LESSON OF THE MONTH

Getting to the heart of the matter

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Case history

A 57 year old man, previously fit, presented to rheumatology clinic in November 1995 with a five year history of flitting arthralgia of the knees, elbows, and shoulders. The rheumatoid factor was negative and C reactive protein (CRP) normal. The diagnosis was unclear but shoulder joint injections with corticosteroid by the family doctor had given temporary symptomatic relief.

The following month he was assessed by a neurologist with complaints of continuous headache and episodic visual blurring. Examination and computed tomography of the brain were normal. Stress related anxiety was diagnosed.

In May 1996 the patient was admitted to hospital with severe central chest pain. Electrocardiogram showed anterior ST increase consistent with a myocardial infarction and streptokinase was given. Subsequent coronary angiography was performed that showed normal coronary arteries and normal left ventricular function; coronary spasm was evoked as the mechanism for the presentation.

The following month, June 1996, the patient was again admitted to hospital this time with complaints of lethargy, generalised headache, proximal myalgia but no stiffness or weakness, visual blurring, and weight loss. There were no abnormalities on examination. Laboratory findings included a normochromic, normocytic anaemia (haemoglobin of 10.2 g/dl, mean cell volume 81.9, and mean cell haemoglobin 26.9), CRP was 59.6 (normal <10) with a negative autoantibody screen. Immunoglobulins were increased with an IgG value of 20 (normal 6.9-16.20), IgA of 4.5 (normal 0.68-3.76), but normal IgM fraction. Urine analysis was normal. Temporal artery biopsy was not done. A putative diagnosis of polymyalgia rheumatica was made and he was discharged home and prescribed prednisolone (30 mg daily). He was kept under regular rheumatological review. His symptoms eased but failed to completely settle and his CRP, although falling from 59 to 23, never completely returned to normal.

In June 1997 the patient was admitted to hospital in a semi-conscious state after three generalised tonic-clonic seizures. He was kept in the intensive care unit for two weeks. Magnetic resonance imaging of his brain showed multiple bilateral lesions in both grey and white matter consistent with ischaemia.

While in the intensive care unit the patient developed acute renal failure. He remained normotensive. Urine analysis showed red cell casts. Ultrasound of the kidneys was normal but renal arteriography showed bilateral cortical infarction and intrarenal micro-aneurysms. Anti-neutrophil cytoplasmic antibodies were negative. A diagnosis of polyarteritis nodosa was made and treatment started with cyclophosphamide and prednisolone. Despite continuous immunosuppressant treatment there were episodic complaints of headache and the CRP continued to fluctuate over the next few months.

In December 1997 the patient was transferred to a rehabilitation hospital. His cerebral disorder had left him cognitively impaired and wheelchair bound. During rehabilitation he developed malaise and a low grade pyrexia. Chest radiography, repeated blood cultures, urine analysis, and lumbar puncture disclosed no signs of infection. Cardiovascular examination was normal. However a transthoracic echocardiogram showed the presence of a large mass in the left atrium (fig 1). Subsequent surgical excision and histological examination showed this to be a left atrial myxoma. After excision the patient's constitutional symptoms

Figure 1 Transthoracic echocardiogram showing mass (M) in left atrium (LA). Left ventricle (LV), aorta (AO).

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settled but there was little improvement in his cognitive capacity.

Discussion

Cardiac myxomas are the most common primary cardiac neoplasm, 75% arising from the left atrium.¹ They may present in three ways: intracardiac obstruction, embolisation, and constitutional symptoms.² Intracardiac obstruction most often mimicks mitral stenosis resulting from obstruction of blood flow across the mitral orifice. Patients can present with dyspnoea and syncope and on examination may have a loud first heart sound, a tumour “plop” in early diastole because of the tumour prolapsing through the valve, and variable mitral diastolic and systolic murmurs. Our patient, however, had no such features and cardiovascular examination can be entirely normal with even large myxomas. About half of patients present with embolic manifestations, half of which involve the brain. Transient visual loss can result from involvement of the retinal arteries and strokes from cerebral infarcts.³ Embolisation to the coronary arteries can cause myocardial infarction.⁴ Peripheral embolisation can not only cause renal infarcts but also arterial aneurysms. The latter is thought to occur from emboli that invade and weaken the arterial wall. Angiographically this can mimic polyarteritis nodosa and occasionally systemic lupus erythematosus, Wegener’s granulomatosis and infective endocarditis.^{5–7} Constitutional symptoms are present in 90% of patients and include fever, myalgia, weight loss and arthralgia, which can cause diagnostic confusion with polymyalgia rheumatica. Laboratory abnormalities include anaemia, an increased erythrocyte sedimentation rate and CRP and hypergammaglobinaemia.¹

The embolic phenomena and the constitutional symptoms of atrial myxomas can emulate the symptomatology of many multisystem disorders. This, together with the visualisation of microaneurysms on renal angiography, can wrongly lead to the diagnosis of polyarteritis nodosa. A high clinical index of suspicion is essential for correct diagnosis.

The lessons

- Atrial myxomas can mimic the clinical features of polyarteritis nodosa and polymyalgia rheumatica.
- Both polyarteritis nodosa and myxomas can cause microaneurysms on renal arteriography.
- Atrial myxomas may produce no cardiac signs.
- Echocardiography should be considered in all cases of polyarteritis nodosa where inflammatory markers do not settle with appropriate treatment.

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