Cardiac Imaging

# Primary Heart Angiosarcoma

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Malignant primary cardiac tumours are rare and usually fatal. The diagnosis is difficult and usually late. We present two patients with primary cardiac angiosarcoma in whom echocardiography suggested the diagnosis, since cardiac tumours with an obviously rich blood supply were recorded.

Case 1 was a 32-year-old male patient with shortness of breath and fatigue over a 3-month period. Case 2 was a 56-year-old male with shortness of breath, haemoptysis and low-grade fever during the last 15 days. Both had histories of massive pericardial effusion 6 and 8 months prior to admission and pericardiocentesis was performed. Histological analysis of the bloody pericardial fluid was cytologically negative for malignant cells, while the chest computed tomography (CT) in each case revealed no abnormal findings at this time.

On admission, the echocardiogram in Case 1 showed a mass attached to the lateral free wall of the left ventricle (LV) and the colour Doppler further recorded blood flow within the mass (Figure 1). A subsequent CT revealed that the mass had invaded the LV. The patient died of excessive bleeding during an open chest biopsy of the tumour. *Post mortem* histological examination showed multiple anastomosing vascular channels lined with malignant endothelial cells, indicative of angiosarcoma.

In the second case, the echocardiogram showed a large mobile mass arising from the right atrial free wall (Figure 2), while colour Doppler recorded blood supply to the mass, probably from the right coronary artery (Figure 3). Subsequent CT revealed multiple secondary metastases in both lungs and liver. The histological examination of an open lung biopsy of a secondary metastasis once again diagnosed an angiosarcoma.

Primary tumours of the heart are rare and necropsy studies show incidences between 0.0017-0.28%.[1] However, only 10-20% of all primary cardiac tumours are malignant[2,3] and the most common histological type is angiosarcoma. Primary heart angiosarcomas are composed of malignant cells that form vascular channels and involve almost exclusively the right atrium but, as in our Case 1, have also been reported in other cardiac chambers.[4-6] They may be associated with dyspnoea, thoracic pain, general fatigue, or symptoms of right heart failure, cardiac tamponade and haemopericardium.[4] Mean survival is less than one year after diagnosis and patients respond poorly to chemotherapy.

The diagnosis of angiosarcoma is usually difficult and late, despite the different methods used. The patients described in this report had a very rare presentation,

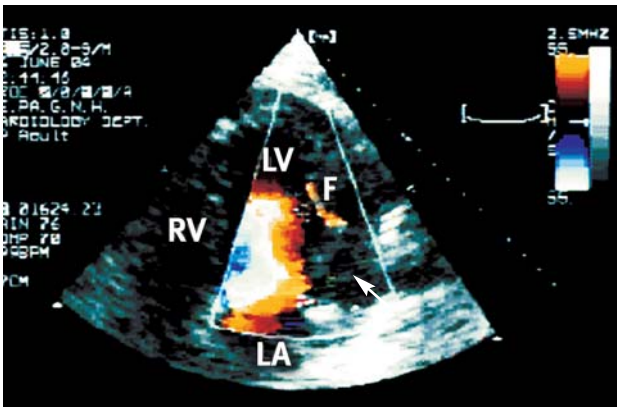



**Figure 1.** Four-chamber view from Case 1, showing a mass (arrow) which invades the free lateral wall of the left ventricle while pericardial fluid around the apex is evident. The colour Doppler reveals the presence of blood flow within the mass. LV – left ventricle; RV – right ventricle; F – blood flow.

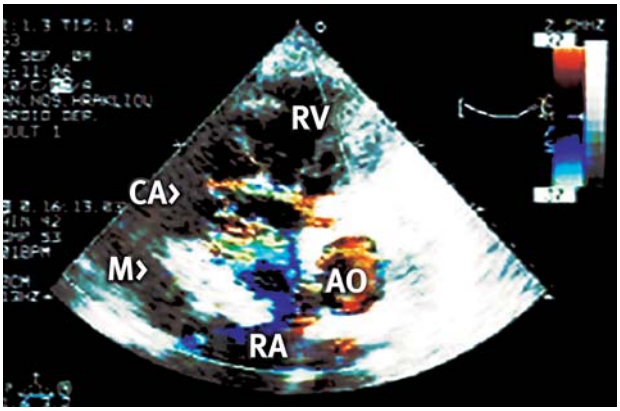
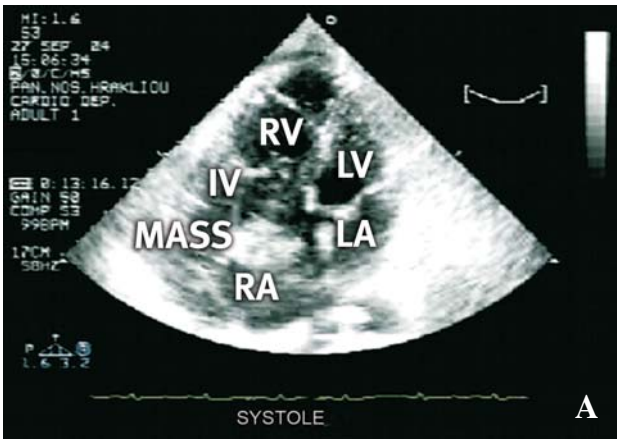



**Figure 3.** A modified left parasternal short axis view from Case 2, clearly showing an enlarged, probably right coronary artery (arising from the aorta at 11 o'clock), which supplies the right atrial mass. AO – aortic root; RV – right ventricle; CA – coronary artery; M – mass; RA – right atrium.



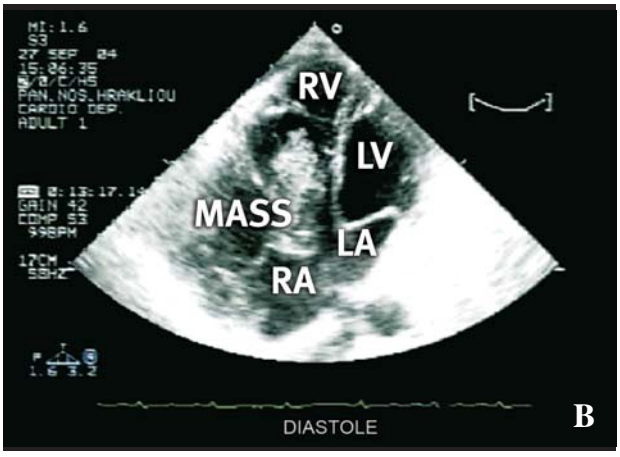




**Figure 2.** Four-chamber view from Case 2, showing a large irregular mobile mass arising from the free right atrial wall (A), that prolapses to the right ventricle in diastole through the tricuspid valve (B). The right chambers are severely dilated and compress the left chamber. LV – left ventricle; RV – right ventricle; LA – left atrium; RA – right atrium; TV – tricuspid valve.

with a massive pericardial effusion 6-8 months prior to the correct diagnosis. Echocardiography suggested the accurate diagnosis, since cardiac tumours with an obviously rich blood supply were recorded. In the literature, to the best of our knowledge, there is only one report that has described the blood supply of an angiosarcoma.[7]

Primary cardiac angiosarcoma should be considered in the differential diagnosis of recurrent haemopericardium, and colour-Doppler flow velocity imaging seems to be a useful tool for the earlier diagnosis of such a tumour.

## References

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