Aortic dissection: an unusual presentation

Introduction

Aortic dissection classically presents acutely with severe chest pain. Rarely, chronic dissection can exhibit atypical features, including persistent fever and malaise. This article describes a case where the patient presented with fever and bilateral pleural effusions associated with chronic dissection of the aorta that was also misdiagnosed on computed tomography of the aorta. This case outlines the difficulty in diagnosing aortic dissection secondary to a circumferential tear of the intima, whereby imaging modalities can indicate a perforated supravalvular aortic membrane rather than dissection. The case also shows the importance of fever and pleural effusions as indicators of pathology of the thoracic aorta.

Discussion

Aortic dissection is a life-threatening condition that is often a diagnostic challenge. Patients are usually in their sixth or seventh decade and classically present with acute onset severe chest or back pain. However, it can also occur in younger patients and can present chronically with symptoms lasting >2 weeks.

In the acute setting, up to one third of patients may exhibit fever (Hirst et al, 1958); however, prolonged pyrexia and generalized malaise may rarely dominate the clinical picture in chronic dissection, thus suggesting infectious, neoplastic or autoimmune conditions, as with the patient in this report and a few cases in the literature (reviewed by Schattner et al (1996) and Gorospe et al (2002)).

It has been suggested that the systemic presentation is secondary to a subacute inflammatory response following continuing dissection and haematoma formation. Alternatively, it occurs secondary to cytokine release following ischaemic damage to the tissues (Schattner et al, 1996).

In the current case, fever and upper and lower respiratory tract symptoms prevailed, suggesting a respiratory tract infection, while the presence of murmurs in the setting of fever suggested associated endocarditis. Interestingly, bilateral pleural effusions were noted. In a study by Schattner and Klepfish (2012), pyrexia of unknown origin together with pleural effusion (predominantly left-sided) suggested pathology of large thoracic arteries. Pleural effusion was also noted in cases where aortic dissection had an atypical presentation with protracted course (Schattner et al, 1996; Yuan et al, 2009).

Various thoracic aortic pathologies can mimic aortic dissection, e.g. intramural haematoma, penetrating aortic ulcer and localized intimal tear without dissection. The vast majority of intimal tears in type A aortic dissection are transverse, sparing more than half of the aortic circumference (Dimarakis et al, 2007). Only a few cases of circumferential intimal tear have been reported (Shah et al, 2006). Similarly to the current case, Dimarakis et al (2007) reported a patient in whom all imaging modalities suggested the presence of a perforated membranous type supravalvular aortic stenosis, with a definite diagnosis of aortic dissection secondary to a circumferential tear only being made intraoperatively.

Useful imaging modalities in the diagnosis of aortic dissection are computed tomography, magnetic resonance imaging and transoesophageal echocardiography, all of which have high sensitivity and specificity. Magnetic resonance imaging is

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Case Report

A 39-year-old hypertensive man presented to the emergency department with generalized malaise and a non-productive cough. He denied associated dyspnoea or chest pain. A week before, he had presented to the emergency department with sudden onset jaw and pharyngeal pain. Following chest X-ray, cardiac markers and ear, nose and throat review, tonsillitis was diagnosed.

During this second presentation, he was tachycardic (100 bpm) and febrile (99.4°F). Arterial blood gases revealed hypoxia (pO_2 57.1 mmHg). Physical examination revealed dullness and decreased breath sounds in the right base together with ejection systolic and early diastolic murmurs. He did not exhibit any phenotypic features of Marfan's syndrome. Chest X-ray showed infiltration in the right lower lung lobe with possible nodules in the right and left upper zones. Biochemical investigations excluded vasculitis. The patient was treated with intravenous antibiotics.

A transthoracic echocardiogram showed the possibility of a supravalvular membrane with severe aortic regurgitation and a dilated left ventricle, but no vegetations (*Figure 1*). Computed tomography angiogram of the aorta excluded aortic dissection and reconfirmed inflammatory changes of the lungs and extensive bilateral pleural effusions. Consequently, a transoesophageal echocardiogram showed a thin mobile membrane above the left main orifice with a central opening (*Figures 2* and *3*). The differential diagnoses were supravalvular membrane and aortic dissection. The patient recovered with conservative treatment and was discharged home.

Repeat transthoracic echocardiogram 3 months later showed severe aortic regurgitation and dilated aortic root and ventricles. Repeat computed tomography angiogram 4 months after the initial presentation showed type A aortic dissection confined to the ascending aorta, bilateral pleural effusions, lung congestion and a pericardial effusion. Surgery was performed 2 days later. A circumferential tear in the intima producing two flaps was noted. Resection and re-suturing of the ascending aorta, and aortic valve replacement were performed. Histology revealed cystic medial necrosis of the aorta consistent with Marfan's syndrome, while the aortic valve showed focal myxoid degeneration. The patient made a good recovery. Repeat transthoracic echocardiogram 2 months after surgery showed improvement in left ventricular dimensions and function.

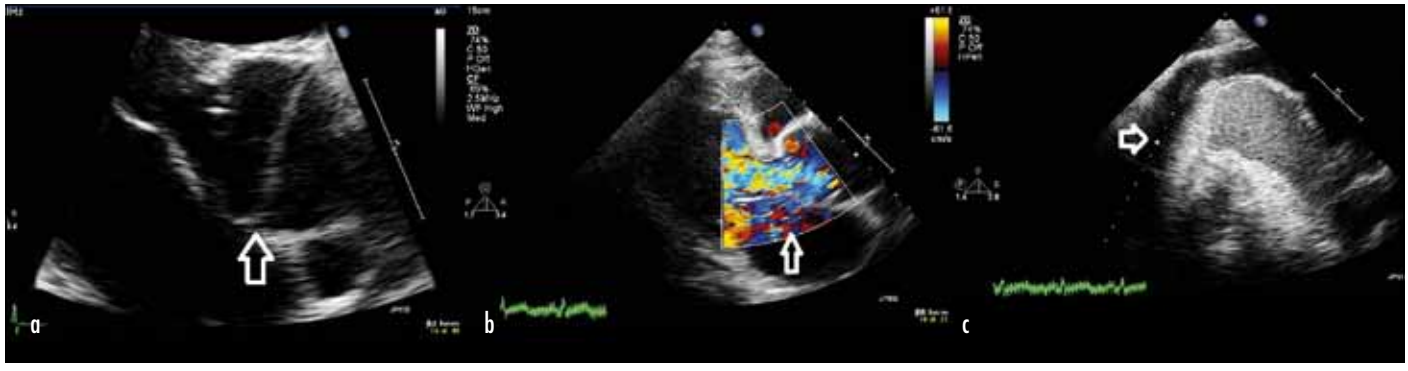

Figure 1. Transthoracic echocardiogram views. a. Dissection flap (indicated by arrow) is seen extending across the aortic root above the aortic valve. b. Turbulence is seen across the left ventricular outflow tract (arrow) in the parasternal long-axis view. c. Pleural effusion (arrow) as seen with echocardiogram.

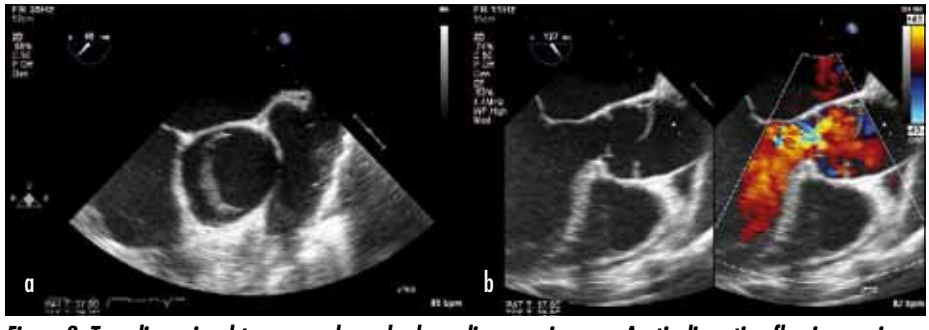

Figure 2. Two-dimensional transoesophageal echocardiogram views. a. Aortic dissection flap is seen in cross-section in aortic valve short-axis view. b. Dissection flap with associated turbulence across it is seen in the long-axis view.

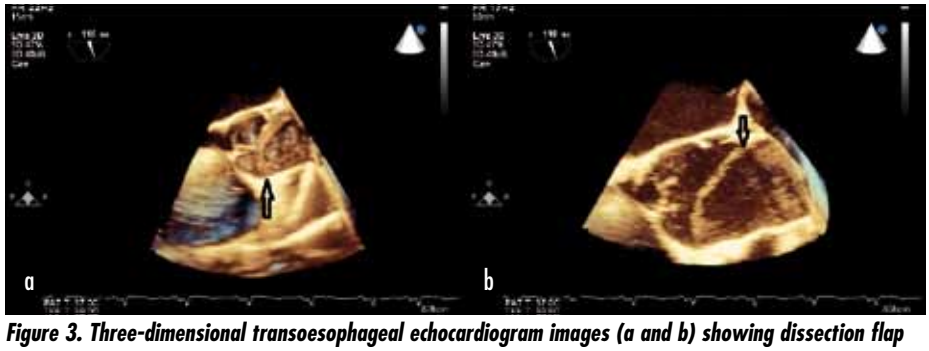

Figure 3. Three-dimensional transoesophageal echocardiogram images (a and b) showing dissection flap (indicated by arrow) above the aortic valve.

probably best for diagnosis of chronic aortic dissection and the most appropriate technique for preoperative evaluation (Deutsch et al, 1994). Nonetheless, magnetic resonance imaging is not widely available and cannot be used on haemodynamically unstable patients. Furthermore, the diagnosis of circumferential tears may still be missed with cardiac magnetic resonance imaging, as in the patient described by Dimarakis et al (2007).

Conclusions

This case highlights the importance of maintaining high clinical suspicion of aortic dissection in the presence of pyrexia of unknown origin, especially with a history of hypertension, in association with pleural effusion(s), despite negative imaging results. Close follow-up of the patient is necessary to avoid misdiagnosis of this potentially lethal condition. BJHM

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LEARNING POINTS

- Chronic aortic dissection can present atypically, including pyrexia of unknown origin and generalized malaise.
- With a circumferential tear of the intima, imaging modalities can indicate perforated supravalvular aortic membrane rather than aortic dissection. In this case, a definitive diagnosis can only be made intraoperatively.
- Fever and pleural effusion are indicators of pathology of large thoracic arteries. The clinician should have a high index of suspicion for possible aortic dissection in the presence of pyrexia of unknown origin, especially if there is a history of hypertension, in association with pleural effusion(s).