Clinical Medicine 2013, Vol 13, No 1: 106–9

lesson of the month (2)

Stroke in a 53-year-old woman: getting to the heart of the problem

This Lesson of the Month illustrates both the complications and the pivotal role that echocardiography has in the diagnosis of a cardiac source of embolism.

Introduction

A 53-year-old woman, who had previously been fit and well, presented to the emergency department (ED) with headache, loss of consciousness and left hemiplegia of sudden onset. Her past medical history included neurosurgery for a left temporo-occipital cavernoma four years previously and a separate incidence of 'labyrinthitis'. Cardiovascular examination did not reveal any abnormalities. The patient underwent routine investigations, including an electrocardiogram (ECG), blood tests, a computerised tomographic (CT) scan of head and a magnetic resonance (MR) scan of head (Fig 1). The latter showed a large cerebral infarct. The patient was found to be in sinus rhythm; 24-hour Holter monitoring and carotid duplex scan were also normal. On further questioning, there was a history of weight loss and night sweats.

A cardioembolic cause for the cerebral infarct was suspected and consequently confirmed by a transthoracic echocardiogram (TTE). The TTE demonstrated a large, mobile echodense mass within the left atrium (LA) (Fig 2). The irregular and frond-like mass, resembling a cluster of grapes, appeared to be attached to the fossa ovalis. Its appearance was considered to be consistent with a LA myxoma. Thrombolysis was not attempted and the patient was unfit for cardiac surgery. Despite cranial decompression on day two, the patient died 35 days after presentation.

The MR image in Fig 1 demonstrates right frontoparietal lobe intensity, indicating acute infarction of territory of the right middle and right anterior cerebral arteries.

A consented hospital post-mortem was performed and showed that the immediate cause of death was bronchopneumonia following a massive right anterior cerebral artery territory infarct. The distal right internal carotid artery and its branches were all occluded by a tumour embolus typical of an embolic myxoma (Fig 3). Acute infarcts were seen in kidneys, spleen and liver, and an old infarct was seen in the myocardium. An old right cystic cerebellar infarct was also confirmed. The LA contained a tan-coloured, elongated, lobulated mass 25-mm long and 8 mm in maximum diameter (Figs 4 and 5). It was firmly attached to the interatrial septum just posterior to the closed fossa ovalis and extended to the orifice of the mitral valve. Histology of both the cardiac mass and the tumour embolus in the right internal carotid artery was identical. Immunohistochemistry for calretinin was positive in both cardiac and arterial samples confirming that this was an atrial myxoma. Myxoid sarcomas (myxoid imitators) and organised thrombus were excluded. Although thrombus was present in both cardiac and cerebral artery lesions, it was not significant in amount.

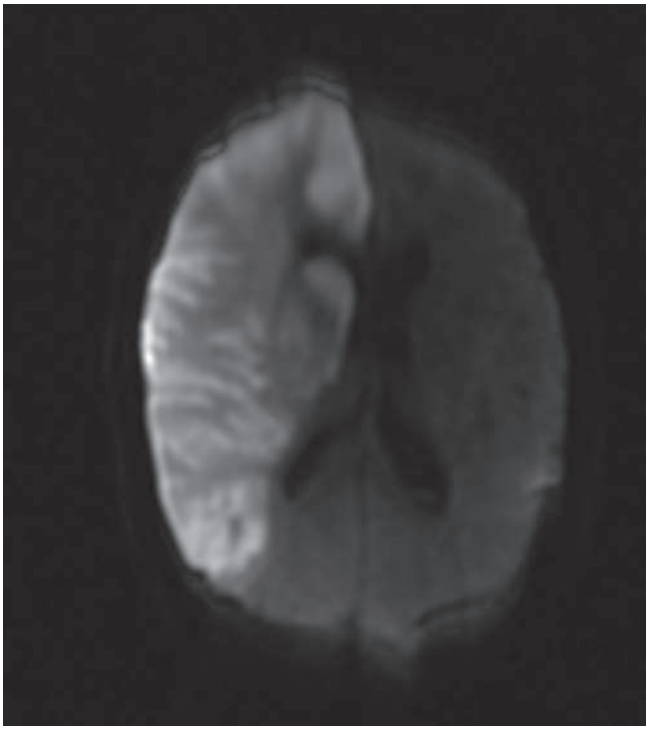

Fig 1. Magnetic resonance axial diffusion-weighted scan of head.

Cardiac source of embolism

The causes of stroke can be classified into two major categories, with approximately 85% being ischaemic and the remaining 15% being haemorrhagic.[1] Of those ischaemic strokes, embolism from a cardiac source accounts for approximately 20% and is shown to carry a poorer outcome compared with other causes of stroke. Echocardiography is indicated in the setting of a suspected cardiac mechanism for stroke or related symptoms of syncope and

Alexander W Pearce,[1] chief cardiac physiologist; **Bushra S Rana,**[1] consultant cardiologist; **Dominic G O'Donovan,**[2] consultant neuropathologist

[1]Papworth Hospital NHS Foundation Trust; [2]Addenbrooke's Hospital, Cambridge

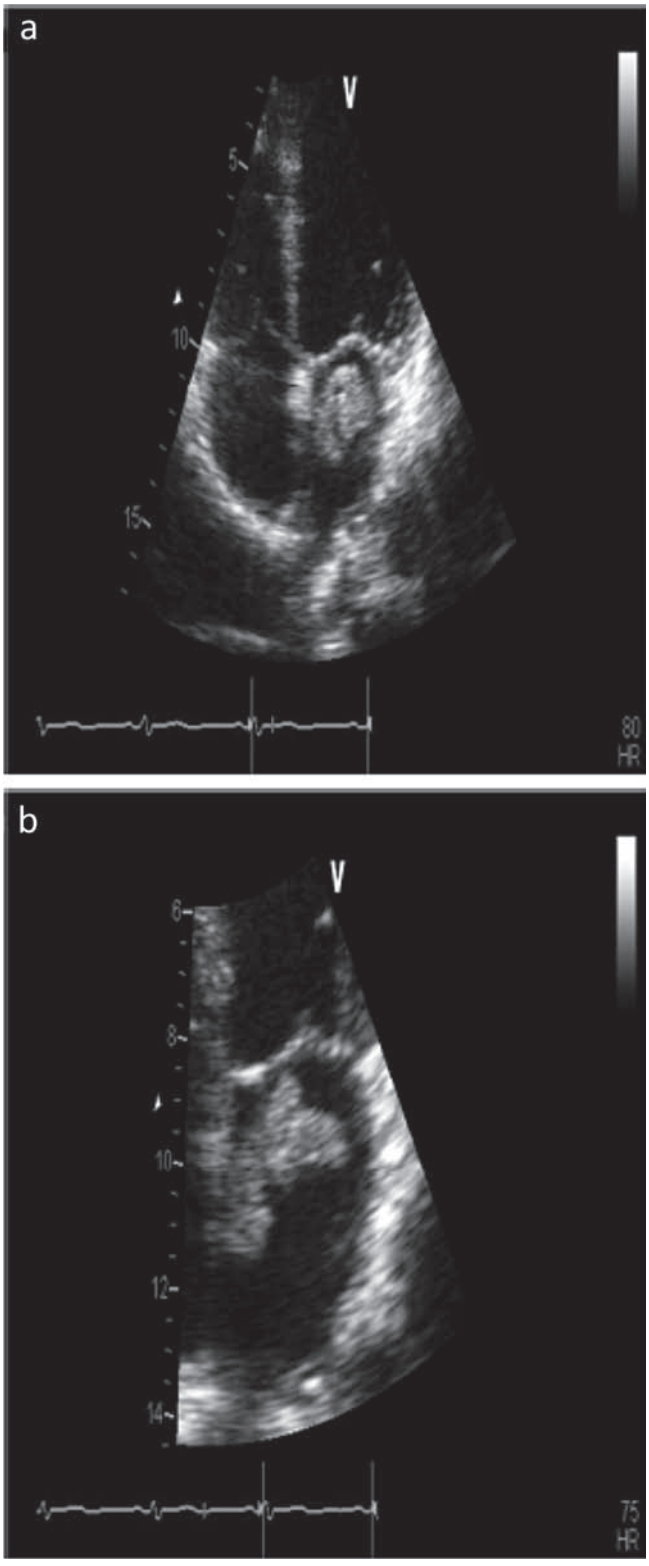

Fig 2. Transthoracic echocardiogram. (a) Apical four-chamber view; (b) left atrial-focused view.

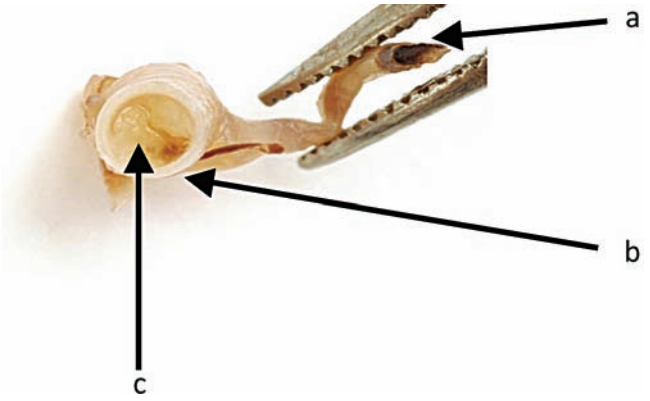

Fig 3. Right internal carotid artery occluded by embolic myxoma from left atrium. a = right anterior cerebral artery; b = right internal carotid artery; c = embolic myxoma.

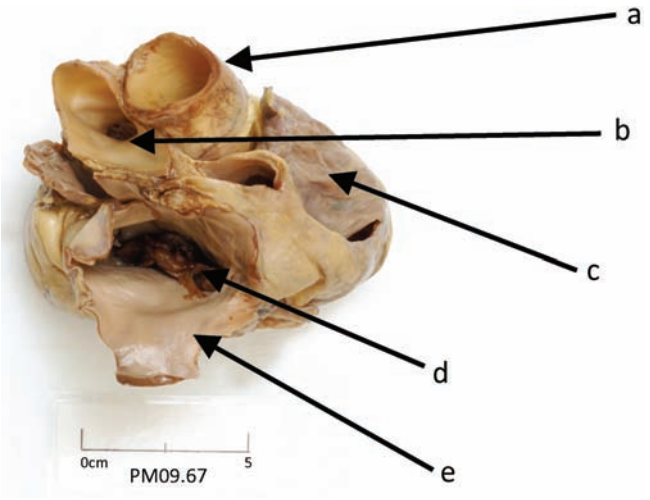

Fig 4. Superior view of heart. a = aortic root; b = pulmonary trunk; c = right atrium; d = tumour and post-mortem clot; e = left atrium.

Box 1. Echocardiography checklist protocol for suspected atrial myxoma.

- Establish dimensions of tumour mass.
- Establish site of attachment (usually fossa ovalis attached via a stalk).
- Determine whether there is valve leaflet involvement (tumour might attach to the surface of the leaflet).
- Exclude other and/or multiple sites (see main text).
- Assess haemodynamic effects: a large tumour that attaches to a valve leaflet or prolapses across the mitral valve orifice might cause a degree of obstruction (assessment as per mitral stenosis).

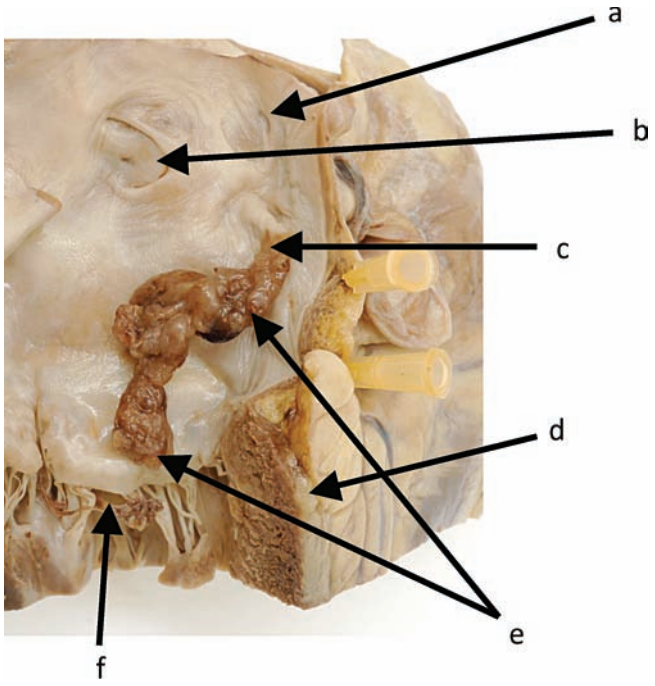

Fig 5. Left atrium and upper left ventricle. a = left atrial wall; b = fossa ovalis; c = point of attachment of myoxma to the posterior interatrial wall posterior to fossa ovalis; d = upper posterior left ventricle wall; e = myxoma polypoid free-floating tumour; f = mitral valve posterior cusp.

Table 1. Causes of cardiac source of embolism.

Sources	Causes
High-risk sources	
Atrial fibrillation	
Left ventricular dysfunction	Recent myocardial infarction
	Left ventricular aneurysm
	Cardiomyopathies
Valvular pathology	Mitral stenosis
	Endocarditis
	Mechanical valve prosthesis
Cardiac masses	Tumours
Proximal aortic atheroma	
Low-risk or undetermined risk sources	
Valvular pathology	Mitral valve prolapse
	Calcific aortic stenosis
	Mitral annular calcification
	Giant Lambl's excrescences
Paradoxical embolism	Patent foramen ovale
	Atrial septum aneurysm

transient ischaemic attack (TIA). In this regard, echocardiography has a pivotal role in determining cardiac sources. Causes of cardiac sources of embolism are summarised in Table 1, and are categorised according to embolic risk; high, low and/or undetermined.

Cardiac masses seen on echocardiography can be broadly classified into one of three categories: tumour, thrombus or vegetation. Cardiac tumours can be primary or secondary, benign or malignant and arise from the heart (primary) or be deposited in the heart from a remote primary (metastasis). They can present with embolic events and stroke, which might be the first indication of their presence, as illustrated by our case. The most common tumours associated with embolic stroke are cardiac myxomas and papillary fibroelastomas of the cardiac valves.

Myxomas

Although sporadic cardiac myxomas present more often in middle-aged (30–70 years) females (male:female ratio is 1:3), 7% are familial and associated with Carney complex. Cardiac myxomas are benign intrinsic neoplasms possibly arising from intracardiac sensory nerve ganglia, which are also calretinin positive.[2] Such myxomas are the most common primary neoplasm of the heart, representing approximately 30–50% of all primary cardiac neoplasms, although metastatic (secondary) malignant neoplasms are more frequent. Most (90%) are found in the LA and are pedunculated, arising from the fossa ovalis. Other sites include the right atrium (approximately 18%), left ventricle (approximately 4%) and right ventricle (approximately 4%), although myxomas can occur at multiple sites simultaneously.[3] Cardiac myxomas cause constitutional symptoms (paraneoplastic reactions), including malaise, arthralgia, pyrexia of unknown origin, night sweats, anorexia and cachexia, and can lead to haemolytic anaemia, dysglobulinaemia and thrombocytopenia. Cardiac presentations include cardiac failure and syncope. If the tumour mass causes significant mitral valve obstruction, then clinical signs of mitral stenosis might be present, including a typical early low-pitched diastolic murmur known as a 'tumour plop' on auscultation.[3] Occasionally, they might also be incidental findings.

The primary objective in the treatment of a cardiac myxoma is to remove it surgically. This is performed by surgical shaving of the tumour from the septum[4] or excision with a small amount of atrial septal tissue to prevent recurrence and the septum repaired, if necessary, with pericardial tissue.[5]

Echocardiographic assessment

TTE is a non-invasive method for assessing cardiac tumours and is useful for establishing their location, extent, attachment and haemodynamic consequences, as well as for excluding multiple-site involvement (see Box 1 for an echo checklist). Myxomas are gelatinous, lobulated and often pedunculated neoplasms that resemble a cluster of grapes, having an irregular surface and usually being attached to the endocardium via a stalk (ie pedunculated). A myxoma is generally heterogeneous in appearance, containing bright echodense regions if there is calcification. Superimposed onto this mass might be thrombus, although this is usually difficult to differentiate from the tumour mass on TTE. If there is suspicion of tumour infiltration, which cannot be confidently assessed, then transoesophageal echocardiography (TOE) might be needed. If all the salient information cannot be acquired with echocardiography, then a further evaluation with a CT or MR scan might be required.

Acknowledgements

The authors wish to thank Caroline Pryke, Cardiology, Addenbrooke's Hospital, Cambridge, for her contributions to this article. The authors also thank Dr S Nagaraju, Neuropathology, Queen Elizabeth Hospital, Birmingham; Dr J Cross, Neuroradiology, Addenbrooke's Hospital,; Dr E O'Brien, Medicine, Addenbrooke's Hospital; Dr B Seymour, Neurology, Addenbrooke's Hospital; and the Departments of Neuropathology, Cardiology and Medical Photography, Addenbrooke's Hospital.

References

1 Adams HP Jr, Bendixen BH, Kappelle LJ *et al.* Classification of subtype of acute ischemic stroke: definitions for use in a multicentre clinical trial. TOAST: Trial of Org 10172 in Acute Stroke Treatment. *Stroke* 1993;24:35–41.

2 Terracciano LM, Mhawech P, Suess K *et al.* Calretinin as a marker for cardiac myxoma. Diagnostic and histogenetic considerations. *Am J Clin Pathol* 2000;114:754–9.

3 Otto C. *Textbook of clinical echocardiography*, 4th edn. USA: Saunders Elsevier; 2009.

4 Lee VH, Connolly HM, Brown RD Jr. Central nervous system manifestations of cardiac myxoma. *Arch Neurol* 2007;64:1115–20.

5 Hanson EC, Gill CC, Razavi M, Loop FD. The surgical treatment of atrial myxomas. Clinical experience and late results in 33 patients. *J Thorac Cardiovasc Surg* 1985;89:298–303.

Address for correspondence: Dr AW Pearce, Papworth Hospital NHS Foundation Trust, Papworth Everard, Cambridge CB23 3RE. Email: alex.pearce@papworth.nhs.uk

Clinical Medicine 2013, Vol 13, No 1: 109–10

Literature and medicine

Tomas Tranströmer's stroke of genius

This column explores the links and synergies between medicine and literature. What roles can literature play in reflecting and influencing good practice, and what sorts of images of doctoring are to be found in drama, poetry, fiction, biography, electronic fora and film? The editors would be pleased to receive short papers, ranging from 500–1,000 words, on relevant topics. Those interested in contributing should email brian.hurwitz@kcl.ac.uk or neil.vickers@kcl.ac.uk

In November 1990, the widely acclaimed Swedish poet Tomas Tranströmer (b. Stockholm, 1931) lost his speech and the use of his right hand as a result of a stroke. As if anticipating his own fate, in his longest poem *Baltics* (1974), he had referred to the story of a composer who became speechless and hemiplegic after a brain bleed:

Then, cerebral hemorrhage: paralysis on the right side with aphasia,
can grasp only short phrases, says the wrong words.
Beyond the reach of eulogy or execration.
But the music´s left, he goes on composing in his own style,
for the rest of his days he becomes a medical sensation.
He wrote music to texts he no longer understood
in the same way
we express something through our lives
in the humming chorus full of mistaken words.[1]

Himself a lifelong amateur pianist, Tranströmer carried on playing the piano with his left hand after the stroke. Some pianists, inspired by his first post-stroke collection of verse, *The sad gondola* (1996), composed left-handed pieces for him, such as the *Tranströmer settings* for the New European Ensemble's 2010 tour of Sweden.

Left-handed piano pieces have been written by great composers, including Prokokiev and Richard Strauss. In 1928, Maurice Ravel composed *The concerto for the left hand in B major* for Paul Wittgenstein, the Austrian musician who had lost his right arm in the Great War, which enabled him to resume concert performances. Ten years later, in 1933, at the age of 58, the French composer came to the end of his musical career after losing his own speech through a brain insult that rendered him unable to express musical ideas in either writing or performance.[2]

Neurology as a medical specialty was founded in the late 19th century when dysfunctioning structures of the brain were localised according to the anatomico-pathological method. A founding cornerstone of this approach was the 1861 structural localisation of expressive language, which was found to reside over the foot of the third left frontal convolution. This finding was confirmed by a brain necropsy on a patient of Paul Broca who had suffered from a 20-year history of non-fluent aphasia.[3] Since then, neurologists have recognised that almost all patients with expressive aphasia have some degree of difficulty in writing (dysgraphia). More challenging and overambitious are recent attempts to localise functions as complex and abstract as identification and appreciation of beauty and creativity by using cutting-edge brain-scanning technology. Such functions are not '*clean-cut. Like/ the shaved parts of a patient's crown before he has a brain operation*' to borrow Tranströmer´s expression referring to artificial landscapes.[1]

What makes Tranströmer a medical sensation — as prophetically announced in *Baltics*, the poem he regarded as his '*most consistent attempt to write music*'[1] — is the fact that he managed to continue translating

Ivan Iniesta, consultant neurologist and honorary clinical lecturer

The Walton Centre NHS Foundation Trust and Liverpool University, UK