Case report

Right atrial blood cyst of a patient with hypertrophic cardiomyopathy

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Received 17 May 2022; accepted 21 October 2022

Summary

Intracardiac blood cysts are generally observed in infants and regress spontaneously with time. Blood cysts are mainly located on the left side of the heart and atrioventricular valves. These findings are extremely rare in older children and adults. We report a rare case of a 45-year-old female patient with a cardiac blood cyst in the right atrium and hypertrophic obstructive cardiomyopathy. To the best of our knowledge, this is the first-ever case report in the literature of blood cyst and hypertrophic cardiomyopathy.

Seminars in Cardiovascular Medicine 2022; 28:5–9

Keywords: cardiac blood cyst, hypertrophic cardiomyopathy, multimodality imaging

Background

Intracardiac blood cysts are generally observed in new-borns and infants but regress spontaneously with time, a few months after birth. Blood cysts are mainly located on the left side of the heart and atrioventricular valves. These findings are extremely rare in older children and adults. Blood cysts are blood-filled diverticula surrounded by endothelium. The origin of cardiac blood cysts is not well understood. Complications of cardiac blood cyst depend on the location of the cyst, and can include syncope, embolism, valvular dysfunction, ventricular outflow tract obstruction, occlusion of a coronary artery, embolism, and/or death [1–4]. We present a rare case of a blood cyst in the right atrium of a patient with hypertrophic cardiomyopathy.

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Case report

A 45-year-old female patient presented to the hospital with chest pain on the left side of the sternum and recurring syncope. Physical examination revealed a systolic murmur on the left sternal region, irradiating to the left axillar line. The patient's blood pressure was 66/40 mmHg and pulse rate 77 beats/minute. Electrocardiogram showed sinus rhythm, left ventricular hypertrophy, inverted T-waves in leads from V1 to V5. Laboratory tests showed Hb levels of 63–75 g/l. The possible cause of severe anaemia was uterine myoma, which was diagnosed and scheduled for surgical treatment. Also, elevated cardiac damage markers were obtained: TnI 3789 ng/l, BNP 922.6 ng/l. The patient's chest X-ray was normal. Coronary angiography revealed non-critical plaques without severe stenosis in the right posterolateral branch and interventricular branch of the left coronary artery. A transthoracic echocardiogram (TTE) was performed for further differential diagnosis. TTE revealed mixed (apical and septal) hypertrophic cardiomyopathy (HCM) with significant left ventricular outflow tract obstruction, moderate mitral valve regurgitation and patent foramen ovale. Furthermore, a mobile 25 × 30 mm oval cystic mass attached above the tricuspid valve in the right atrium was ob-

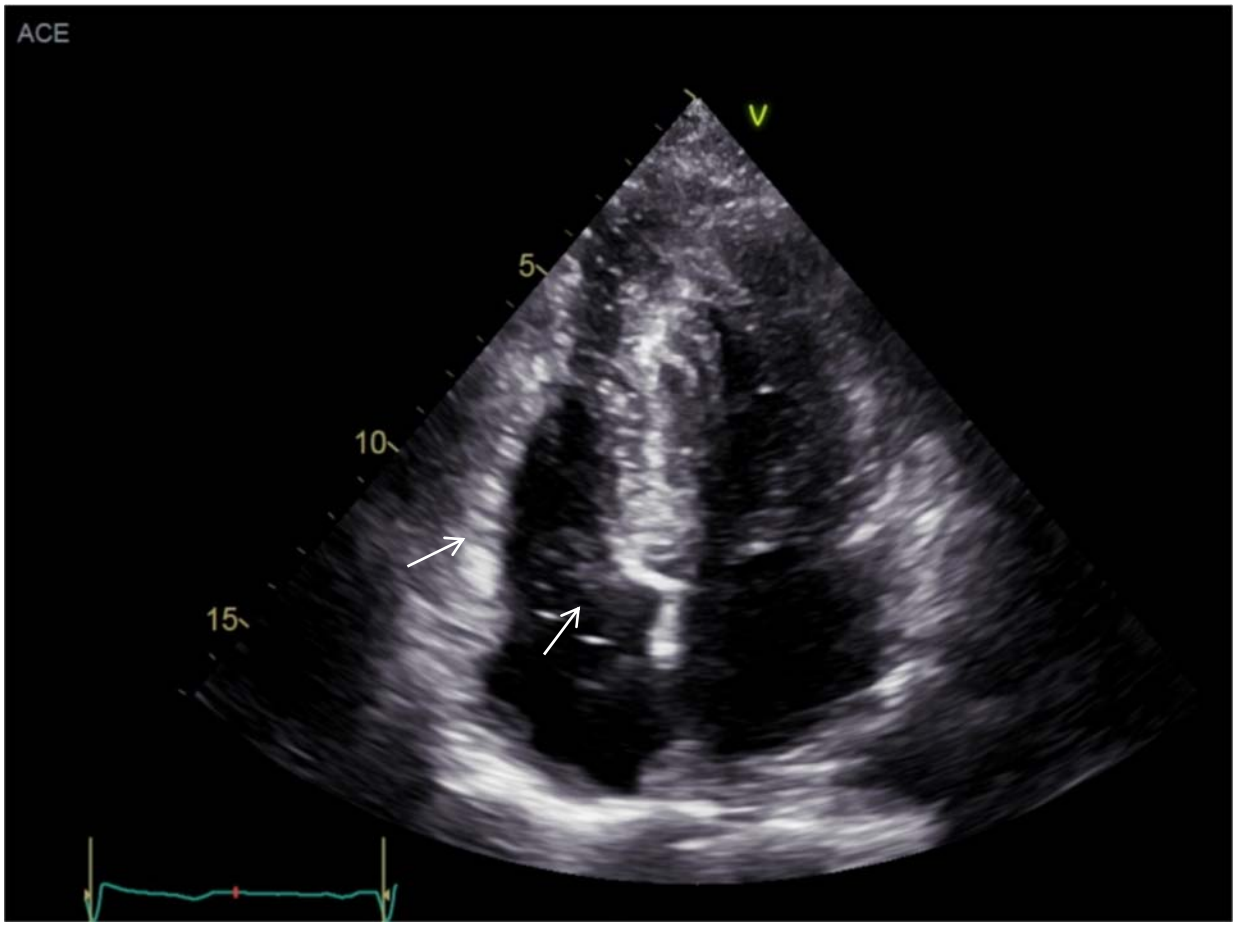

Figure 1. Transthoracic echocardiography image in apical 4-chamber view. In the right atrium, there is a thin-walled, oval-shaped cystic structure measuring ~2.5 × 3.0 cm, with a smooth surface and completely anechogenic content. During diastole, the mass moves through the tricuspid valve into the right ventricle. Left ventricular apical and septal hypertrophy is seen

served. Thin-wall, anechogenic, non-obstructive blood cyst-like structure prolapsed into right ventricular cavity during diastole in every heart cycle (Fig. 1). In addition, for better cystic structure visualisation, contrast-enhanced transoesophageal echocardiography (TEE) was performed (Fig. 2). After administration of contrast media, no uptake was observed. There was no tricuspid valve blood flow abnormality. Cardiac magnetic resonance imaging (MRI) confirmed the intracardiac mobile mass (Fig. 3). The right atrial mass demonstrated features suggestive of cystic nature of the mass. Combining multiple imaging techniques (TTE, TEE and MRI), atrial mass was most likely a blood cyst. 24-hour Holter monitoring was performed to determine the cause of syncope. Non-sustained ventricular tachycardia was observed. Calculated HCM-related sudden death risk for this patient was 11.4%. Due to the high 5-year risk of sudden cardiac death, ICD implantation was recommended. Because the cystic mass did not cause right ventricular outflow tract obstruction and did not interfere with the planned ICD lead implantation site, surgical removal of the mass was not performed. For symptomatic HCM with severe left ventricular outflow tract (LVOT) obstruction treatment with a maximum outflow tract gradient of 90 mmHg, septal reduction therapy was indicated. To improve symptoms, the percutaneous alcohol septal ablation was performed. After procedure, the LVOT gradient was reduced to 10 mmHg. As a result of alcohol septal ablation syncope did not recur, better tolerance of physical activity was achieved, but general weakness remained. Further treatment strategy was discussed by the multidisciplinary team. After evaluation of the patient, the team has decided to postpone ICD implantation and first do the follow-up due to multiple reasons. Firstly, the presence of a blood cyst repeatedly migrating between the right ventricle and right atrium. Placing the ICD lead through the tricuspid valve has a high risk to damage the cyst's capsule. That could result in paradoxical thromboembolic events through patent foramen ovale. Secondly, the usual location for screwing the ICD lead is the interventricular septum, which recently received alcoholic ablation. After these

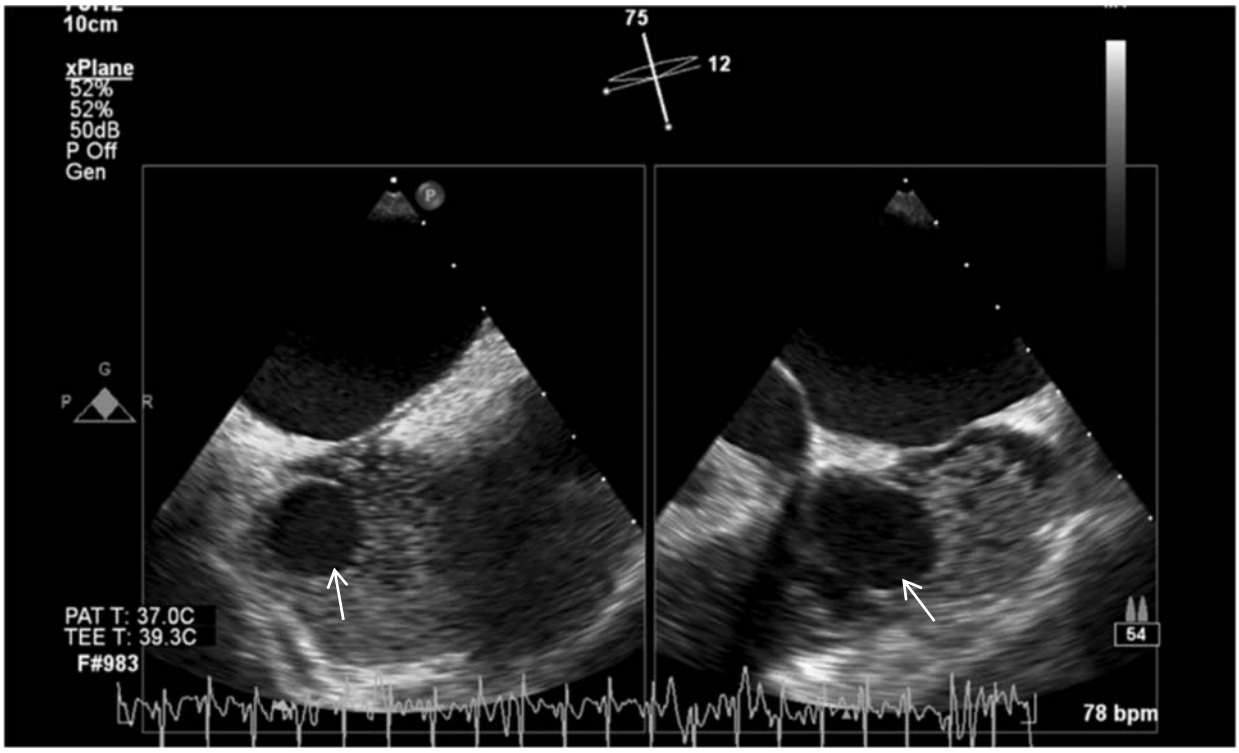

Figure 2. Contrast transesophageal echocardiography images in midesophageal modified bicaval view. The use of contrast highlights the oval-shaped cystic mass in the right atrium attached between the tricuspid valve annulus and the orifice of the inferior vena cava. The mass does not show contrast uptake

procedures, the scar formation and myocardial reorganization is a prolonged process of a few months. Implantation of the ICD might result in an increased pacing threshold and the need for additional surgery to further reposition the lead in the near future. For these reasons, follow-up after one month was planned. After health reassessment, the patient was registered for ICD implantation and one more outpatient visit until the procedure. The patient did not show up for a planned health condition evaluation and ICD implantation. The patient died, presumably, of out-of-hospital sudden cardiac death.

Discussion

Primary cardiac tumours are rare disorders. The incidence of cardiac tumours varies between 0.0017% and 0.028% [5]. Approximately 70% of cardiac tumours are benign. The incidence of cardiac blood cyst is reportedly 0.07%. Blood cysts of the heart were first described by Elsasser in 1844 [6]. Cardiac blood cyst is believed to be benign cardiovascular neoplasm. Intracardiac blood cysts are generally observed in infants and regress spontaneously with time. These findings are extremely rare in older children and adults representing 1.5% of intracardiac tumours [3]. The clinical symptoms of the patient are atypical. Blood cysts are usually small and may develop in any heart cavity. Complications of cardiac blood cyst depend on the location of the cyst. Blood cyst can cause syncope, embolism, valvular dysfunction, ventricular outflow tract obstruction, occlusion of a coronary artery, embolism, and/or death. The differential diagnosis of a blood cyst includes myxoma, vegetation, hydatid cyst, cardiac malignancies, abscess, and thrombus. Almost in all reported cases of cardiac blood cysts have involved the valvular apparatus or papillary muscle of the tricuspid, pulmonary, or mitral valve. In the current literature we found fifteen cases of cardiac blood cysts located in the right atrium [1–3,7–18]. There are several exceptional clinical cases of blood cyst with total occlusion of the right coronary artery [14], double blood cyst [18], blood cyst with ischaemic heart disease [10], blood cyst with simultaneous fibroelastoma [1]. To our best knowledge, a case of a cardiac blood cyst with HCM as in our case has never been previously reported.

The origin of cardiac blood cysts is not well understood. Various hypotheses have been proposed about the development of such cysts. One theory suggests that blood cysts are formed during valve development by blood being compressed in crevices that are later sealed off [19]. Another theory suggests that blood cysts are the result of heteroplastic change in tissues originating from the

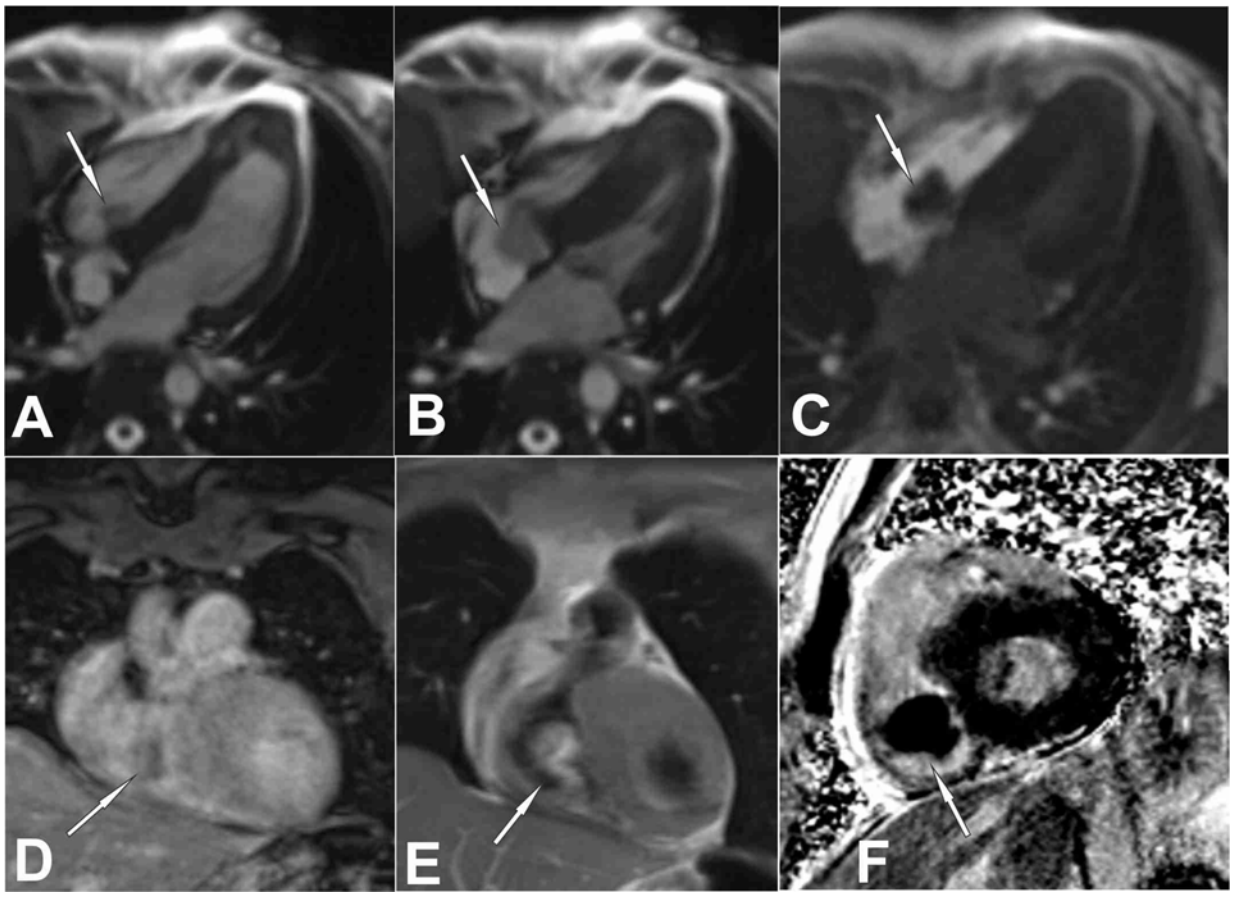

Figure 3. Cardiac MRI images. Cine 4 chamber heart views in diastole (A) and in systole (B) depicted the mass (arrow) and its relationship with the tricuspid valve, marked asymmetrical septal and apical hypertrophy is clearly seen. Perfusion 4-chamber heart view (C) does not show mass uptake of gadolinium (arrow) during the first pass perfusion. On the contrast-enhanced fat-saturated T1-weighted vibe sequence (coronal plane) the mass (arrow) showed a signal isointense compared with myocardium (D) and hyperintense signal on coronal T2 haste image (E). On the late gadolinium enhancement sequence (inversion recovery) the mass (arrow) showed no contrast enhancement, suggestive of the cystic nature of the mass (F)

primitive pericardial mesothelium [20]. Sakakibara et al. suggested injury-related hypothesis, they suggested that hypoxia, inflammation, and bleeding tendencies are possible mechanisms of the blood cysts formation [21]. It is likely that the true aetiology of cardiac blood cysts is a combination of several mentioned mechanisms.

Multimodality imaging by echocardiogram, including transoesophageal contrast echocardiography, and magnetic resonance imaging is useful to make a diagnosis. Echocardiography is useful in detecting and examining intracardiac tumours [22,23]. A thin-walled cystic mass with an echo-free space within the lumen can be distinguished from a solid tumour. Two-dimensional echocardiography provides information about the size, location, movement, and characteristics of blood cyst. Transthoracic and transoesophageal echocardiography can increase the diagnostic accuracy and transoesophageal echocardiography is sensitive to small cysts less than 5 mm [22]. Magnetic resonance imaging is the most effective tool for the differential diagnosis of solid tumours. A blood cyst may be suspected when a round homogeneous image is observed with no uptake of intravenous contrast medium, which indicates a hematic and cystic nature [18]. MRI is a powerful tool for tissue characterization of cardiac masses, because of case rarity, differentiating blood cyst from myxoma or metastases based on its results is difficult. In this case multimodality imaging was a key feature in differential diagnosis. Most cases described in literature made diagnose of blood cyst after surgical cyst removal and histological examination. In our case diagnosis was made based on typical blood cyst features in different imaging modalities described in the literature [24].

There is no consensus or guidelines about the optimal management of cardiac blood cysts. There are two different opinion sides of the optimal management of symptomatic or complicated blood cyst in asymptomatic patient. Surgical resection in all patients to prevent complications such as emboli and exclude malignancy is recommended by most of the authors [1,3,8,10,

14]. Other authors prefer surgical operation in symptomatic patients with cardiac blood cysts, because of benign nature of the tumour [2,12, 17]. Although the indications for surgical intervention include progressive increase in mass size, obstruction of flow, or embolization [7,23]. In our patient, we found a cystic lesion without hemodynamic effects of tricuspid valve, and we opted for imaging follow-up.

Conclusions

We report a rare case of a 45-year-old female patient with a cardiac blood cyst in the right atrium with hypertrophic obstructive cardiomyopathy. To the best of our knowledge, this is the first-ever case report in the literature of blood cyst with hypertrophic cardiomyopathy. Characterisation of an atrial blood cyst using multimodality imaging is recommended. There is no consensus on the optimal management of cardiac blood cysts. Cardiac blood cysts are rarely detected preoperatively and should be included in the differential diagnosis of cardiac masses.

Consent

Consent was obtained from the hospital to use anonymized patient information.

Conflict of interest

The authors declare that they have no conflict of interest.

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