To the Editor,

A 68-year-old man was admitted for dyspnoea at rest and ankle oedema. Chest x-ray showed bilateral pleural effusion. Electrocardiogram and routine laboratory tests were normal. Transthoracic echocardiography demonstrated an echogenic mass containing anechogenic areas involving the right atrium (RA) and right ventricle (RV), and causing a reduction of the right ventricle in-flow. Left ventricular (LV) dimensions and function were normal.

The patient was referred for cardiac magnetic resonance imaging (MRI).

Dynamic imaging using steady-state free-precession cine MRI in the horizontal long-axis plane (Figure 1A) and in the right out-flow plane (Figure 1B) showed a large (8 cm x 13 cm) and inhomogeneous mass located in the RA and RV causing a reduction of RV inflow. Furthermore, there was akinesia of the basal and mid segments of the right anterior (Figure 1A) and right inferior ventricular walls due probably to the mass infiltration (Figure 1B).

Tumour infiltration was observed, particularly in the RA and RV inferior wall (Figure 1B).

A T1-weighted fast spin-echo (Figure 1C) showed a multilobate strongly hyperintense mass. A T1-weighted image with fat saturation (Figure 1D) demonstrated that the hyperintensity was not due to fat and was an expression of the presence of metahaemoglobin due to diffuse subacute-chronic haemorrhage.

Finally, delayed enhanced contrast inversion-recovery depicted an inhomogeneous enhancement involving almost exclusively the tumour periphery. On the other hand, a large part of the mass showed no significant delayed enhancement (Figure 1E). On the basis of the above findings, we made a presumptive diagnosis of a diffusely haemorrhagic malignant heart tumour suggestive of cardiac angiosarcoma.

The patient was scheduled for surgery. However, at surgical exploration through median sternotomy, the mass appeared unsuitable for complete resection. The RA wall appeared completely invaded by the mass that was extending to the atrioventricular junction and to the anterior and posterior RV wall (Figure 2). Further surgical manoeuvres were withheld. Histology confirmed the diagnosis of primary cardiac angiosarcoma with intratumoural haemorrhage. The patient was then referred for medical treatment.

Primary malignant cardiac tumours are rare. Sarcoma is the most common type (about 76%-78%) while angiosarcoma is the second most common type (about 31%). The symptoms are variable. Imaging morphology plays a key role in the preoperative differential diagnosis of cardiac tumours.
To the Editor,

We have read with great interest and admiration the work of Dr Pastora Gallego on vascular disease of the arterial wall in congenital cardiopathies, recently published in the supplement that the journal dedicated to congenital cardiopathies in adults. Of special interest was the paragraph dedicated to the bicuspid aortic valve (BAV), which has great relevance due to both its prevalence (between 0.5% and 2% of the population, as mentioned in the article) and its clinical implications. However, this remains a poorly understood disease with regard to its pathogenic mechanisms and genetic basis.

As mentioned by other authors, BAV patients also represents a heterogeneous population with a wide spectrum of symptoms. As Dr Gallego mentioned with regards to pathogeny, Fedak et al observed a reduction in fibrillin 1 content in the vascular walls of BAV patients. This led to increased production of matrix metalloproteases (MMP) capable of fragmenting and weakening extracellular matrices, which finally leads to degeneration and dilation of the aortic wall.

The analysis of MMP gene expression in BAV patients has also been studied with no definitive results; on the other hand, a study was recently published correlating the plasma concentrations of matrix metaloprotease-2 (MMP-2) with proximal dilation of the aorta in BAV patients.

In connection with the genetic basis of BAV and the increased expression of MMP in the aortic wall of these patients, our group is currently analyzing the differential diagnosis is very broad and must include thrombus, vegetations, intracardiac metastases, infectious and nonbacterial thrombotic or marantic endocarditis as well as other sarcomas (unclassified spindle cell sarcomas, leiomyosarcoma, fibrosarcomas, malignant fibrous histiocytoma, mixoma). Discrimination between these pathologies by echocardiography may at times not give straightforward results. Use of a comprehensive MRI approach is helpful to characterize the different tissue components of angiosarcoma.

As in the reported case, some particular characteristics may help in recognizing an angiosarcoma on MRI:

1. Heterogeneous signal intensity originating from intra-tumoural hemorrhage
2. Heterogeneous enhancement and a cauliflower appearance on T1 weighted SE
3. A strong irregular enhancement appearance on contrast MRI

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