Case Report

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Atrial myxoma masquerading as Takayasu's arteritis

Kenneth Fung[1], Stephen Edmondson[2] and David S Wald[1,3]
[1]Department of Cardiology, London Chest Hospital, London E2 9JX, UK
[2]Department of Cardiothoracic Surgery, St Bartholomew's Hospital, London EC1A 7BE, UK
[3]Wolfson Institute of Preventive Medicine, Barts and the London School of Medicine, Queen Mary University of London, London EC1M 6BQ, UK
Corresponding author: David S Wald. Email: d.s.wald@qmul.ac.uk

Lesson

We describe the case of a 48-year-old woman whose atrial myxoma was mistaken for vasculitis. The case report highlights the reasons why these two disorders may become confused, the dangers of initiating the wrong treatment and a simple means of avoiding misdiagnosis.

Keywords

Atrial myxoma, vasculitis, endocarditis

Introduction

Atrial myxoma is the most common cardiac tumour, affecting two per 10,000 individuals.[1] Although it is histologically benign and causes no symptoms in about a fifth of patients with the disorder, serious complications can arise through systemic arterial embolisation in 30–40% of cases and obstruction to blood flow through the mitral valve leading to dyspnoea and orthopnoea in over 50% of patients.[2] Cases of sudden death have also been reported.[3] Diagnosis is confirmed by imaging the heart, using echocardiography or magnetic resonance imaging, and surgical removal is required as soon as the diagnosis is made. The outlook following surgery is good, with a recurrence rate of 5%.[4]

Case history

A 48-year-old woman of African descent presented to her local London hospital after an episode of collapse. She experienced dizziness while sitting, became sweaty and lost consciousness, sustaining a head injury with bruising and swelling to her right forehead and eye. On admission to the hospital, she was fully conscious and complained of central chest pain. She had never collapsed before but had a history of sickle cell anaemia and beta thalassemia trait and had suffered an unexplained pulmonary embolism 20 years earlier.

Two months prior to the collapse, while taking no regular medications, she was diagnosed with vasculitis following a 10-week period of progressive weight loss and a spontaneous left superficial femoral artery thrombosis. The possibility of vasculitis was raised following a computed tomography (CT) scan of her pelvis and abdomen that showed inflammatory changes around the left femoral artery. A subsequent ultrasound scan showed a stenosis (>85% luminal narrowing) of the left superficial femoral artery, which together with the finding of a normocytic anaemia (haemoglobin 6.3 g/dL), high C-reactive protein (CRP) of 269 mg/L and erythrocyte sedimentation rate (ESR) of 100 mm/h were judged sufficient for a rheumatologist to diagnose large vessel vasculitis (Takayasu type). Immediate treatment with oral prednisolone (60 mg per day) and low-molecular-weight heparin (enoxaparin) was started. A magnetic resonance angiogram found no other peripheral arterial abnormalities, and an autoimmune screen (anti-nuclear antibody, anti-neutrophil cytoplasmic antibody, extractable nuclear antigens cardiolipin antibody, complement C4) and immunoglobulin levels were normal. Complement C3 was mildly elevated (1.75 g/L).

After one week on prednisolone, her CRP level reduced to 7 mg/L and her ESR to 17 mm/h after eight weeks. But, her symptoms did not resolve. Weight loss, left leg weakness and pain, particularly around the popliteal fossa, persisted. She remained under rheumatology outpatient care on a dose-reducing regimen of prednisolone.

Investigations

On the day of her collapse, emergency investigations included an electrocardiogram, which showed T-wave inversion in limb leads II, III and aVF, a CT brain scan which was normal and a CT pulmonary angiogram which demonstrated no evidence of pulmonary embolism. A troponin measurement, taken 12 h after her admission, was elevated (>1200 ng/L), a provisional diagnosis of non-ST-segment-elevation myocardial infarction was made and she was referred to a specialist cardiac referral centre for invasive coronary angiography. The recent diagnosis of vasculitis

and her collapse prompted the cardiology team to arrange an echocardiogram to exclude aortic dissection and valvular disease, prior to angiography. This, unexpectedly, demonstrated a 2×3 cm echogenic mass in the left atrium, which prolapsed through the mitral valve causing a functional stenosis during diastole (Figure 1). Its lobulated appearance and connection, by a short stalk, to the interatrial septum was suggestive of an atrial myxoma. The mitral valve leaflets were thickened with a severe posteriorly directed jet of mitral regurgitation.

Treatment

Emergency surgical excision of the myxoma was undertaken that evening, and it was noted by the surgeon that the tumour appeared to be infected (Figure 2) and the posterior mitral valve leaflet was thickened with soft nodules. Endocarditis affecting the myxoma and the mitral valve was suspected, but because the mitral valve was functionally competent, it was not replaced. The mass was sent for histological and microbiological analysis that confirmed the diagnosis of atrial myxoma together with

Figure 1. Transthoracic echocardiogram demonstrating the atrial myxoma: (a) seen in left atrium at the beginning of diastole and (b) prolapsing through the mitral valve later during diastole. M: atrial myxoma; LA: left atrium; LV: left ventricle; RV: right ventricle; AMVL: anterior mitral valve leaflet; PMVL: posterior mitral valve leaflet; Ao: aorta.

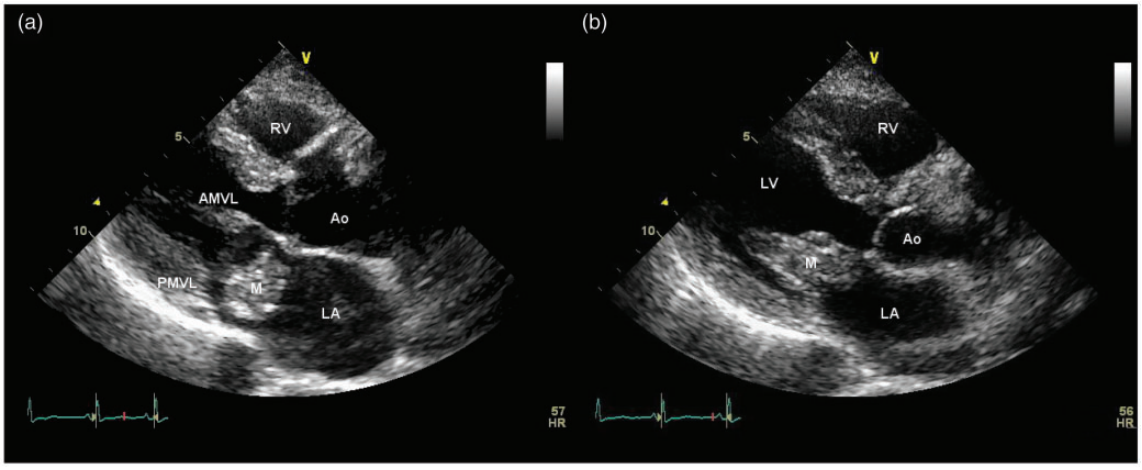
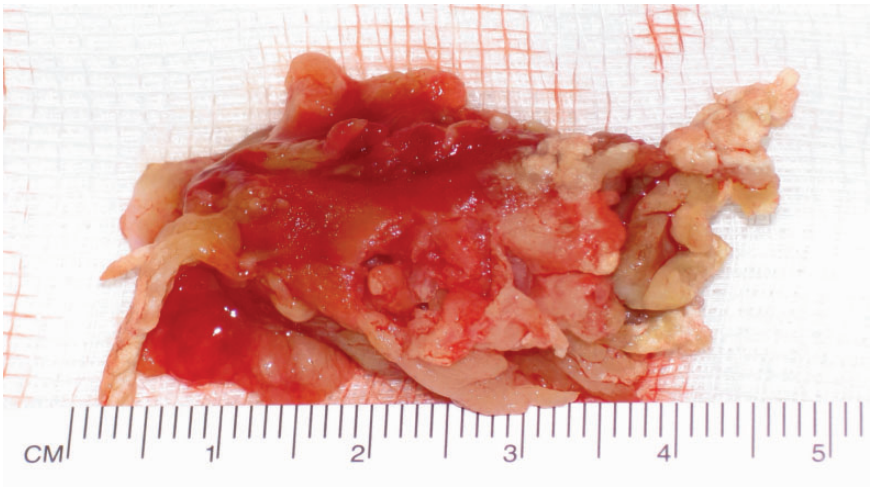


Figure 2. Surgically resected atrial myxoma.



superimposed infection by *Streptococcus parasanguinis*. A sample of one of the mitral valve nodules showed mild inflammatory cell infiltrate and endocarditis with the same organism. Given the prior diagnosis of vasculitis, a tissue biopsy of the ascending aorta was also taken during surgery, but this was histologically normal with no features of vasculitis.

Antibiotics (amoxicillin and gentamycin) were started, and the patient made a rapid recovery. One week after surgery, a repeat echocardiogram showed no evidence of the atrial mass and no mitral regurgitation. Given the negative biopsy for vasculitis, her prednisolone was reduced in dose and then discontinued completely over two weeks. She was discharged home after completing a four-week course of antibiotics and remained free from symptoms.

Discussion

This case report illustrates the overlap between atrial myxoma and Takayasu's arteritis, two separate disorders that share clinical features but have quite different aetiologies and treatment requirements.

Table 1 summarises the similarities between the general and specific clinical features of atrial myxoma and Takayasu's arteritis.[4–9] Our patient presented with weight loss and fatigue and was found to be anaemic with elevated inflammatory markers. These features are common to both disorders. Her superficial artery occlusion, on which the diagnosis of Takayasu's arteritis depended, can in retrospect, be attributed to a thromboembolic complication of the myxoma or hypercoagulability due to high levels of myxoma-related interleukin-6 and interleukin-8.[5,6]

This is not the first time a patient with atrial myxoma has been diagnosed with vasculitis.[10–13] Table 2 summarises four other reports which have been published over the past 40 years in which diagnostic confusion led to the wrong diagnosis. The consequences of misdiagnosis are serious, and in the case reported here, led to steroids being started. The high dose and long duration of steroid treatment may have predisposed this patient to endocarditis on the

Table 1. Comparison of clinical features between atrial myxoma and Takayasu's arteritis.

	Atrial myxoma	Takayasu's arteritis
General features	• Arthralgia • Fever • Malaise • Weight loss • Dizziness • Chest pain	• Arthralgia • Fever • Malaise • Weight loss • Headache and dizziness • Chest and abdominal pain
Specific features	• Thrombembolic arterial occlusion, affecting large or small arteries • Arterial thrombosis due to hypercoagulability • Collapse (mitral valve obstruction) • Sudden death (8%)	• Inflammatory arterial occlusion, affecting aorta or its large arterial branches. • Arterial thrombosis due to hypercoagulability • Limb claudication • Visual disturbance • Sudden death (2–3%)
Blood tests	• ↑CRP • ↑ESR • ↓Hb • ↑IL-6	• ↑CRP • ↑ESR • ↓Hb

Table 2. Published case reports of atrial myxoma misdiagnosed as vasculitis.

Author	Year	Age	Gender	Symptoms	Initial diagnosis
Tan et al.[10]	2010	23	Female	Headache, dizziness, lower limb weakness	Takayasu's arteritis
Gravallese et al.[11]	1995	59	Male	Fever, arthralgia, nausea	Polyarteritis nodosa
Taylor and Deutsch[12]	1992	63	Female	Collapse, hemiplegia, facial palsy	Giant cell arteritis
Leonhardt and Kullenberg[13]	1977	19	Male	Arthralgia, myalgia	Polyarteritis nodosa

myxoma and surrounding mitral valve leaflet, which is an extremely uncommon complication of atrial myxoma.

The key clinical implication arising from this case is that any patient with a suspected diagnosis of vasculitis should undergo an echocardiogram to rule out atrial myxoma. This simple, non-invasive and safe investigation would avoid misdiagnosis and the serious consequences of failing to identify and treat the true underlying disorder.

Declarations

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