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## CASE REPORT

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# Primary Pericardial Osteosarcoma in an Elderly Patient

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### ABSTRACT

*Primary pericardial osteosarcoma is exceedingly rare, with only a few reported cases in the literature. We describe the clinical, radiological, and pathological features of a left primary pericardial osteosarcoma in a 72-year-old man.*

*Key Words:* Cardiovascular diseases; Magnetic resonance imaging; Osteosarcoma; Pericardium; Tomography, spiral computed

## 中文摘要

### 一名原發性心包骨肉瘤老年病例

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原發性心包骨肉瘤極其罕見，文獻中只有寥寥數個病例報告。本文報告一名原發性左心包骨肉瘤的72歲男性病例，並討論其臨床、影像和病理特徵。

### INTRODUCTION

Primary cardiac tumours are rare, with an estimated prevalence of about 0.002% to 0.3% in large autopsy and echocardiographic series.<sup>1,2</sup> Approximately 75% of all primary cardiac tumours are benign, of which myxoma is the most common tumour in adults. Malignant tumours account for 25% of all primary cardiac neoplasms, and approximately 95% of which are sarcomas.<sup>3,4</sup>

Tumours arising primarily from the pericardium are even less common than primary cardiac tumours, with an estimated prevalence of about 0.001% to 0.007%.<sup>5</sup> Primary pericardial osteosarcoma is extremely rare, with only few reported cases in the literature.<sup>6,7</sup>

This report describes the clinical, radiological, and pathological features of a left primary pericardial osteosarcoma in a 72-year-old man.

### CASE REPORT

In July 2014, a 72-year-old man with a history of hypertension and diabetes was admitted to Queen Elizabeth Hospital, Hong Kong for fever with cough and sputum production. He also had poor appetite, weight loss, and decreased exercise tolerance with shortness of breath in the previous year. His vital signs and blood biochemistry were otherwise unremarkable.

Chest radiograph (Figure 1) showed an enlarged cardiac silhouette with left pleural effusion. There was no

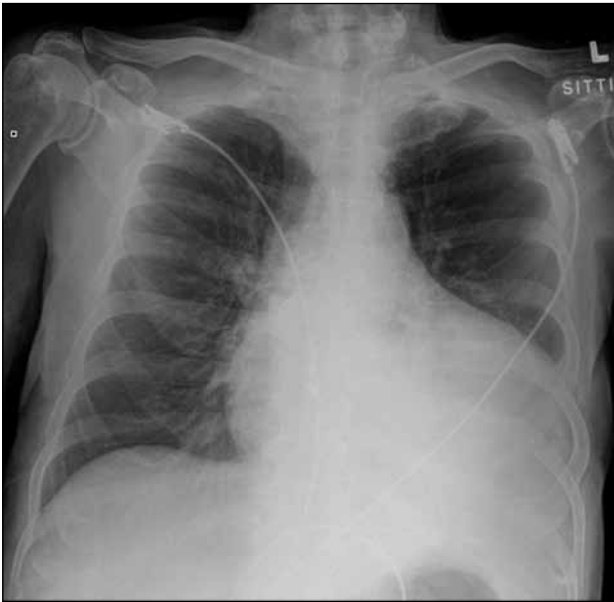
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Submitted: 6 Jan 2015; Accepted: 13 Mar 2015.

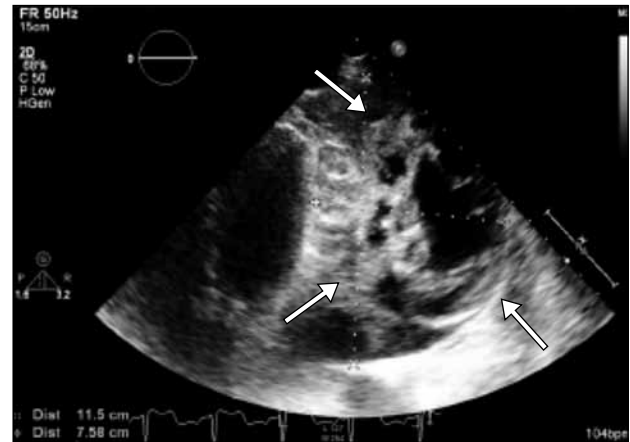


**Figure 1.** A posteroanterior chest radiograph of a 72-year-old patient with an enlarged cardiac silhouette and left pleural effusion.

lung mass or consolidation. The imaged skeleton was normal. The patient was treated for chest infection and given antibiotics (Augmentin 375 mg 3 times a day; GlaxoSmithKline, London, UK). Left pleural tapping and pleural biopsy showed blood-stained fluid with an exudative nature. The adenosine deaminase level was not elevated. Pleural fluid cytology and biopsy results were negative for malignancy. The pleural culture was negative.

Echocardiography was performed to investigate for suspected left pericardial effusion due to globular heart enlargement on chest radiograph. Ejection fraction was normal (62%). No pericardial effusion was detected. However, a large multiloculated cystic mass with solid component measuring up to 11.5 cm was detected adjacent to the left ventricle (Figure 2).

Contrast computed tomography (CT) of the thorax was performed to further delineate the periventricular mass lesion (Figure 3). A large (approximately 12 cm) mixed solid and cystic lesion was noted closely abutting the left pericardium. The solid component showed heterogeneous contrast enhancement. Multiple enhancing septae and blood-fluid levels were seen within the mass lesion. A small calcified focus was noted in the posterior aspect of the mass. The mass compressed and displaced the left atrium and ventricle.



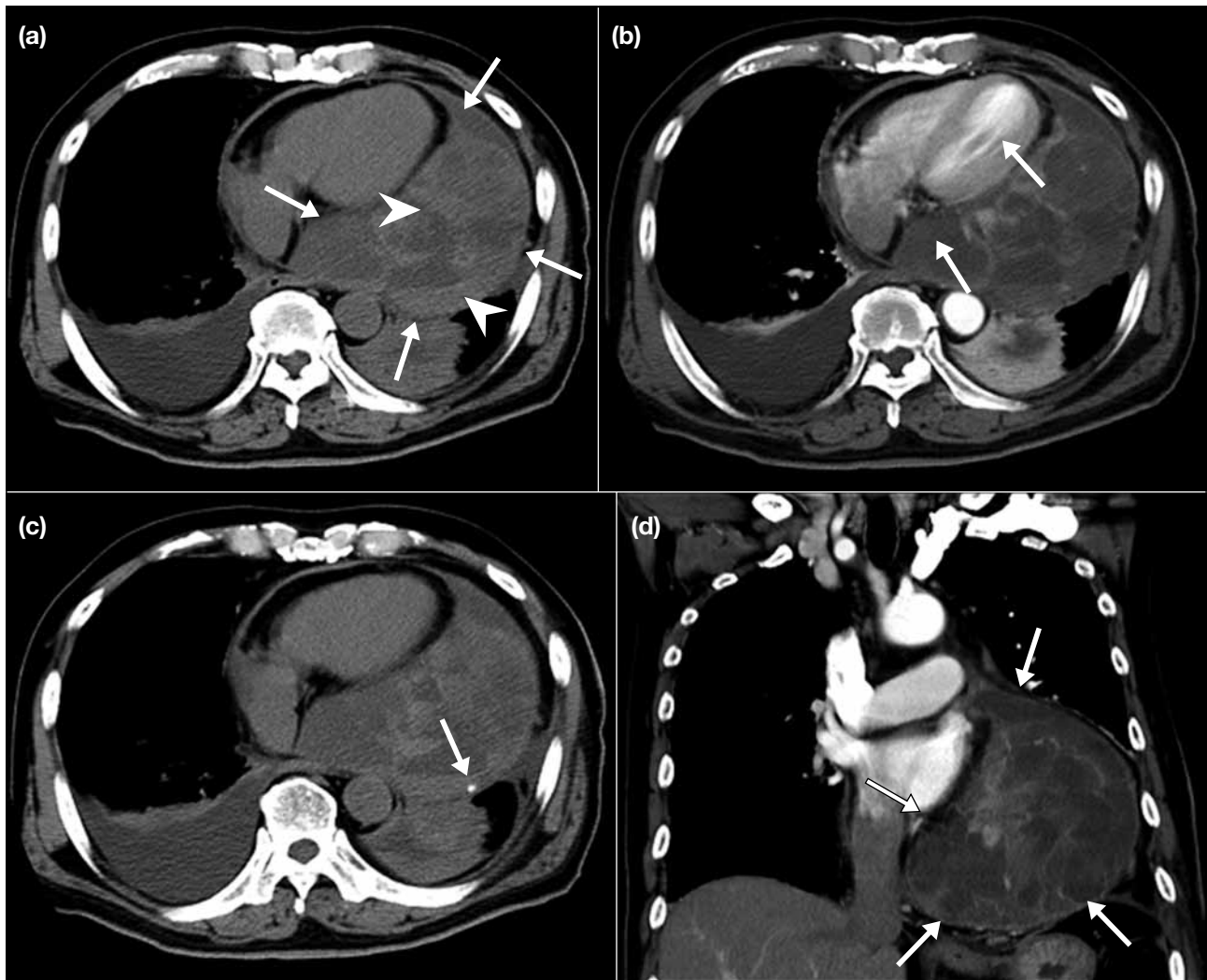
**Figure 2.** An echocardiogram shows a large multicystic lesion with solid component in the left periventricular region (arrows). No pericardial effusion is noted. The left lateral ventricular wall is hypokinetic.

There was no invasion into the heart chambers. The pulmonary arteries and veins were patent with no sign of invasion. No pericardial effusion was detected. No lung mass or enlarged mediastinal, hilar, or supraclavicular lymph nodes were seen. No destructive bone lesion was detected in the imaged skeleton.

Magnetic resonance imaging (MRI) of the heart was subsequently performed to better define the anatomical relationship (Figure 4). A large multicystic mass with solid component was noted arising from the left pericardium extending to the lateral and posterior aspects of the mediastinum. The mass measured approximately 13 cm in its greatest dimension. Heterogeneous signal intensity was noted on T1- and T2-weighted and cine true fast imaging with steady-state precession sequences.

There were multiple cystic areas with thin septae and irregular walls. No definite internal fat was seen. Multiple fluid levels were also noted, which may have been altered blood products corresponding to the CT findings of blood-fluid levels. The septae and solid areas showed heterogeneous contrast enhancement in delayed images. There was significant mass effect on the lateral wall of the left ventricle and atrium. There was no invasion into the cardiac chambers. The lateral ventricular wall was severely hypokinetic. No pericardial effusion was detected.

The diagnosis was a complex cystic mass with unknown nature, and elective operation for removal of the mass

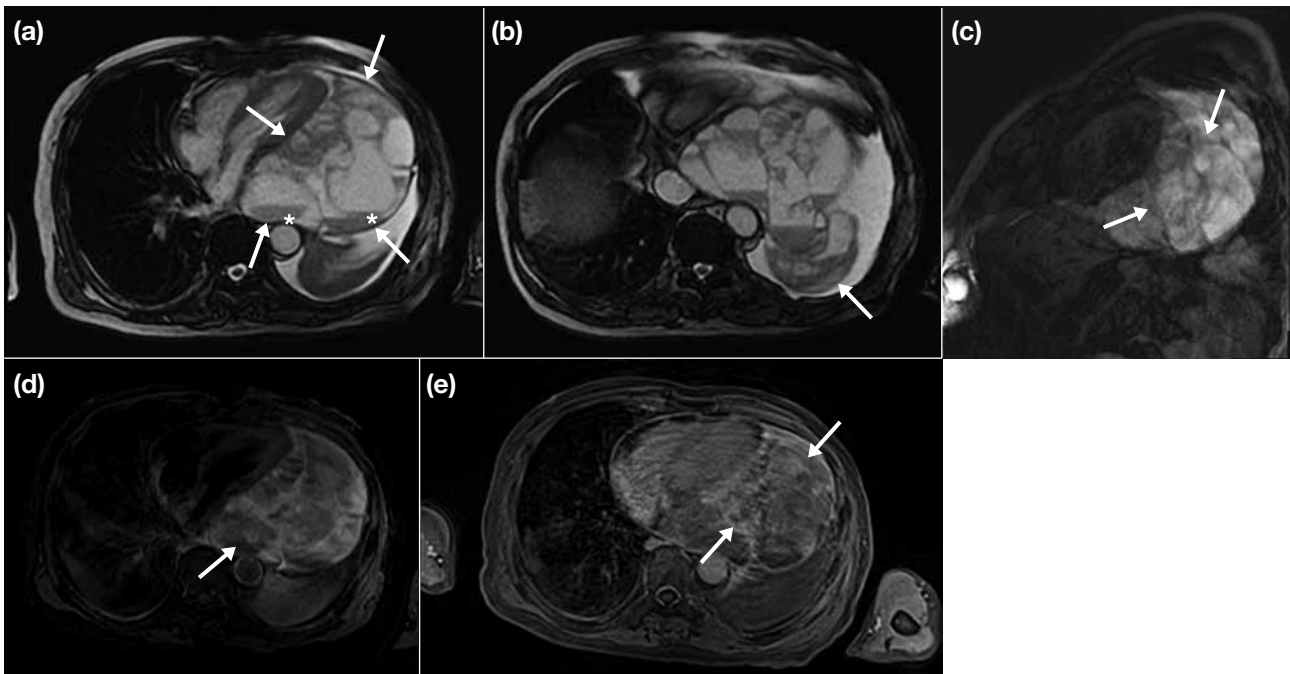


**Figure 3.** Computed tomography images of the thorax. (a) An unenhanced image shows a large multicystic mass (arrows) with solid component abutting the left ventricle — blood-fluid levels can be seen (arrowheads); (b) a contrast-enhanced image shows enhancement of the septae and solid component — the left ventricle and left atrium are compressed (arrows); (c) a small calcification (arrow) is noted in the posterior aspect of the multicystic mass lesion; and (d) a contrast-enhanced coronal image of the same patient shows a large multicystic mass (arrows) with solid component abutting the left ventricle.

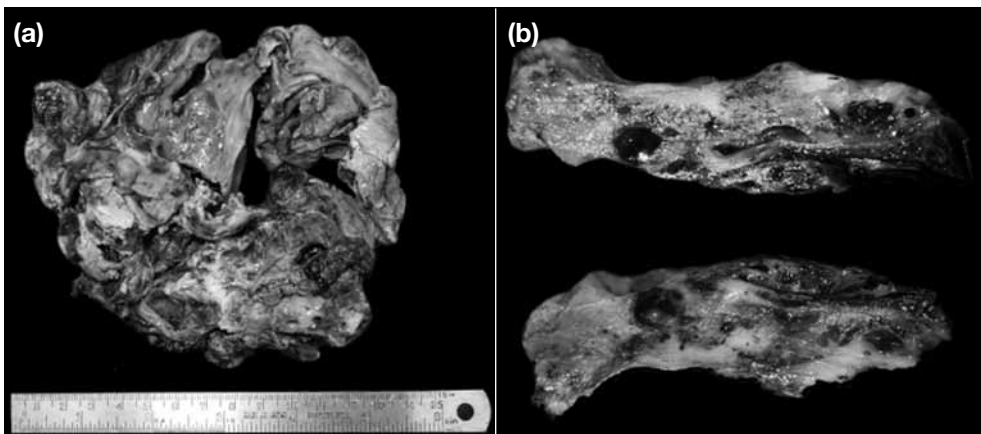
was planned for the patient. His condition, however, deteriorated steadily and he had a cardiac arrest, possibly due to the effect of the mass on the left ventricle. Emergency operation was arranged for excision of the left pericardial mass after initial stabilisation. A tumour arising from the parietal pericardium near the left ventricular apex was confirmed. A large amount of organising blood clot was noted inside the pericardial sac. Intrapericardial adhesions were noted.

The patient died after the surgery due to hospital-acquired pneumonia. However, a diagnosis of high-

grade osteosarcoma was confirmed histologically. Gross examination showed multiple fragments of tumour tissue, with the largest piece measuring 12 x 12 x 3 cm. Cut surfaces showed solid tan-coloured tumour with focal haemorrhagic cystic change (Figure 5). Microscopic examination showed a tumour consisting of markedly pleomorphic spindle-to-polygonal tumour cells, occasional osteoclast-like giant cells, and focal osteoid formation. Frequent telangiectatic areas with haemorrhagic cystic change were also seen. Carcinoma or a well-differentiated liposarcomatous component was not identified (Figure 6).



**Figure 4.** Magnetic resonance images of the thorax. T2-weighted true fast imaging with steady-state precession transverse image (a) shows the left pericardial mass with multiple cystic areas (arrows) and fluid-fluid levels (asterisks) — the left atrium and left ventricle are compressed, but are not involved and (b) at a lower level shows the pericardial mass extending laterally and posteriorly — the solid component shows heterogeneous signal intensity (arrow); (c) in T2-weighted fat-saturated sagittal image, the tumour shows heterogeneous signal intensity (arrows); (d) in T1-weighted fat-saturated transverse image, the tumour shows heterogeneous signal intensity (arrow); and (e) T1-weighted VIBE contrast-enhanced image with fat saturation shows heterogeneous contrast enhancement in the solid component of the lesion (arrows).



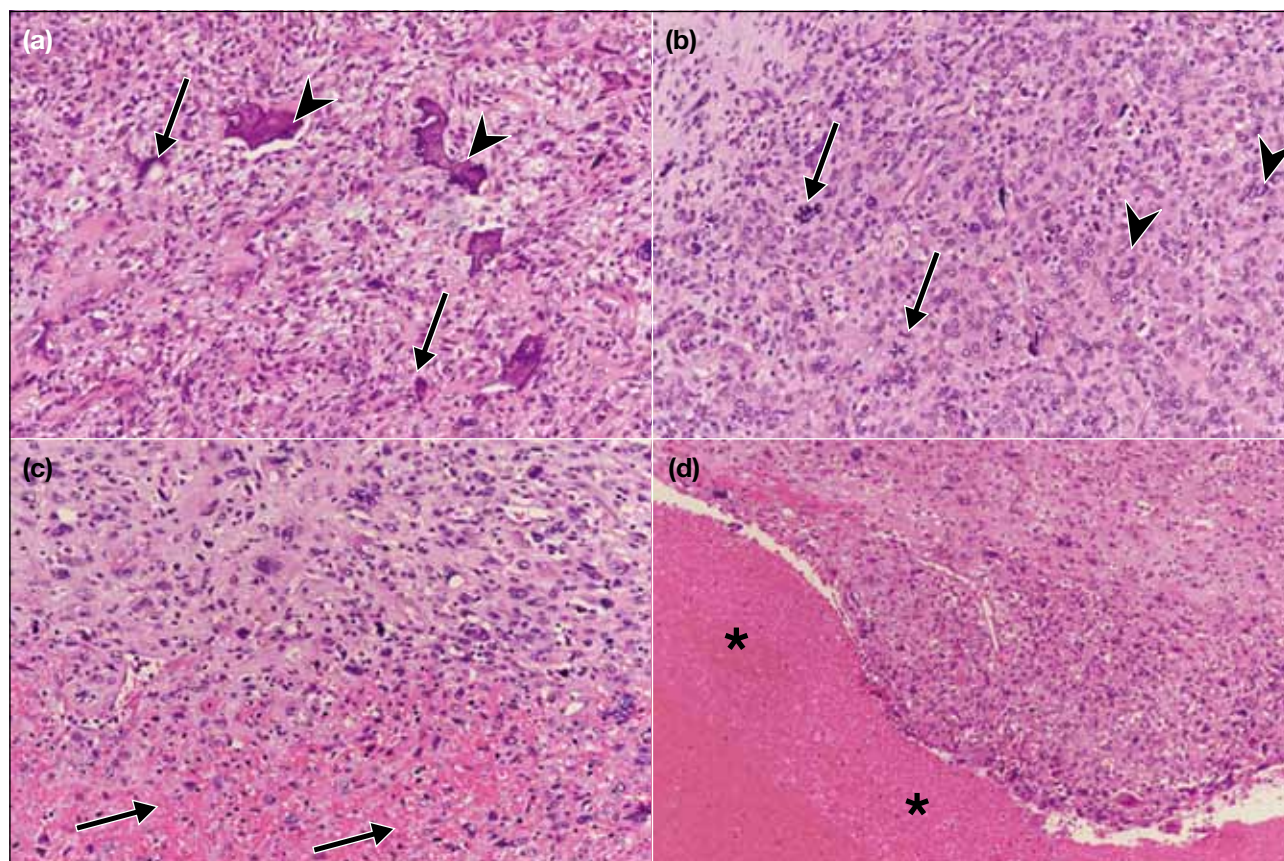
**Figure 5.** Surgical specimen. (a) Gross specimen; and (b) tumour cut surfaces with solid tan-coloured appearance and focal haemorrhagic cystic change.

## DISCUSSION

Primary cardiac tumours are rare, and tumours arising primarily from the pericardium are even less common. The most common benign pericardial tumour is pericardial cyst, followed by lipoma. The most common primary pericardial malignancy is mesothelioma followed by a variety of sarcomas and lymphoma.<sup>5</sup>

Metastatic involvement of the heart and pericardium is more common than primary cardiac tumours.

Primary pericardial osteosarcoma is extremely rare. In 1984, Schneiderman et al<sup>6</sup> described a patient with right ventricular epicardial osteosarcoma with correlation to technetium bone scan and gallium scan. In 2013, Wang



**Figure 6.** Microscopy images show (a) tumour with markedly pleomorphic spindle / polygonal cells (arrows), with focal areas of osteoid formation (arrowheads); (b) tumour with spindle and polygonal markedly pleomorphic cells (arrowheads), together with atypical mitoses (arrows); (c) tumour with haemorrhage (arrows); and (d) area of tumour with haemorrhagic cystic change (asterisks) [H&E; original magnification: (a to c) x 40, (d) x 20].

et al<sup>7</sup> described a patient with pericardial osteosarcoma originating in the left side of the heart posteriorly with positron emission tomography–CT correlation. The tumour was found to be heavily calcified.

Most of the reported cases of primary cardiac osteosarcoma showed a predilection for the left atrium.<sup>8,9</sup> A literature search by Takeuchi et al<sup>10</sup> in 2007 revealed 36 cases of primary cardiac osteosarcoma worldwide. The prognosis of cardiac osteosarcoma is generally poor. The presenting symptoms of cardiac tumours generally depend on tumour size, location, and presence of distant metastasis.

This patient initially presented with non-specific chest and constitutional symptoms. Later, he developed cardiac arrest presumably as a result of mass effect onto the left ventricle. He had no history of other malignant disease. Radiologically, the echocardiography, CT, and MRI findings of multicystic mass with blood-fluid

levels were consistent with the histological findings of frequent telangiectatic areas with haemorrhagic cystic changes. The presence of markedly pleomorphic spindle / polygonal cells, with focal areas of osteoid formation, confirm the diagnosis of osteosarcoma. It is noted that necrosis and haemorrhage are common in extraskeletal osteosarcoma and that the tumour tends to be of high-grade spindle malignancy.<sup>11</sup>

Heterogeneous signal intensities are noted on T1- and T2-weighted imaging on MRI, probably secondary to underlying haemorrhage and complex tumour constituents. Only a tiny focus of calcification is noted on CT imaging which is different from conventional osteosarcoma seen in appendicular or axial skeleton in which coarse and more extensive calcifications are more commonly seen.

Heterogeneous contrast enhancement is noted on both CT and MRI imaging. There were no other lytic bone

lesions detected in the radiographs and CT images and the patient did not have any abnormal mass or pain in the extremities and other regions to suggest a metastatic origin.

Cross-sectional imaging is useful for investigation of suspected pericardial mass, and plays an important part in differentiating a mass of myocardial or pericardial origin. MRI and CT can delineate the full extent of the pericardial mass, to look for myocardial invasion, involvement of cardiac and mediastinal vasculature, adjacent lungs, and any evidence of distant metastases.

The diagnosis of pericardial cyst and lipoma can readily be made by imaging. However, histological proof is usually needed for diagnosis of other pericardial tumours as they can have non-specific imaging findings. Complications associated with pericardial mass such as pericardial haemorrhage, pericardial effusion, or mass effect can also be made by imaging. The diagnosis of cardiac tamponade is, however, usually clinical and imaging is rarely indicated.

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