Torrential aortic regurgitation: a first presentation of an old disease

Introduction

A 62-year-old, previously healthy man presented with dyspnoea. His examination findings were in keeping with congestive cardiac failure secondary to severe aortic regurgitation, which was confirmed on echocardiography. A comprehensive assessment was carried out to identify the aetiology of his aortic regurgitation, and in view of his sexual history cardiovascular syphilis was considered and diagnosed. The patient was treated with benzylpenicillin and referred for aortic valve replacement, but he turned down the procedure and was treated conservatively for congestive heart failure. He died 1 year after diagnosis.

This case highlights the importance of identifying the aetiology of aortic regurgitation in cases where early treatment can prevent irreversible damage.

Discussion

Golden et al (2003) state that since the advent of penicillin, cardiovascular syphilis has become exceedingly rare, with early treatment of disease successful in preventing progression to tertiary syphilis. Cardiovascular syphilis may also be under-diagnosed; a post-mortem study by Roberts et al (2009) found that out of 90 patients with morphological findings of syphilitic aortitis, evidence of clinical aortic regurgitation or heart failure was reported in only 26% and 31% of patients respectively.

Clinical manifestations of cardiovascular syphilis range from chest pain secondary to rapid expansion of an aortic aneurysm to dyspnoea secondary to heart failure. Saraiva et al (2010) describe a case of a 48-year-old man who presented with dyspnoea and was found to have aortic regurgitation. They proceeded with aortic valve replacement; perioperatively the aortic root was noted to be severely damaged and lymphocytic infiltrate reported on histopathology. Because of a history of promiscuity, diagnostic tests for syphilis were taken which returned positive and the patient required a course of benzylpenicillin postoperatively.

Cardiovascular syphilis has also been associated with the formation of coronary ostial stenosis. Tanaka et al (2016) describe a case of acute myocardial infarction as a result of bilateral coronary ostial stenosis that was also complicated by aortic regurgitation. The patient required a coronary artery bypass as well as an aortic valve replacement. *Treponema pallidum* haemagglutination test levels were taken and found positive. Coronary angiography of this patient also

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CASE REPORT

A 62-year-old man presented to accident and emergency with increasing dyspnoea on exertion over the past 5 months. This had become significantly worse in the previous week. He also reported orthopnoea, but denied any episodes of chest pain or cough. He described a history of significant weight loss over the past few months, with no reported fever, chills or rigors.

The patient was a heavy smoker, but he had no respiratory problems in the past. He did not report any relevant medical history and denied illicit drug use. He was a single, homosexual man who lived alone. There was no family history of cardiovascular problems.

He was hypoxic on room air with saturations of 93%. He was tachycardic at 110 beats per minute and blood pressure readings revealed a wide pulse pressure (145/50 mmHg).

On inspection the patient had gross finger clubbing. Chest examination revealed crepitations throughout with reduced air entry at the bases. On auscultation of the precordium, an ejection systolic murmur was heard followed by a soft S2 and an early diastolic murmur. Abdominal examination was normal and there was no peripheral oedema.

Routine blood investigations were normal apart from a microcytic anaemia. There was no significant rise in two 6-hour troponins. Electrocardiography showed a sinus tachycardia and chest X-ray confirmed congestion of the lung fields with small bilateral pleural effusions.

Echocardiography revealed a dilated left ventricle with an end-systolic diameter of 50 mm (58 mm in end-diastole), global left ventricle hypokinesia and an ejection fraction of 25%. *Figure 1* illustrates the apical 5-chamber view on echocardiogram in colour Doppler mode, identifying the culprit problem as aortic regurgitation – with free blood flow from aorta to left ventricle during diastole. *Figure 2* depicts the continuous wave Doppler values, calculating a pressure half-time (P½T) of 83 ms in keeping with severe aortic regurgitation. Degenerative changes of the aortic valve were noted with a dilated aortic root (41 mm at the annulus) but no vegetations seen.

On angiography, an aortogram confirmed echocardiographic findings with severe, torrential aortic regurgitation filling the entire left ventricle. Coronary angiography showed a totally occluded ostial right coronary artery, which filled well from collaterals from the left anterior descending coronary artery. The left coronary artery showed no significant lesions.

Further investigation into the cause of aortic regurgitation was sought. In view of his sexual history he was screened for HIV and syphilis. HIV serology was negative, but *Treponema pallidum* haemagglutinin titres were positive (>1:10 240) and syphilis IgM also returned positive. It was concluded that the patient was suffering from cardiovascular syphilis resulting in severe valve degeneration and was treated with intramuscular benzylpenicillin administered once weekly over 3 weeks.

Following treatment for syphilis, he was referred to cardiothoracic surgeons for aortic valve replacement. However, following review, the patient turned down surgical treatment because of the risks of the procedure. Follow up echocardiography showed little improvement in ejection fraction and he died within 1 year of diagnosis.

Figure 1. Aortic regurgitation as seen on apical 5-chamber view on echocardiogram in colour Doppler mode.

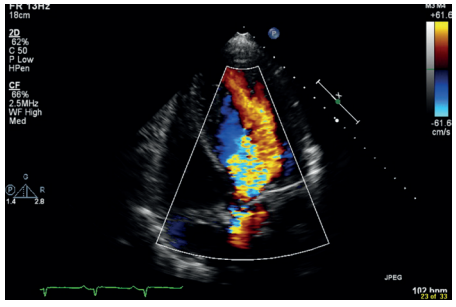
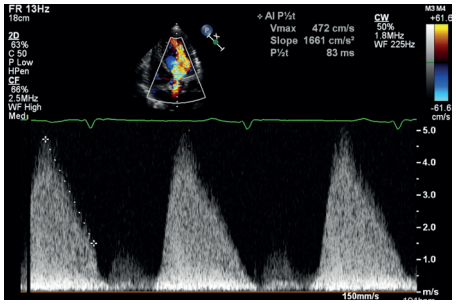


Figure 2. Continuous wave Doppler with pressure half-time of 83 ms – quantifying aortic regurgitation as severe.



noted an ostial stenosis at the right coronary artery with left-sided arteries unobstructed.

Cardiovascular syphilis should be considered as a diagnosis in new onset aortic regurgitation, especially when present in combination with coronary ostial stenosis. A simple blood test can provide a timely diagnosis and early treatment with antibiotics and/or aortic valve replacement will help prevent accelerated aortic regurgitation with irreversible damage and subsequent heart failure. Unfortunately this patient presented late, when degeneration of the aortic root and valve was extensive. The patient refused surgery because of procedural risk and despite diagnosis and treatment of syphilis, he died. BJHM

LEARNING POINTS

- New cardiac valve pathologies should be investigated for an underlying cause before changes are considered to be age-related. A thorough history is needed to guide the clinician in investigation and an open mind must be kept to include rare causes in the differential diagnosis.
- At times, rare pathologies can be easily managed. Early diagnosis of such pathologies will result in early treatment and thus better prognosis.
- Cardiovascular syphilis should be considered in a case of new aortic regurgitation, especially combined with coronary ostial stenosis on angiography.

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Giant liver cyst causing haemodynamic instability

An otherwise healthy 56-year-old woman presented acutely with multiple episodes of loss of consciousness, breathlessness, cyanosis and tachycardia. She was acutely hypoxic, with blood oxygen saturation of 84% on 15 litres high flow oxygen.

On computed tomography pulmonary angiography there was no haemodynamically significant pulmonary embolism, but a giant, simple-appearing liver cyst causing significant pressure effect (*Figure 1*), as confirmed by echocardiogram. The patient improved clinically following drainage of the cyst.

Symptomatic cysts can cause right upper quadrant pain, nausea and vomiting (Kanai et al, 1999), portal hypertension and biliary stasis (Ishikawa et al, 2002). Pressure over the inferior vena cava can lead to thrombosis with lower limb oedema, and very rarely a pulmonary embolism (Buyse et al, 2004).

This rare presentation of a common disease can be added to the wide range of common and rare presentations of simple liver cysts. BJHM

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Kanai T, Kenmochi T, Takabayashi T et al (1999) Obstructive jaundice caused by a huge liver cyst riding on the hilum: report of a case. *Surg Today* **29**(8): 791–794. https://doi.org/10.1007/BF02482330

Figure 1. **a**. Axial, (**b**) coronal and (**c**) sagittal contrast-enhanced computed tomography of the abdomen and pelvis in the portal venous phase showing a giant simple liver cyst replacing the right lobe of the liver and causing pressure effect. Note the ill-defined hypodensities at the left lobe of the liver (black arrows, **a**) as a result of hypoperfusion. Compression of the right lung, right side of the heart and inferior vena cava and displacement of the right kidney also seen.