Pericardial Cyst : A Rare Cause of Pericardial Effusion

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SUMMARY

Pericardial cysts occur rarely, with an incidence rate of 1 per 100,000. They are usually detected by chance and clinically silent in most cases. Pericardial cysts are the most common benign tumours of the pericardium and presents by the third or fourth decade of life, and equally common in males and females. In principle, they only require follow-up, however, an enlarging or symptomatic cyst requires surgical removal. We report a case of a 32 year-old Malay lady, who presented with history of recurrent pericardial effusion followed by right pleural effusion. Computed tomography (CT) thorax identified a large mediastinal cyst as the cause of her problem, requiring exploratory thoracotomy.

KEY WORDS:
Pericardial cyst, pericardial effusion, pleural effusion, mediastinal cyst

INTRODUCTION

Pericardial cysts are rare benign lesions, often asymptomatic and usually found incidentally on routine chest radiography. They are the most common benign tumours of the pericardium. They are usually identified on the third or fourth decade of life and are equally common in males and females. In principle, a pericardial cyst only requires follow-up, however, growing cases or symptomatic cases require surgical removal.

CASE REPORT

A 32 year-old Malay lady, presented with history of chest pain and dyspnoea. An echocardiography revealed significant pericardial effusion with no evidence of cardiac tamponade, requiring pericardial tapping, which later caused complete resolution of symptoms. Three months later, she presented again with similar symptoms. Postero-anterior chest radiograph showed right pleural effusion and once again echocardiography demonstrated recurrent pericardial effusion. Both effusions were drained; all cultures and specimens sent were unremarkable.

Subsequent computed tomography (CT) thorax (Figure 1) revealed a well-defined homogenous cystic lesion arising from the right side of the mediastinum, in close proximity of the large vessels and right heart border, pushing the heart to the left side. The problem recurred eight months later; thus a repeated CT thorax was done. The cyst has increased in size, causing mass effect to the mediastinal structures which shifted the mediastinum to the left.

Since the cyst was enlarging and causing worsening of her symptoms, she was referred for surgical intervention. Preoperative echocardiography showed good ejection fraction of 61%, mild left ventricular hypertrophy with loculated pericardial effusion at anterior right atrium, posterior left atrium and apical around 0.5 cm each. The right atrium looks compressed by an extra cardiac mass (Figure 2).

An exploratory right thoracotomy was performed with double lumen intubation. Intra-operatively, she was noted to have a huge cyst compressing the upper, medial and lower lobes of the right lung, extending from the apex to the diaphragm. The cyst wall was thick and well formed. The cyst contained 700cc of brownish colour fluid with brown coloured cheesy necrotic tissue within it. The medial cystic wall adhered to and compressed the right atrium, and was released intra-operatively. The superior cystic wall adhered densely to part of the ascending aorta and superior vena cava, and was laid opened. The inferior cystic wall adhered to the diaphragm. The cyst, thought to be pericardial, was finally excised.

Histopathology examination demonstrated that the cyst has thick fibro-collagenous wall with no apparent lining epithelium, necrotic debris is seen attached to the inner surface (Figure 3). It was reported as infected benign pericardial cyst.

DISCUSSION

Pericardial cysts are rare mediastinal cysts [1] with an incidence of approximately 1 in 100,000 [2, 3] and constitute 7% of all mediastinal lesions [2]. They are the most frequent benign tumours of the pericardium and generally they are of congenital origin [1]. However, some believe that it may be acquired, as a result of inflammation or injury [3]. They are often discovered on routine chest radiograph of asymptomatic adults [1, 2, 4], in the third or fourth decade of life. They are usually benign cystic lesion, but must be differentiated from other lesions, often requiring an exploratory thoracotomy. There is no report of malignant transformation [1, 2].

Although the majority of patients are asymptomatic, about one third exhibit symptoms [1, 5] when they reach a large size. The most common symptoms are chest pain, dyspnoea and cough [1, 4, 5]. However, complications can be life threatening, including cardiac compression [4], right ventricular outflow tract obstruction, cyst rupture with cardiac tamponade, cyst infection with cardiac or large vessel erosion, atrial fibrillation and even sudden death [1, 2, 5, 6].

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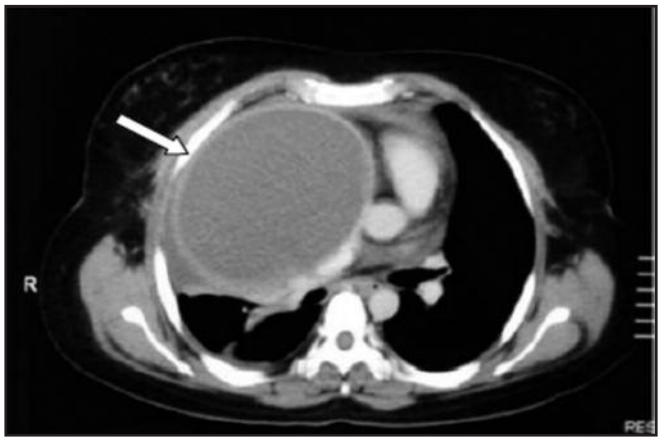

Fig. 1 : Computed Tomography demonstrating a pericardial cyst (arrow), shifting the mediastinum to the left.

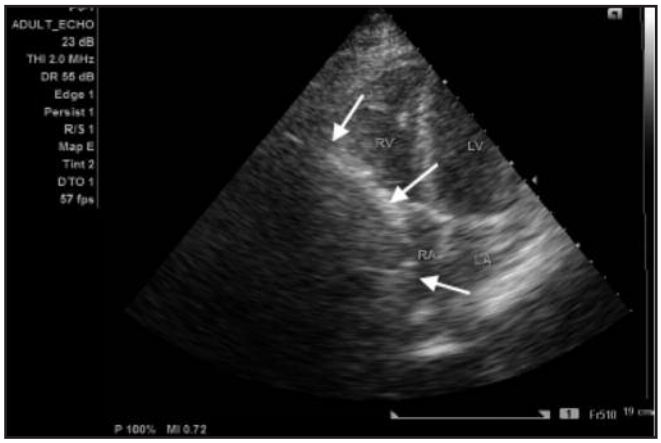

Fig. 2 : Transthoracic echocardiogram showing extra cardiac mass (arrow), compressing the right atrium.

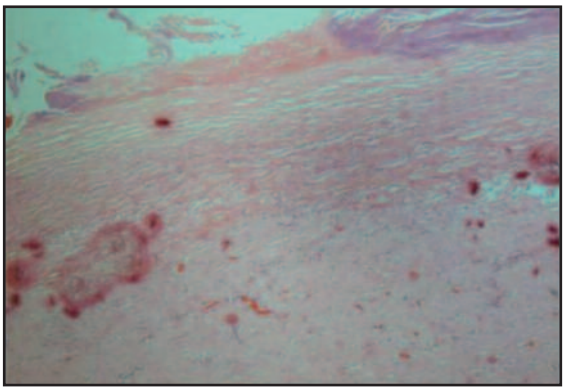

Fig. 3 : Section shows thick fibro-collagenous cyst wall devoid of epithelial lining, with attached fibrinous exudate. No garuloma noted. (H&E x 4 objective).

Most pericardial cysts are situated at the right cardiophrenic angle (70%) [1, 2, 5, 6]. However they can occur throughout the mediastinum such as the left cardiophrenic angle (22%), or in the posterior or anterior superior of mediastinum (8%) [6].

The imaging studies useful for diagnosis of pericardial cysts are chest radiography, echocardiography, CT, and magnetic resonance imaging (MRI) of the chest [5]. A pericardial cyst is usually suspected because of abnormal findings on chest radiography. They appear as well-defined round or oval masses in contact with the heart. Transthoracic echocardiography is helpful in showing the exact location of the cyst and in differentiating a cyst from other entities, such as a fat pad, aneurysm, or solid tumour [5]. On CT and MRI, a pericardial cyst typically appears as a non-enhanced, oval, well-defined homogenous mass adjacent to the pericardium. They provide detailed anatomical description of pericardial lesion and are useful during preoperative assessment as they can evaluate associated extra cardiac disease [6].

Generally, close follow up is sufficient in asymptomatic pericardial cyst. Treatment is required in symptomatic patients, a large size cyst or in those with an unclear diagnosis[1, 2]. Treatment options include surgical resection or percutaneous aspiration of the cyst [5].

In this case, our patient presented with history of recurrent pericardial and pleural effusion with no clinical evidence of cardiac tamponade. The cyst has thick fibro-collagenous wall and filled with brownish fluid. There was no apparent epithelial lining with necrotic debris is seen attached to the inner surface. Considering the location of the cyst found intra-operatively, it is likely that this was originally a pericardial cyst whose epithelial lining was destroyed by inflammation. It is believed that the patient had developed pericarditis that later caused inflammation of the cyst. The inflammed cyst which abutted the parietal surface of the pleura, led to pleural effusion. Inflammatory process resulted in progression of both the pericardial and pleural effusions, and the enlarging inflammed cyst producing her symptoms. There were two reported cases of pericardial cyst presented with pericardial effusion complicated with cardiac tamponade. These authors hypothesized that a small patent connection was established between the cyst and pericardium, causing progressive pericardial effusion[4]. Later this connection was spontaneously obliterated and sealed itself off from the pericardial space by fibrosis [4].

CONCLUSION

In conclusion, although it is rare, large pericardial cyst should be considered in the differential diagnosis of pericardial and/or pleural effusion after the more common causes have been excluded.

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