CASE REPORT

Fibroelastoma in an unusual location: a rare cause of multiple cerebrovascular events

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Summary

Fibroelastomas are rare, primary cardiac tumours with a predilection for valvular endothelium and a propensity to embolise. We present the case of a 72-year-old male with multiple cerebrovascular events (CVA) despite oral anticoagulation. Transoesophageal echocardiography (TOE) revealed a small highly mobile left atrial mass with frond-like projections attached by a stalk to the orifice of the LAA. The mass was surgically excised and confirmed to be a fibroelastoma on histological examination. This case report describes a rare but treatable source of multiple cerebrovascular events and highlights the utility of TOE in the assessment of cardiac embolic source.

Key Words

- fibroelastoma
- left atrial appendage
- 2D transoesophageal echocardiography
- transient ischaemic attack

Learning points:

- Fibroelastomas are most commonly found on left-sided heart valves (aortic > mitral) and have the potential to cause systemic emboli associated with significant morbidity and mortality.
- A left atrial appendage (LAA) mass in a patient presenting with cerebrovascular events does not always represent thrombus. Uncommon aetiologies such as a cardiac tumour should be considered in the differential diagnosis.
- Transthoracic echocardiography (TTE) does not provide an accurate assessment of the LAA and should not be used to detect pathology within this structure. Transoesophageal echocardiography (TOE) is superior to TTE in imaging the LAA and provides a complete delineation of its anatomy. In addition, TOE can detect very small highly mobile lesions (as described in this case), which may be missed on other imaging modalities.

Background

Fibroelastoma is a histologically benign primary intra-cardiac tumour, which can present with cerebrovascular events due to systemic embolisation. Surgical excision is required in symptomatic patients. TOE plays a pivotal role in the detection of these tumours, which most commonly originate from the aortic valve and are rarely found in the LAA.

Case presentation

A 72-year-old male with well-controlled NIDDM and no prior history of neurological or cardiac disease was referred for a TOE following multiple admissions to our hospital with a cerebrovascular event. The CVAs continued to occur despite the initiation of and compliance with Apixaban. There was no history of febrile illness or cardiac symptoms. Physical examination was unremarkable.

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Investigation

Comprehensive serial neurological imaging demonstrated new multifocal infarcts consistent with emboli. There was no evidence of arrhythmia on cardiac monitoring during hospital admission or Holter monitoring performed in the community. Chest X-ray and routine blood tests including inflammatory markers, autoimmune and coagulation panel were unremarkable. Transthoracic echocardiography (TTE) did not demonstrate any abnormalities.

TOE revealed a solitary, small, well-defined highly mobile mass (measuring 0.7 × 0.7 cm) in the left atrium (Fig. 1). It was attached by a broad stalk to the orifice of the LAA at the Coumadin ridge, adjacent to the left upper pulmonary vein (Figs 2 and 3). The mass had multiple thin fronds attached to its surface and did not occlude the LAA or left upper pulmonary vein (Video 1). The LAA was contractile with an emptying velocity of >40 cm/s. Atrial and ventricular size and function were normal, and no valvular pathology was identified. There was no inter-atrial shunt or significant aortic atheroma. Given the history of recurrent CVAs (with no identifiable source on previous investigation) in the presence of a mobile left-sided cardiac mass, prompt referred for surgical excision of the mass was made.

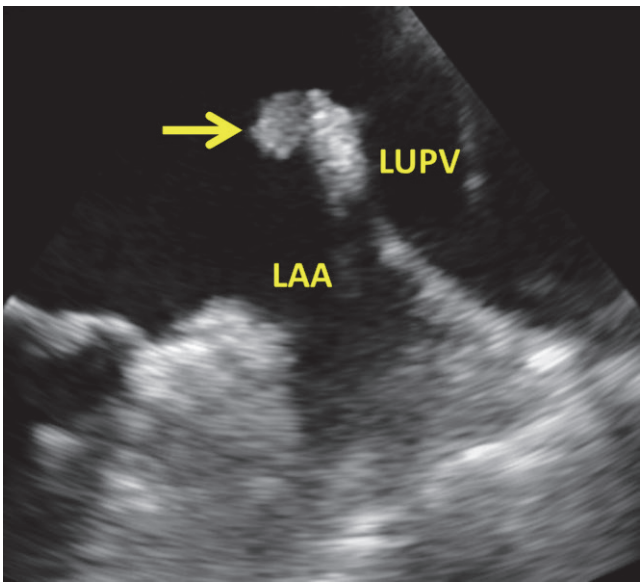

Figure 1
Mid-oesophageal TOE view. The anatomical position of the fibroelastoma (arrow) is seen in relation to the left upper pulmonary vein (LUPV). The mass is well defined, with a heterogeneous texture. LAA, left atrial appendage.

Video 1
The heterogeneous texture and the mobile nature of the fibroelastoma are illustrated in this video. Multiple thin frond-like structures can be seen emanating from the surface of the mass. View Video 1 at http://movie-usa.glencoesoftware.com/video/10.1530/ERP-19-0012/video-1.

Treatment and outcome

The mass was excised during an uneventful surgery, and macroscopic examination revealed it to be a fibroelastoma (with no superimposed thrombus). The patient made a good post-operative recovery, with no further neurological deficits following discharge.

Discussion

The major differential diagnosis considered in our patient was thrombus or cardiac tumour (myxoma or fibroelastoma). There is an overlap in the appearance of thrombus, myxoma and fibroelastoma on echocardiography, with mobility and the presence of a stalk being shared features. The differential diagnosis had significant clinical relevance as the management strategies between thrombus and tumour differ significantly. Although the LAA is a common location for thrombi, we felt that it was less likely to be the case in this patient for the following reasons: left atrial and LAA size were normal, no evidence of spontaneous echo contrast (a precursor for thrombus), the LAA was contractile with normal emptying velocity and the patient had been on anticoagulation for several months with no evidence of arrhythmia. Thus, changing or increasing the dose of anticoagulation therapy with TOE follow-up (to detect reduction/resolution of the potential 'thrombus') was not thought to be a safe strategy. The probability of the mass being either lipoma or vegetation was thought to be low as the frond-like surface protrusions and stalk are not typical features of a lipoma (which are usually broad-based and immobile) (1). The absence of a febrile illness, elevated inflammatory markers and upstream valvular lesions made endocarditis less likely.

Fibroelastomas are small solitary slow-growing lesions which typically arise from the downstream surface of left-sided valves and rarely cause valvular obstruction. They are composed histologically of a superficial endothelial layer, an intermediate proteoglycan layer and

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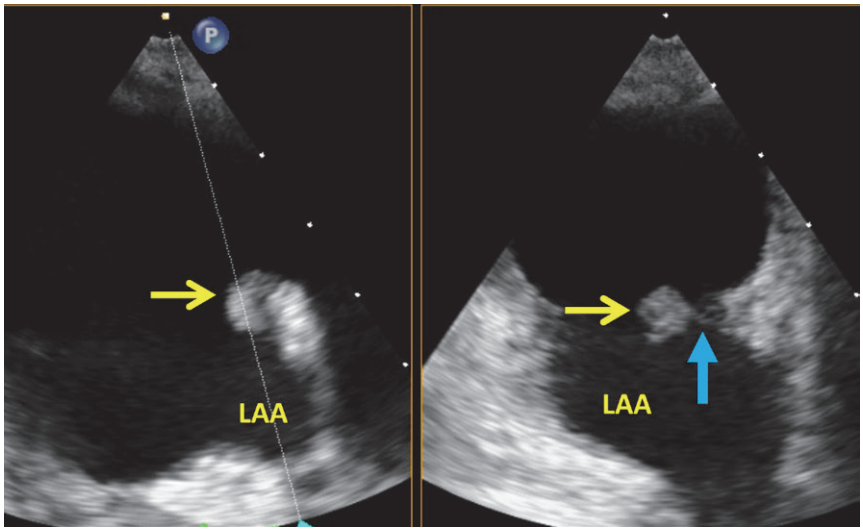

Figure 2
Mid-oesophageal TOE view with 'X-plane' (bi-plane) of the fibroelastoma (yellow arrow) at a 75° view. The stalk at the base of the fibroelastoma is visualised (blue arrow). LA, left atrium.

a central avascular collagen core. Acute and organising thrombi may be seen on the surface. Macroscopically, fibroelastomas have multiple thin papillary fronds attached to the endocardial surface by a short single stalk, and when immersed in water they typically resemble a sea anemone on gross examination (2).

They are the second most common primary cardiac tumour. However, fibroelastoma contribute to less than 1% of all cardioembolic strokes (3). The clinical presentation varies widely from an incidental finding in an asymptomatic individual, to cardiac and/or neurological manifestations relating to embolisation or obstruction. The mechanism of embolisation is thought to be a combination of tumour fragments and superimposed thrombus. In symptomatic individuals, cerebrovascular events are a common presenting symptom, and a number of studies have highlighted the association between CVA and fibroelastoma (4, 5, 6). In a retrospective review of 88 patients who underwent surgical excision of a histologically confirmed fibroelastoma, over 50% had presented with CVA (7). In another single-centre study, CVA was the presenting symptom in one-third of patients found to have fibroelastoma detected on echo (8). The stroke risk of conservatively managed fibroelastoma is not insignificant. Tamin *et al.* (4) found the incidence of CVA was 6% at 1 year in patients with unoperated fibroelastoma, increasing to 13% at 5 years. Sun *et al.* (5) found the incidence of CVA in 45 patients with untreated fibroelastoma to be 6.6% over 11–22 months from diagnosis.

Echocardiography remains the primary form of cardiac imaging in the diagnostic work up of suspected cardiac source of emboli. TOE is superior to TTE in imaging the LAA (as demonstrated in this case) and in detecting small cardiac lesions with embolic potential due to its greater resolution. It is therefore appropriate to proceed to TOE in the presence of a negative TTE if there is clinical suspicion of a cardioembolic source of CVA. Two-dimensional (2D) TOE depicts the typical echocardiographic features of fibroelastoma (described below) and is supplemented by three-dimensional (3D) TOE which provides anatomical information such as proximity of the mass to adjacent structures and attachment point, which may be useful for surgical

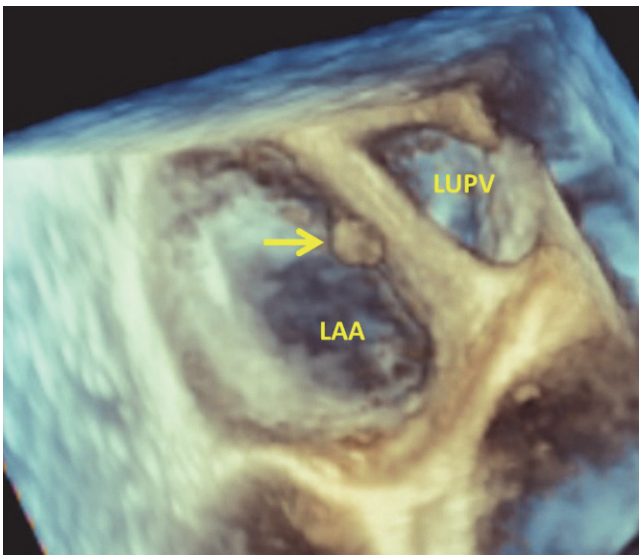

Figure 3
Three-dimensional (3D) TOE reconstruction of an *en face* view of the orifice of the LAA and left upper pulmonary vein (LUPV). The fibroelastoma (arrow) is seen to originate from the orifice of the LAA at the Coumadin ridge.

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planning. A multi-modality imaging approach using cardiac magnetic resonance (CMR) or computed tomography (CT) for tissue characterisation of the mass may be required in some cases of suspected fibroelastoma to differentiate between thrombus and other cardiac tumours. In contrast to TOE, the reduced temporal resolution of these modalities may limit their accuracy in the detection of small highly mobile lesions (which are typical characteristics of fibroelastoma).

The echocardiographic features of fibroelastoma described in the literature are small (<2 cm), mobile, round/oval lesion of predominantly valvular origin, with well-demarcated borders. A speckled texture with 'stippling' on the perimeter is usually seen, and >50% have stalks (associated with increased mobility) (9). Fibroelastoma mobility has been reported to be an independent predictor of death and embolisation (6), but it remains unclear whether tumour volume has prognostic significance, with conflicting data in the literature (7).

In symptomatic patients, surgical excision is the accepted first-line treatment to prevent further embolic events. Surgical excision is considered curative, and recurrence following resection is very rare. There is a paucity of data evaluating the efficacy of long-term anticoagulation in conservatively managed patients, and its role should be limited to patients deemed unsuitable for surgery. In asymptomatic patients, there is significant controversy in the literature on management strategy, with no robust data to guide risk stratification and optimal timing for surgical intervention. Different management algorithms have been proposed, taking into account the location, mobility and size of the fibroelastoma, with emphasis placed predominantly either on location (left vs right side) or mobility (10). Using a case-by-case approach, prophylactic surgery may be considered in asymptomatic low risk surgical patients with a mobile left-sided fibroelastoma, or in those undergoing cardiac surgery for other reasons.

Conclusion

Fibroelastomas are histologically benign cardiac tumours with a high risk of embolisation and should be considered in the differential diagnosis of cerebrovascular events. The LAA is a most unusual site for this lesion, and as demonstrated in this case TOE had a crucial role in the detection of the cardioembolic source. The management for asymptomatic patients remains controversial, though surgical excision is an accepted approach in symptomatic patients to prevent further embolic events.

Declaration of interest
The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this case report.

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Patient consent
Informed consent has been obtained from the patient for publication of the submitted article and accompanying images.

Author contribution statement
M M wrote the first and revised draft of the manuscript and acquired the images. Z E, V Z and J K critically appraised the manuscript. All authors approve of the submitted manuscript.

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