Unusual association of diseases/symptoms

A heart within the heart – a rare congenital cause of an embolic stroke

Tina Khan,¹ Samy El-Sharkawy,² Frank Schafer²¹Cardiology Department, Harefield Hospital, London, UK;²Cardiology Department, Barnet Hospital, London, UK

Correspondence to Dr Tina Khan, tinakhaled@hotmail.com

Summary

The authors report the case of an 87-year-old lady who presented with a right middle cerebral artery territory infarct in whom the authors diagnosed a probable ruptured blood cyst attached to her anterior mitral valve leaflet, with the use of trans-thoracic and trans-oesophageal echocardiography. Embolic strokes are recognised as a rare complication of intracardiac blood cysts, with very few cases previously reported. Echocardiography was first used to identify a blood cyst in 1983 and remains an excellent modality to diagnose this rare condition.

BACKGROUND

Intracardiac blood cysts are thin-walled cysts which are lined by cobble-stone shaped epithelium, filled with non-organised blood.¹ They were first reported by Elsasser in 1844² and are mostly congenital in origin. During autopsy, they are found on cardiac valves in approximately 50% of infants under the age of 2 months and are very rarely found after the age of 2 years.³

Although they are typically asymptomatic, rarely they can cause complications such as valvular dysfunction, left ventricular outflow tract obstruction and embolic stroke, for which there are very few documented cases.^{4–6} Trans-thoracic and trans-oesophageal echocardiography remain useful diagnostic tools in diagnosing this rare condition. We describe the case of an 87-year-old lady who presented with an embolic stroke in whom trans-thoracic and trans-oesophageal echocardiography enabled us to diagnose the presence of an underlying blood cyst attached to the mitral valve which appeared to have ruptured.

CASE PRESENTATION

An 87-year-old lady was admitted to hospital after waking up with new onset left-sided arm and leg weakness. She denied any chest pain, palpitations or breathlessness.

Her past medical history was only remarkable for sensori-neural deafness and previous urinary tract infections. There was no known history of cerebrovascular events or cardiac history and no known family history of cardiac conditions.

Physical examination revealed that she was afebrile with a heart rate of 65 beats per min, blood pressure of 120/60 mm Hg and a respiratory rate of 14 breaths per min with oxygen saturations of 98% on air. Upon neurological examination, she had left-sided weakness with power reduced to 2/5 in her left upper limb and 3/5 in her left lower limb. Her sensation was intact and her plantars were both downgoing. She was unable to mobilise. Her cardiovascular examination was normal, with normal jugular venous pressure, no carotid bruits and normal heart sounds with no added sounds.

INVESTIGATIONS

Laboratory values demonstrated normal blood count, biochemistry, liver function and clotting profile. Her fasting glucose and lipid profile were within normal limits. Her C reactive protein was elevated at 59 mg/l. Her urine dipstick was positive for nitrites, leucocytes and protein suggesting she had a urinary tract infection. Admission ECG showed normal sinus rhythm with no ischaemic changes and her chest radiograph showed normal cardiac contours with normal pulmonary vascular markings. CT of the head showed hypo-attenuation of the periventricular white matter compatible with chronic micro-vascular disease. There was a focus of encephalo-malacia involving the right caudate nucleus, anterior to the caudo-thalamic groove, compatible with a subacute infarct. Carotid artery dopplers did not demonstrate any significant stenosis. Her 24 h ECG demonstrated that she remained in sinus rhythm throughout with no arrhythmias.

A diagnosis of subacute right middle cerebral artery territory infarct was made and she was initially given aspirin 300 mg. She was not thrombolysed in view of her late presentation. She was treated for her urinary tract infection with intravenous piperacillin and tazobactam.

Subsequently she developed low-grade fevers with temperature spikes ranging from 37.5°C to 37.8. Numerous blood cultures were obtained from different sites on different occasions. Two of the bottles grew *Streptococcus viridans* and a third bottle grew *S sanguinis*. She had no new cardiac murmurs or clinical stigmata of bacterial endocarditis. Aside from the positive blood cultures, she did not fulfil any other Duke's criteria for bacterial endocarditis.

In order to exclude intracardiac thrombus as a potential cause for her cerebro-vascular infarct, as well as to exclude bacterial endocarditis, we performed a trans-thoracic and trans-oesophageal echocardiogram. The transthoracic echocardiogram demonstrated that her left ventricle was non-dilated with mild concentric hypertrophy with good systolic function, with an ejection fraction of 60–70%. She was found to have a cystic structure attached to the anterior mitral valve leaflet. This was further characterised

upon performing a trans-oesophageal echocardiogram, which confirmed the presence of a ring-like partially mobile cystic structure attached to the atrial base of the anterior mitral valve leaflet, measuring 1.7 cm squared in area (videos 1–3). The cyst was not causing any stenosis or significant regurgitation of the mitral valve. The impression of communication of the cyst with the intracardiac cavities by colour flow was then impressively confirmed via injection of sulphur hexafluoride contrast (video 4). The appearance was very suggestive of a ruptured blood cyst. There was no evidence of vegetations or any features of bacterial endocarditis. She had no intracardiac thrombus and no inter-atrial or inter-ventricular shunt was demonstrated.

Video 1 Close-up trans-oesophageal image of blood cyst attached to the anterior mitral valve leaflet.

Video 2 Trans-oesophageal colour flow image demonstrating communication of the cyst with the intracardiac cavity, suggesting rupture of the cyst (1).

Video 3 Trans-oesophageal colour flow image demonstrating communication of the cyst with the intra-cardiac cavity, suggesting rupture of the cyst (2).

Video 4 Injection of sulphur hexafluoride contrast confirming communication of the cyst with the intracardiac cavity, consistent with a ruptured cyst.

The presence of the blood cyst was confirmed upon performing a cardiac MRI which again showed a cystic structure measuring 15×15mm attached to the anterior mitral valve leaflet (figure 1).

In the absence of an alternative explanation for her ischaemic stroke and given that there is a reported association between intracardiac blood cysts and embolic events, we postulated that the rupture of this cyst with embolisation of thrombus from within it may have been the cause of her cerebrovascular event.

DIFFERENTIAL DIAGNOSIS

The echocardiographic features of blood cysts consist of an echo-free space surrounded by a thin wall. An important differential diagnosis ought to be a mitral valve leaflet aneurysm, but this tends to occur in the context of infective endocarditis of the aortic valve with significant aortic regurgitation,⁷ which was not present in this case. Differential diagnoses should also include myxoma, vegetation, hydatid cyst, thrombus or malignancy. However, these entities tend to present as echo-dense structures or structures with variable echogenicity.

TREATMENT

Surgical resection is the treatment of choice for blood cysts causing embolic strokes. However, in view of our patient's age, frailty and comorbidities, we felt that the risk of surgery would outweigh the benefits. Hence, in order to reduce her risk of further embolic events we commenced her on full anticoagulation with initiation of heparin and warfarin.

OUTCOME AND FOLLOW-UP

Her left-sided weakness has subsequently improved to 4/5 in her left lower limb only and she is now able to mobilise

Figure 1 Cardiac MRI showing attachment of the blood cyst to the anterior mitral valve leaflet.

with the aid of a walking frame. After 2 weeks of treatment with intravenous benzyl-penicillin and gentamycin her fevers fully resolved and her inflammatory markers normalised and subsequent blood cultures did not grow any organisms.

DISCUSSION

The first case of intracardiac blood cyst was reported by Elsasser in 1844.² In an autopsy series of foetal and infant hearts, blood cysts were found to be present in 56% of cases.⁸ The cysts regress spontaneously in most patients and are rare in adults. They are normally found on the pulmonary, tricuspid or mitral valves and are rarely found in the ventricles or atria. Echocardiography was first used to identify a blood cyst in 1983.⁹

Although complications of blood cysts are rare, they include valve dysfunction, left ventricular outflow tract obstruction and ventricular dysfunction.¹⁰ To the best of our knowledge, there have only been three previously published cases in the literature of embolic stroke felt to be caused by the presence of underlying intracardiac blood cysts.^{4–6}

We postulate that due to the relative stasis of blood within the cyst, thrombus develops, which can then travel into the intracardiac cavities via breaches in the walls of the cyst. In our case we were able to demonstrate the evident rupture of the mitral valve cyst with the use of sulphur hexafluoride contrast and colour flow. Particles of thrombi can then enter the systemic circulation, causing embolic events such as ischaemic strokes.

Due to the rarity of the condition, there are no clear guidelines regarding management of blood cysts. However, the general consensus is that in the context of embolic stroke, surgical resection should be considered.⁶ However, in this case we felt that our patient would not be a suitable surgical candidate and hence we opted to manage her medically with the use of anticoagulation. This strategy was used successfully in two of the previously reported cases of embolic stroke in the context of blood cysts.^{4–5} It has been suggested that asymptomatic cysts should be monitored with serial echocardiograms, in view of potential complications.⁴ Roberts *et al* proposed that the cyst should be removed routinely to exclude malignancy and to avoid the potential risk of embolism.¹¹

Learning points

- Intracardiac blood cysts are thin-walled cysts which are lined by cobble-stone shaped epithelium, filled with blood. They are mostly congenital in origin and are very rarely found in adults.
- Rarely they can cause complications such as valvular dysfunction, left ventricular outflow tract obstruction and embolic stroke, for which to date there are only three documented cases.
- Echocardiography was first used to identify a blood cyst in 1983 and remains an excellent modality to diagnose this rare condition.
- In the context of embolic stroke, surgical resection should be considered. However, if surgery is considered inappropriate, a strategy of anticoagulation may be adopted, as in our case example, to reduce the risk of future embolic events.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Burke A**, Virmani R. *Tumors of the Heart and Great Vessels. Atlas of Tumor Pathology*. Third Series. Fascicle 16. Washington, DC: Armed Forces Institute of Pathology 1996:80.
2. **Elsasser C**. Bericht über die Ereignisse in der Gebäreranstalt des Katherinen-Hospital im Jahre 1844. *Med Korrespondenzblatt* 1844;**14**:297.
3. **Boyd TA**. Blood cysts on the heart valves of infants. *Am J Pathol* 1949;**25**:757–9.
4. **Lodha A**, Patel J, Haran M, *et al*. Blood cyst of the mitral valve: a rare cause of stroke. *Echocardiography* 2009;**26**:236–8.
5. **Jacob JJ**, Jose J, John B. Intracardiac blood-filled cysts of the heart: a rare cause of embolic stroke. *Singapore Med J* 2007;**48**:e125–6.
6. **Kuvin J**, Saha P, Rastegar H, *et al*. Blood cyst of the mitral valve apparatus in a woman with a history of orthotopic liver transplantation. *J Am Soc Echocardiogr* 2004;**17**:480–2.
7. **Decroly P**, Vandenbossche JL, Englert M. Anterior mitral valve aneurysm perforation secondary to aortic valve endocarditis detected by Doppler colour flow mapping. *Eur Heart J* 1989;**10**:186–9.
8. **Begg JG**. Blood-filled cysts in the cardiac valve cusps in foetal life and infancy. *J Pathol Bacteriol* 1964;**87**:177–8.
9. **Hauser AM**, Rathod K, McGill J, *et al*. Blood cyst of the papillary muscle. Clinical, echocardiographic and anatomic observations. *Am J Cardiol* 1983;**51**:612–3.
10. **Arnold IR**, Hubner PJ, Firmin RK. Blood filled cyst of the papillary muscle of the mitral valve producing severe left ventricular outflow tract obstruction. *Br Heart J* 1990;**63**:132–3.
11. **Roberts PF**, Serra AJ, McNicholas KW, *et al*. Atrial blood cyst: a rare finding. *Ann Thorac Surg* 1996;**62**:880–2.

This pdf has been created automatically from the final edited text and images.

Copyright 2012 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Khan T, El-Sharkawy S, Schafer F. A heart within the heart – a rare congenital cause of an embolic stroke. *BMJ Case Reports* 2012; 10.1136/bcr.10.2011.5027, Published XXX

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow

Keep up to date with all published cases by signing up for an alert (all we need is your email address) <http://casereports.bmj.com/cgi/alerts/etoc>