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CASE REPORT
Cardiac imaging

A papillary fibroelastoma with myxoma camouflage: a case report

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Background Benign cardiac tumours are infrequent in clinical practice and, of these, cardiac myxoma is the one with the highest incidence. Given that a left intraventricular presentation is rare, other differential diagnoses such as papillary fibroelastoma should be considered.

Case summary A 73-year-old woman patient with cardiac mass detected in transthoracic echocardiography (TTE) after a transient ischaemic attack. At TTE 2D–3D, a left intraventricular mass anchored at the level of the anterolateral papillary muscle was detected. Subsequently, cardiac magnetic resonance (CMR) was performed for mass characterization. This revealed behaviour in T1 (isointense with respect to myocardium), T2 (hyperintense), very prolonged T1-mapping (1848 msg), and T2-mapping (161 msg) values, without gadolinium uptake in the first-pass perfusion sequence, but with intense uptake in late enhancement sequences. Previous findings were compatible with a diagnosis of papillary fibroelastoma. The mass was resected intraoperatively and, although its macroscopic appearance pointed to a diagnosis of cardiac myxoma, it was finally confirmed to be a papillary fibroelastoma by pathological anatomy.

Discussion In cases where the size of the mass and its mobility allow tissue characterization by CMR, a diagnosis of papillary fibroelastoma and its differentiation with cardiac myxoma are feasible by this cardiac imaging technique.

Keywords Cardiac mass • Papillary fibroelastoma • Cardiac magnetic resonance • Case report • Primary cardiac tumours • Echocardiography

ESC Curriculum 2.3 Cardiac magnetic resonance • 2.1 Imaging modalities • 6.8 Cardiac tumours

Learning points

- In cases where the size of the mass and its mobility allow its tissue characterization, a diagnosis of papillary fibroelastoma is feasible by cardiac magnetic resonance.
- When the mass is located at the left intraventricular level anchored to papillary muscle, papillary fibroelastoma should be included in the differential diagnosis.
- Cardiac magnetic resonance mapping sequences, as well as the behaviour of the mass after gadolinium administration, are key for an accurate diagnosis.
- The definitive diagnosis will always be made by pathological anatomy.

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Introduction

Benign cardiac tumours are infrequent in clinical practice (various postmortem studies report rates between 0.001 and 0.28%) and, of these, cardiac myxoma is the one with the highest incidence.[1,2] However, at the left intraventricular level, its presentation is not common, as it is generally found in the left atrium attached by a pedicle surrounding the fossa ovalis in the interatrial septum.

On the other hand, papillary fibroelastoma is the benign tumour that most frequently affects cardiac valves.[1] Its incidence is slightly higher in males (55%), most prevalent in the 8th decade of life, and the predominant tumour location is the valvular surface, with the aortic valve being the most affected valve, followed by the mitral valve.[3] The left ventricle is the predominant nonvalvular site involved.[3] Clinically, it can present with TIA, stroke, peripheral embolism, myocardial infarction, among others, and tumour mobility and size are independent predictors of embolization.[1,3] They tend to be small masses (<1.5 cm), composed of collagen, highly mobile, narrow, elongated/branching tumours and, like myxomas, attached to the endocardial surface by a short pedicle.[1] Symptomatic patients should be treated surgically with an excellent prognosis.[3] Patients who are not candidates for surgery could be offered long-term oral anticoagulation/antiplatelet therapy, but efficacy is doubtful and no randomized controlled data are available. Asymptomatic patients could be treated surgically if the tumour is mobile, in view of the risk of embolization.[3]

We present the case of a patient with a left ventricle intracavitary tumour with macroscopic aspect of myxoma, finally diagnosed as a papillary fibroelastoma by CMR and anatomical pathology (AP).

Timeline

Time point	Event
Day 1	Referred from another hospital for transient ischaemic attack (TIA) and mass finding in transthoracic echocardiography (TTE) 2D
Day 1	Normal neurological clinical examination/laboratory tests at the time of our first contact
Day 1	New TTE 2D and 3D were performed for our group, detecting a left intraventricular mass anchored at the level of the anterolateral papillary muscle
Day 1	Cardiac magnetic resonance (CMR) was conducted, and mass characterization was compatible with papillary fibroelastoma
Day 2	Surgical resection by cardiovascular surgery department
Day 3	Pathological anatomy compatible with papillary fibroelastoma
Day 8	Hospital discharge. Asymptomatic, without neurological sequelae
Day 30	Outpatient control with transthoracic echocardiography without evidence of cardiac masses
Month 8	Clinical control, asymptomatic, without recurrence of masses

Case presentation

A 73-year-old woman patient was transferred to our centre to study a cardiac mass detected in TTE after a TIA (transient binocular diplopia). The patient had no previous history of any similar neurological/embolic events and dyslipidaemia and arterial hypertension were the only risk factors. At the time of our first contact, the patient had a normal cardiological/neurological examination and no relevant alteration in laboratory tests. On the day of arrival, our group performed a new TTE2D (*Figure 1A*, Supplementary material online, *Video S1*) and 3D (*Figure 1B*, Supplementary material online, *Video S2*), which showed a left intraventricular mass (long arrow) of 10 × 9 mm, pedunculated (short arrow *Figure 1B*), irregular morphology, with wide anarchic and independent movement, anchored at the level of the anterolateral papillary muscle. An adequate contractility of all the myocardial segments, as well as the absence of visualization of the mass after infusion of Sonovue® (see Supplementary material online, *Video S3*), ruled out the possibility of thrombus. With the differential diagnoses of papillary fibroelastoma vs. myxoma of atypical location, CMR was performed the same day for mass characterization.

In CMR (*Figure 2A–F*), the left intraventricular mass described in TTE was identified with the following behaviour in specific sequences: hyperintense (black blood-T2) (*Figure 2B*) and isointense (black blood-T1) (*Figure 2C*) with respect to myocardium, with very prolonged intramass values in myocardial mapping sequences, native myocardial T1 of 1848 msg (*Figure 2D*) and T2 mapping of 161 msg. After gadolinium infusion, no gadolinium uptake was observed in the first-pass sequence (*Figure 2E*); on the contrary, its intense uptake was evidenced in late enhancement sequences (*Figure 2F*). With the diagnosis by CMR of a benign cardiac mass of primary origin of papillary fibroelastoma type, the patient underwent surgery the following day by our cardiovascular surgery department, resecting the mass in question, which macroscopically resembled a cardiac myxoma (*Figure 3A*). Finally, in the pathological anatomy (*Figure 3B*) performed after the immunohistochemical study, the surface lining cells showed immunoreactivity with CD34, CD31, and factor VIII, but were negative for calretinin, excluding the possibility of cardiac myxoma and confirming the presumptive diagnosis by CRM of papillary fibroelastoma.

On the eighth day after admission, the patient was discharged under treatment with acetylsalicylic acid, angiotensin-converting enzyme inhibitor and statins, cardiological/neurologically asymptomatic and without evidence of masses in control echocardiography. The patient has remained asymptomatic and without new evidence of intracardiac masses in clinical control to date.

Discussion

Cardiac magnetic resonance is usually used in the evaluation of cardiac masses because of its advantages for tissue characterization and high-contrast resolution, being noninvasive and not requiring ionizing radiation. In the present case, although the appearance of the mass could be confused with a possible cardiac myxoma of atypical location, the MRI and a multidisciplinary approach were key to reaching an accurate diagnosis of papillary fibroelastoma. On the one hand, although their appearance in black blood T1–T2 sequences (isointense–hyperintense, respectively), and in first-pass perfusion after gadolinium infusion (hypointense, absence of gadolinium uptake) is the same for both cardiac masses,[4] the behaviour of fibroelastomas in late enhancement sequences (intense uptake due to their high collagen content, multifocal vs. heterogeneous patchy uptake for myxomas due to their mixed necrotic content, haemorrhagic foci, and calcification), as well as very prolonged native myocardial T1 and T2 map values in myocardial mapping sequences in fibroelastomas,[5] inclined to its definitive diagnosis, which was finally confirmed by AP, the gold standard in these cases.

Papillary fibroelastomas constitute 11.5% of all primary cardiac tumours.[6] Transoesophageal echocardiography (TEE) is more sensitive at identifying papillary fibroelastomas compared with TTE because of the typical small

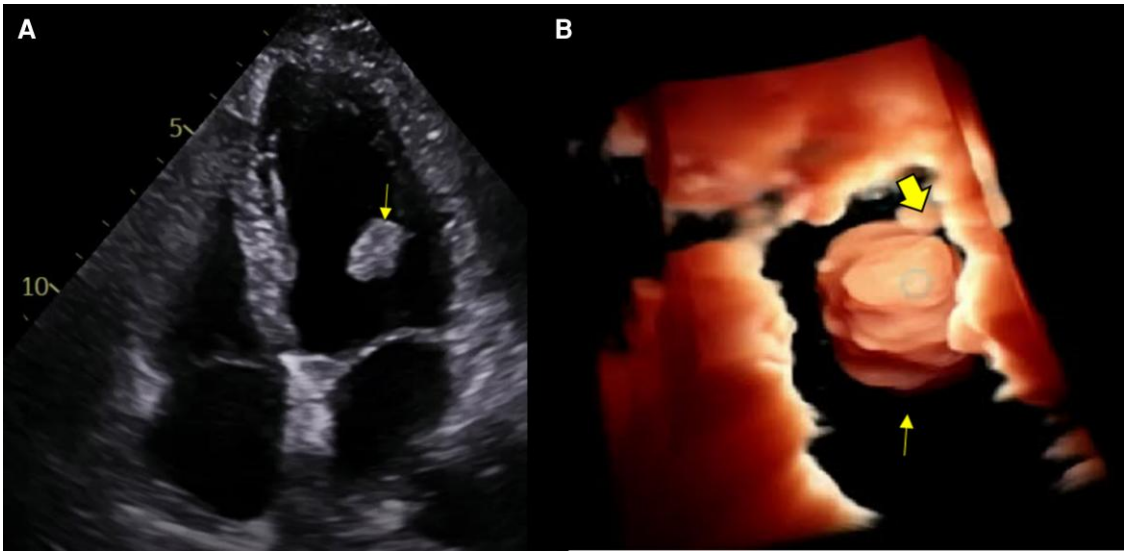

Figure 1 (*A*) Two-dimensional transthoracic echocardiography. Left intraventricular mass with cardiac myxoma-like appearance (*thin arrow*). (*B*) Three-dimensional transthoracic echocardiography. Papillary fibroelastoma (*thin arrow*), small pedicle (*thick arrow*) optimally visualized by three-dimensional technology.

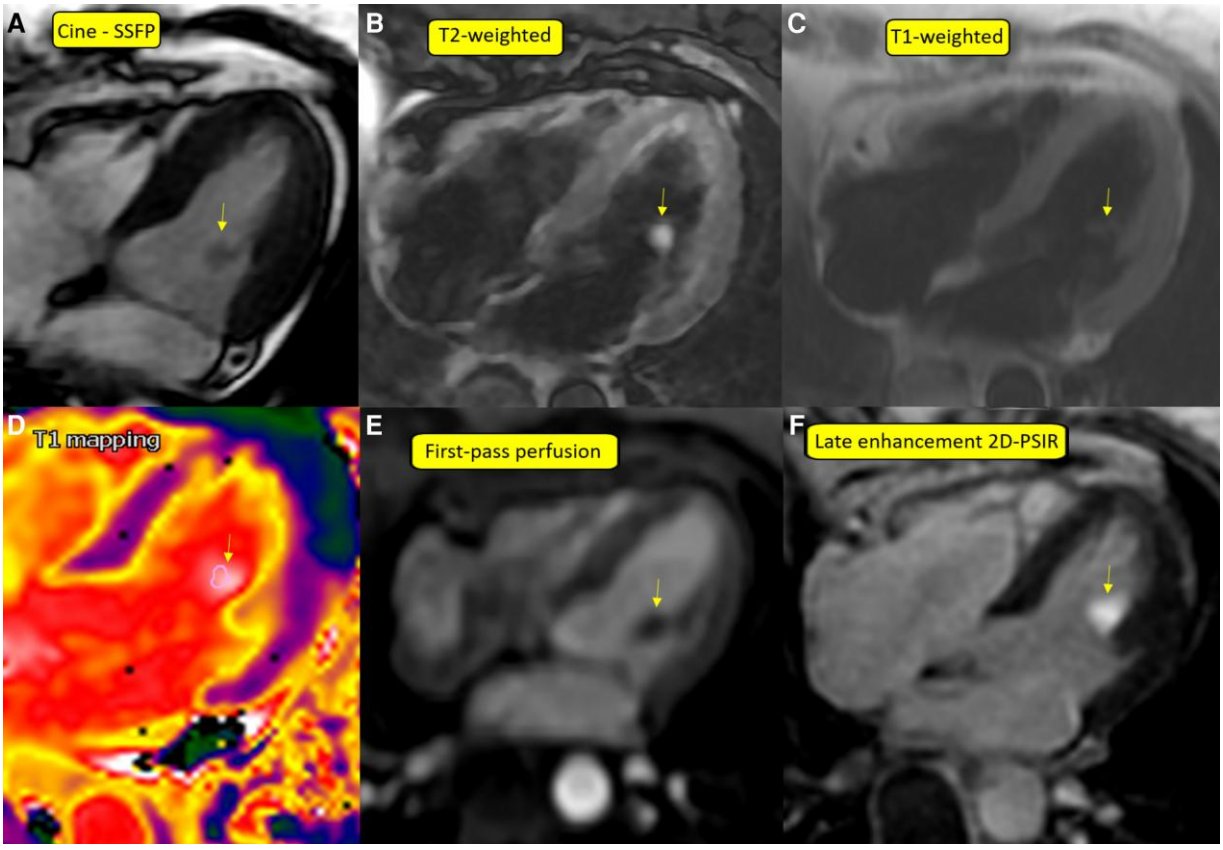

Figure 2 Cardiac magnetic resonance. Tissue characterization compatible with papillary fibroelastoma. (*A*) Cine-SSFP, isointense mass. (*B*) T2-weighted, hyperintense. (*C*) T1-weighted, isointense. (*D*) Very prolonged T1-mapping values. (*E*) Absence of gadolinium uptake in first-pass perfusion. (*F*) Intense gadolinium uptake in late enhancement sequence.

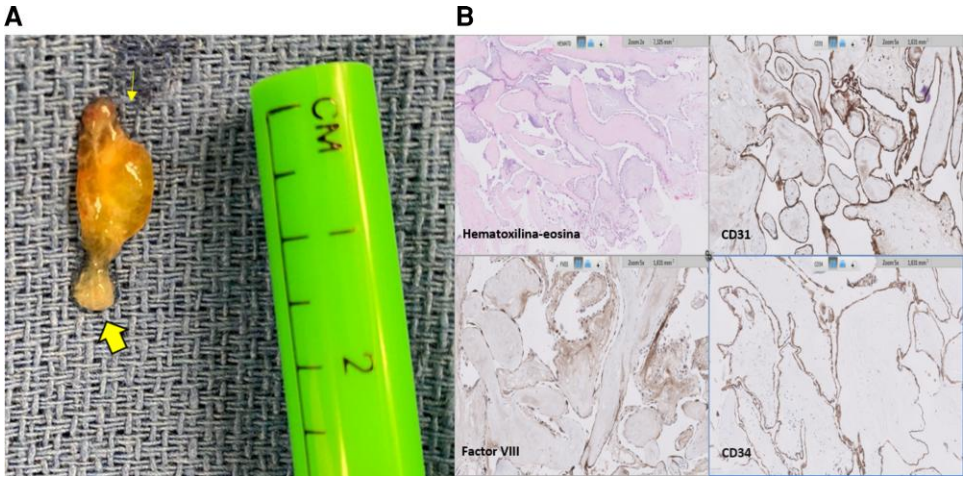

Figure 3 (*A*) Surgical specimen, macroscopic appearance of cardiac myxoma. (*B*) Anatomical pathology compatible with papillary fibroelastoma.

size of these tumours, which reinforces the role of TEE in the evaluation of an embolic event.[6] However, in cases where TTE is the initial study used to detect the mass, TEE is not usually necessary when techniques such as CMR imaging are available. Surgical excision is recommended for larger (≥1 cm) left-sided papillary fibroelastomas in candidate patients at low surgical risk,[3,6] with excellent prognosis/survival, and low recurrence/morbidity.

Although it is difficult to demonstrate that the neurological event presented was caused by the intracardiac mass found, its characteristics: pedunculated form, high mobility, and size (≥1 cm), represent characteristics of high risk of embolism and, therefore, according to the previous literature,[2,3,6] are an indication for surgical resection which, indeed, was performed in our case.

Conclusion

Cardiac magnetic resonance is an excellent tool for the evaluation of cardiac masses, although the final diagnosis will always be made by AP. In cases where the size of the mass and its mobility allow its tissue characterization by MRI, a diagnosis of papillary fibroelastoma and differentiation with cardiac myxoma is feasible in this type of patient.

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Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including the images and associated text have been obtained from the patient in line with COPE guidance.

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