Unusual association of diseases/symptoms

An unusual cause of stroke in an older patient

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Summary

The case of an older patient admitted with an acute stroke in the presence of atrial fibrillation is presented. Subsequent transthoracic echocardiography and advanced imaging suggested the diagnosis of a large papillary fibroelastoma on the anterior mitral valve leaflet (AML). The patient underwent surgical resection and made an excellent clinical recovery.

BACKGROUND

Cardioembolic events are implicated in the aetiology of up to a third of all strokes.¹ Atrial fibrillation with thrombus embolisation from the left atrial appendage is the most common cardiac cause.² Other embolic sources include vegetations, tumours, aortic atheromatous plaques and paradoxical embolisation across a patent foramen ovale. Most authorities advocate screening echo studies in younger patients with cryptogenic stroke but screening in older patients is less frequently performed.³ This case highlights the importance of always considering an embolic cause for stroke and performing a routine echocardiogram in patients who have experienced stroke, regardless of age, even in the presence of atrial fibrillation.

CASE PRESENTATION

A 79-year-old woman, with a background of mild hypertension, presented with sudden onset of drowsiness and left-sided weakness. Clinical examination revealed signs of a left hemiparesis, nominal dysphasia and a left inferior quadrantanopia. Atrial fibrillation was noted clinically and confirmed on electrocardiogram. An initial CT scan ruled out any haemorrhage and demonstrated the presence of small vessel disease and lacunar infarcts in the deep white matter. Her Barthel score (for activity of daily life) was 7 on admission. The working diagnosis was of an embolic stroke in the right middle cerebral artery (anterior circulation) territory, presumed from the left atrial appendage, secondary to atrial fibrillation. She presented outside the current time window for administering thrombolysis. There was steady clinical improvement and oral anticoagulation was given after 10 days and the warfarin dose adjusted to achieve an international normalised ratio (INR) of between 2.5 and 3.5. Over the next 7 weeks, she made excellent progress with almost complete resolution of her presenting symptoms and signs. Her Barthel score was 16 on discharge. A later CT scan confirmed a right middle cerebral artery territory infarct.

INVESTIGATIONS

Routine laboratory investigations were essentially unremarkable. Carotid Doppler studies showed normal

flow patterns. She remained in atrial fibrillation. Later, a routine transthoracic echocardiogram demonstrated a significant abnormality in the form of a mobile echo density adherent to the anterior mitral leaflet, prolapsing into the left ventricular outflow tract. Transoesophageal echocardiography demonstrated a polypoid mass, approximately 1.5 cm in diameter, adherent to the chordal attachments of the anterior mitral leaflet. This was highly mobile and was flopping into the outflow tract of the left ventricle just below the aortic valve (figure 1A, yellow arrow). The echo appearances were suggestive of a tumour, which was felt most likely to be a papillary fibroelastoma (PFE) based on the echo features, position and clinical presentation. Live matrix 3D transoesophageal echocardiography dramatically demonstrated the highly mobile nature of this polypoid tumour and significant potential for further embolisation (video 1–2).

Video 1 TOE 0 degree mid oesophageal view (Real time 3D) showing mobile polypoid mass on ventricular surface of anterior mitral leaflet. 10.1136/bcr.11.2009.2476v1

Video 2 TOE(2D) mid oesophageal view showing polypoid mass prolapsing into LVOT. 10.1136/bcr.11.2009.2476v2

The left atrial appendage was clearly visualised and was free of thrombus.

DIFFERENTIAL DIAGNOSIS

A cardiac tumour was diagnosed by echocardiography. By its position, echo appearance and clinical context a PFE was felt to be the most likely diagnosis. Though more prevalent, this would be a highly unusual position for a cardiac myxoma. Other causes of a mass on the mitral leaflet such as vegetation or even thrombus were felt to be unlikely in this clinical context.

TREATMENT

A decision was made to refer her for surgical excision. Pre-operative coronary angiography excluded any critical coronary artery disease. At surgery, a gelatinous mass 2.0 × 1.5 cm was identified on the ventricular surface of the anterior mitral leaflet involving the chordae and free margin of the

Figure 1 Composite slide showing 2D transoesophageal echocardiography (TOE) appearance (A), surgical view (B), excised specimen (C) and histology (D).

leaflet (figure 1B,C). Repair was not technically achievable so the anterior mitral leaflet was excised along with the mass, and the valve replaced with a 29 mm perimount magna valve (bioprosthesis) with external radiofrequency ablation of the pulmonary veins performed. Histology confirmed the typical features of a PFE (figure 1D).

OUTCOME AND FOLLOW-UP

The patient made an uneventful recovery following surgery. She remains well. Despite undergoing perioperative radiofrequency ablation, she has remained in atrial fibrillation and therefore has been maintained on oral anti-coagulation.

DISCUSSION

PFEs are histologically benign endocardial papillomas that predominantly affect cardiac valves.⁴ They are the second most common benign primary cardiac tumours (after myxomas) and account for three-quarters of all cardiac valve tumours. PFEs are gelatinous masses with ‘sea anemone’ appearance with a size usually less than 1 cm, seen on echocardiography as a single mobile mass attached to the valve with a small pedicle.⁵ There are no characteristic clinical features of the condition, with most of them found incidentally during echocardiography, cardiac surgery, cardiac catheterisation or autopsy. Although usually asymptomatic they can cause chest pain, transient ischaemic attacks or stroke, dyspnoea or even sudden death.

Embolic fragments may arise either from the tumour itself or from fibrin forming on the tumour. Surgical resection is the treatment of choice even in patients who are asymptomatic because of the risk of embolisation.⁴

Transthoracic echocardiography is part of the routine investigation of cardiac source of embolus in patients who have experienced stroke in many centres across the world,^{6 7} but guidelines vary. A recent hospital-based study from Canada reports that transthoracic echocardiography has a low yield, diagnosing abnormalities suggestive of cardioembolic aetiology in only 4% of patients who are ischaemic and have experienced stroke.⁸ In contrast, another single centre study from Portugal revealed 37.2% of such patients had echocardiography abnormalities suggestive of a cardioembolic aetiology and the authors recommended the use of echocardiography as a routine procedure in all patients who have experienced stroke.⁹

In the UK, the Royal College of Physicians National Clinical Guidelines for Stroke 2008 mentions echocardiography as part of investigations for transient ischaemic attacks but does not advocate routine transthoracic echocardiography for all patients who have experienced stroke.^{2 10 11} However, any clinical suspicion of a cardioembolic cause for a stroke should prompt an urgent transthoracic echo regardless of the age of the patient. Contrast bubble echo should be employed where there is consideration of a patent foramen ovale as a possible cause of cryptogenic stroke, which may also occur in older patients.¹²

Learning points

- ▶ Although atrial fibrillation is the commonest cause of embolisation from the heart, its presence does not rule out other cardiac pathology.
- ▶ Echocardiography should be carried out in most patients who have experienced stroke regardless of age.
- ▶ Cardiac tumours are rare and although papillary fibroelastomas (PFEs) and myxomas and are both histologically benign, they should be considered malignant in view of their embolic potential.
- ▶ Careful cardiological assessment should be carried out in suspected cases of cardiac tumours as advanced imaging can often suggest the diagnosis and optimal management plan.

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