infection and she received low molecular weight heparin and antibiotics. A subsequent CT pulmonary angiogram ruled out pulmonary embolism but revealed an abnormal finding in the ascending aorta, suggestive of a penetrating aortic ulcer. Urgent transoesophageal echocardiography was consistent with an intramural haematoma and the patient underwent emergency aortic root replacement with imminent aortic rupture confirmed at surgery. This case highlights the fact that acute aortic syndromes may have atypical presentations and also emphasises the fact that D-dimer levels are elevated in aortic syndromes.

BACKGROUND

'Common things are common' but occasionally what seems to be a definite diagnosis may be incorrect. D-dimer is a breakdown of thrombus and may be elevated in conditions other than acute pulmonary embolism. In fact, D-dimer levels are significantly elevated in acute aortic syndromes associated with intramural haematoma. Aortic dissection is not the only acute aortic syndrome and some patients may present with very atypical symptoms. Urgent specialist consultation should be requested in suspicious cases.

CASE PRESENTATION

A 69-year-old, previously fit and healthy, female presented with a 2 day history of acute right calf pain and sharp left-sided chest pain, associated with progressive worsening shortness of breath. The chest pain was pleuritic in nature, the patient describing it as sharp and worse on inspiration. The chest pain did not radiate. She had a history of flu-like symptoms, with the onset coinciding with a recent 4 h flight from a beach resort. She had no major medical history apart from recently diagnosed mild hypertension. She smoked 15 cigarettes daily and was not on any regular medication. Family history was unremarkable.

On examination, she had a tachycardia of 120/min, blood pressure was 139/81 mm Hg, initial temperature was normal at 37.2 °C but later she developed a low-grade fever. She was tachypnoeic with a respiratory rate of 24/min and pulse oximetry was consistent with mild hypoxia with a saturation of 94% on room air. Clinically, she appeared to be mildly short of breath but was able to complete full sentences. The cardiovascular examination was essentially unremarkable, jugular venous pressure was not raised and there were no murmurs. Pulses were equal and there was no radio-radio delay or blood pressure differential between the two arms. She had mild tenderness of the

right calf but no obvious swelling, heat or other signs of venous thrombosis.

INVESTIGATIONS

Inflammatory markers were raised with a C-reactive protein of 168 mg/dl (normal <5 mg/dl). White cell count was 15.9 (normal 4–11 $10^9/L$) with neutrophils of 13 (normal 2–7.5 $10^9/L$). Haemoglobin was 11.7 g/dl (normal 13–18g/dl) and D-dimer of 1849 (normal 0–200 ng/ml).

Arterial blood gas on room air showed a pH of 7.49; pCO₂: 4.6 kPa; pO₂: 7 kPa; Sat: 93%; base excess: 2.7; HCO₃: 26.8; lactate: 0.8. The ECG was normal and the chest x-ray revealed cardiomegaly and a small left-sided pleural effusion.

Other routine laboratory tests including electrolytes, hepatic and renal function tests were all normal.

DIFFERENTIAL DIAGNOSIS

The initial working diagnosis was of an acute pulmonary embolism and /or left-sided chest infection.

TREATMENT

The patient was managed initially on the medical assessment unit as a case of probable acute pulmonary embolism with a possible chest infection. She was commenced on full treatment dose tinazeparin (175 mg/kg sc) and was also given oral antibiotics in the form of amoxicillin. Immediate access to CT pulmonary angiography (CTPA) was not available and as the patient was haemodynamically stable, therapy was initiated.

CTPA was performed 36 h after hospital admission and showed normal pulmonary arteries with no evidence of thrombi but did reveal a significant pericardial effusion and bilateral pleural effusions (more marked on the left side). There was also evidence of moderate pulmonary

Figure 1 (A) Chest x-ray showing cardiomegaly and a small pleural effusion. (B) CTPA image showing ascending aorta with out pouching (yellow arrow) consistent with a penetrating aortic ulcer. (C) Transoesophageal echo mid oesophageal 120 degree view demonstrating intramural haematoma on anterior wall of aorta (yellow arrow). (D) Transoesophageal echo mid oesophageal position with short axis view of ascending aorta just above aortic valve with intramural haematoma (yellow arrow).

emphysema. An unusual appearance was noted in the ascending aorta but opacification of the remainder of the aorta was normal. There was no evidence of an aortic dissection flap. The results of the CTPA were relayed to the patient's managing physicians who requested an immediate cardiology review. The cardiology consultation raised the possibility of an acute aortic syndrome with a penetrating aortic ulcer and further imaging was performed (figure 1).

Immediate transthoracic echo images confirmed a 2 cm pericardial effusion without signs of tamponade. Left ventricular function was normal and there was no aortic valve regurgitation. Some non-specific thickening of the immediate ascending aorta was noted. Transoesophageal echocardiography (TOE) was performed within 1 h of consultation and demonstrated an intramural haematoma of the anterior wall of the ascending aorta extending from just above the aortic valve. The aortic arch and descending thoracic aorta appeared normal and there was no evidence of any dissection flap (figure 1). At this stage, the patient had the working diagnosis of an acute aortic syndrome in the form of a large intramural haematoma of the ascending aorta arising from a probable penetrating aortic ulcer. There was a moderate pericardial effusion which was felt to be a sympathetic effusion. She was immediately commenced on anti impulse therapy in the form of intravenous labetalol and transferred to a regional cardiothoracic centre for immediate surgery.

Emergency surgery was performed shortly after transfer and within 5 h of the cardiology consultation. The surgical findings confirmed the diagnosis of a large intramural haematoma of the ascending aorta in association with a

penetrating aortic ulcer. There was no evidence of a dissection and the aortic valve was normal. There was a serous pericardial effusion. However, there were signs of imminent aortic rupture with areas of haematoma extending to the adventitial surface. The ascending aorta was replaced with a graft.

OUTCOME AND FOLLOW-UP

The patient made an uneventful recovery and was discharged well. She remains well at follow-up 6 months postoperatively.

DISCUSSION

Acute aortic dissection is the commonest acute aortic syndrome (AAS). A DeBakey type I dissection involving the ascending aorta can be rapidly fatal. The classic presentation of acute rendering chest pain is familiar to acute physicians but many cases may have an atypical presentation. An AAS may present as a myocardial infarction when an intimo-medial flap extends into the coronary ostium causing coronary occlusion, ECG changes and raised cardiac markers, including cardiac troponin. Here we describe a patient who presented with a clinical picture similar to a pulmonary embolus and who actually had an AAS with impending aortic rupture. High clinical suspicion is required to avoid the administration of thrombolytic or anticoagulation which has potential detrimental/harmful effects in the presence of an AAS¹. Early diagnosis is critically important as emergency surgery may help transform what is otherwise a dismal prognosis. It is estimated that up to 40% of AAS

including dissections may have delayed diagnosis due to atypical clinical presentation.^{2–4} For this reason, it is important to stress clinical vigilance and awareness of abnormal D-dimer levels in the absence of pulmonary embolism.^{3,4}

D-dimer is a breakdown product of thrombus. With an acute aortic dissection where there is an intramural haematoma, D-dimer levels will be raised. In fact D-dimer levels are characteristically five to ten times higher than in other conditions, including acute pulmonary embolism, in the first 6 h of symptom onset.³ D-dimer levels in acute aortic dissection are comparable to levels seen in acute pulmonary embolism beyond the first 6 and up to 24 h. A normal D-dimer level in fact has a strong negative predictive value in excluding an acute aortic dissection.^{3,4}

Our knowledge of AAS has increased in recent years, particularly through easier access to advanced imaging with CT, MR and TOE. In addition to classic dissection, cases of penetrating aortic ulcer, intramural haematoma and more recently incomplete dissection are being recognised. These different syndromes are part of a spectrum of pathology that can affect the aorta and one entity may potentially progress to another or even to aortic rupture. It is important to be aware that there is no dissection flap seen in cases of intramural haematoma or penetrating aortic ulcer. This fact can mislead radiologists and other physicians into a false sense of security with potentially disastrous consequences.

Penetrating aortic ulcers generally occur in the descending thoracic aorta where there is a high burden of atherosclerosis. They are most commonly described in older males with hypertension. It is assumed that an atheromatous plaque ulcerates into the aortic lumen but that development of a dissection flap or intramural haematoma is prevented by the amount of degenerative disease in the aorta. Penetrating aortic ulcers have been described in the ascending aorta, such as in our patient, but are less common.

It is well established that acute aortic syndromes involving the ascending aorta (Stanford type A) needs urgent surgical repair. Furthermore, there is evidence to suggest that we ought to differentiate between intramural haematoma with or without a penetrating aortic ulcer, as these progress differently and may potentially be managed in different ways. In this setting the presence of ongoing chest pain and a pleural effusion have been found to be independent predictors of disease progression. Hence urgent surgical repair should also be sought when an intramural haematoma is associated with a penetrating aortic ulcer, (regardless if the lesion is in the ascending or descending aorta), the patient

has ongoing chest pain or a pleural effusion is increasing in size⁵.

In this patient, CT had been performed as a CTPA for a presumed pulmonary embolus with a penetrating aortic ulcer in the ascending aorta discovered as an incidental finding. CTPA with multislice CT is well established in the diagnostic workup of patients with suspected pulmonary embolism.⁶ In practice, CTPA examinations may not be always be immediately available and patients are often started on anticoagulation based on clinical suspicion and D-dimer levels. This could potentially be disastrous in the setting of an acute aortic syndrome.

Learning points

- ▶ D-dimer levels are elevated in acute aortic syndromes including acute aortic dissection and intramural haematoma.
- ▶ Classic aortic dissection is not the only form of acute aortic syndrome and it is important to consider other variants including penetrating aortic ulcer and intramural haematoma.
- ▶ Many cases of acute aortic syndrome may present in an atypical fashion and one should always have a high index of suspicion as early diagnosis may be life saving.
- ▶ Anticoagulation may have disastrous consequences in the setting of an acute aortic syndrome and it is important to consider this possibility in the differential diagnosis of acute coronary syndromes and pulmonary embolism.

Competing interests None.

Patient consent Obtained.

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