Unusual localization of a malignant fibrous histiocytoma on the mitral valve

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The present report describes the case of a 55-year-old woman who suffered from cardio-embolic stroke originating from malignant fibrous histiocytoma (MFH) localized on the mitral valve. The patient underwent transthoracic two-/three-dimensional and transoesophageal echocardiography which demonstrated the mass protruding in the outflow tract of the left ventricle. Differential diagnosis had to be made with other masses in the left ventricle, such as thrombi, vegetations, and cardiac tumours. Surgery was performed to remove the tumour and the surgery findings confirmed echocardiographic images. Primary cardiac tumours are a rare entity, and their incidence is 0.0017–0.019%. The majority of them are benign, but in a quarter of cases they are malignant. This case is an example of an MFH which caused embolism to the central nervous system.

Keywords Echocardiography • 3D-echo • Cardiac tumours • Cardiac sarcomas • Malignant fibrous histiocytoma

Case report

A 55-year-old woman was admitted to hospital because of ictus cerebri at favourable evolution. She was under treatment for arterial hypertension, and 1 year before the present episode, she was operated for breast cancer. Cerebral computerized tomography (CT) at admittance showed a vanishing low-density area in the right frontal white matter of ischaemic meaning. The patient was in sinus rhythm, and there was no history of arrhythmia.

Nothing was relevant at clinical examination: the woman was in sinus rhythm at 80 bpm and no cardiac murmurs were present; her blood pressure was 120/60 mmHg as expected because of her therapy with angiotensin-converting enzyme inhibitors; peripheral pulses were all present and valid; she was not dyspnoic and no pathologic respiratory sounds were heard at chest evaluation; no oedema or liver enlargement was present. She was classified as being in NYHA I class.

Chest X-ray and CT showed an oval low-density thickening of 0.34 cm in the pulmonary parenchyma, with clean borders in the dorsal segment of the left superior pulmonary lobe, adherent to the posterior thoracic wall and to the great scissor. Echocardiographic evaluation revealed the presence of a floating mass hung to the anterior leaflet of the mitral valve. Suspecting a cardio-embolic event, she was treated with anti-coagulant therapy and transferred to our hospital. We performed a complete echocardiographic study with transthoracic two-dimensional (2D) and 3D echo and transoesophageal echo (TEE). Two-dimensional echo demonstrated a normal systolic function of the left ventricle and normal valve morphology and function. The major finding was the presence of a filamentous pedunculated mass on the anterior leaflet of the mitral valve that was competent (Figure 1). We also performed a TEE, which made possible a better characterization of the mass. It appeared pedunculated, originating from the subvalvular mitral valve apparatus, measuring 5 cm. It was coarsely shaped as a rosary crown, composed of six oval elements with a maximum diameter of 0.7 cm protruding during systole in the left ventricular outflow tract (LVOT) (Figure 2). Echocardiographic characterization of the mass was completed by real-time transthoracic 3D echocardiography which demonstrated that the mass during systole occupied LVOT and entered for 1 cm in the aortic root; in diastole, it retreated inside the ventricle (Figure 3). Correlation with the patient's previous tumoural disease could not be further investigated due to the lack of past medical history documents. According to the medical history of the patient, the most probable diagnostic hypothesis was cardiac tumour, and the therapeutic option chosen was the surgical
removal of the mass. The patient underwent elective surgical treatment to remove the tumour, with free resection margins as confirmed microscopically by cytohistological evidence; on gross examination, it measured $8 \times 2$ cm. No associated procedures such as prosthetic mitral valve replacement or mitral ring annuloplasty were necessary. No post-operative complications occurred. The microscopical analysis showed a pleomorphic malignant tumour composed of spindle-shaped cells, with round–oval nuclei and prominent nucleoli. Giant cells were also present. Focal necrotic areas were seen. Mitotic index was 40–50% with some atypical mitoses. Immunohistochemistry revealed positivity with vimentin, and non-reaction with desmin and smooth muscle actin. These findings oriented the diagnosis towards malignant fibrous histiocytoma (MFH).

**Discussion**

We discuss the case of a cardio-embolic stroke originating from an MFH of the mitral valve.

Tumours of the heart are rare. They were first described by Yater.1 Primitive tumours of the heart have an incidence of 0.0017–0.019%: 70–75% are benign, mostly myxomas, and 25–30% are malignant, in the majority of cases sarcomas. Metastasis to the heart from other tumours is much more frequent than primary cardiac tumours.3 The patient is usually asymptomatic until the advanced stage of the disease, when the tumour interferes with cardiac activity. The main symptoms are connected to heart failure, embolization, and generally compromised situation; arrhythmias are rare.4

Among malignant cardiac tumours, the most frequent one is angiosarcoma. Primary MFH is the second most common primary cardiac sarcoma, with an estimated incidence of $\sim 11.7\%$ of the cases. It usually affects middle-aged people from 14 to 77 years old.5 When it is small, no clinical manifestations are evident.6 As the tumour grows, symptoms can vary according to its location. The most frequent symptoms are shortness of breath, palpitation, or chest discomfort. Histologically, MFH is a variously shaped and multilobulated mass, sessile or pedunculated. It is a tumour of the fibroblasts with giant cells and nuclear and...
Localization of MFH

It is common in the left heart, and embolism is a frequent way of manifestation; also, metastasis is frequent and precocious, particularly to the lungs. Diagnosis needs confirmation with immunohistochemical and ultrastructural examination. The term MFH has now become synonymous of undifferentiated high-grade pleomorphic sarcoma, as the fibrohistiocytic differentiation is not a characteristic of a specific tumour type. The therapy plan depends on the tumour status (primary or secondary), size, histology, location, and metastatic spread. Surgical resection is the gold standard for diagnosis and treatment, and it should be attempted if technically feasible. It often consists in an atriotomy under cardiopulmonary bypass. Radical resection carries a higher risk of peri-operative and post-operative complications. Although the prognosis of MFH is poor and the possibility of local recurrence and metastasis is high, the patients benefit from surgery. Chemotherapy and radiation therapy do not clearly affect outcome, but these treatments can be useful palliative therapies especially when only an incomplete resection is possible. Multiple studies have reported a median survival of 6 months for the right-sided tumours, whereas left heart tumours seem to have a better prognosis. Left-sided tumours, a low mitotic index, and the absence of metastases are associated with a better outcome. Age, gender, the presence of differentiation, and histological type have no impact on prognosis. The tendency of MFH to early metastasis is not as prominent as with angiosarcoma.

The distinction between a benign and a malignant lesion is extremely difficult on a clinical basis, as symptoms are strictly dependent on the location rather than on the microscopic appearance of the tumour. The non-septal origin of the mass strongly supports the suspicion of sarcoma, but multiple attachment sites, infiltration of the mitral valve and/or of the atrial and ventricular walls, are also indicative of malignancy. Differential diagnosis of a cardiac mass should also consider intracavitary thrombi. Contrast echocardiography can reveal the abundant neo-vascularization of tumours (benign vascular tumours and malignant ones in particular) compared with the complete lack of enhancement of thrombi.

Conclusions

This report presents a rare case of cardiac tumour. Echocardiography allowed us to study its morphological appearance and its links to cardiac structures. In the search for embolic sources, transoesophageal and transthoracic echocardiography clearly identified the mass as the origin of the stroke. Three-dimensional echocardiography in particular showed the exclusive involvement of the ventricular side of the mitral valve, and the absence of the mass in the atrium. It also confirmed the commitment of LVOT from the mass. The diagnostic hypothesis oriented towards surgical operation, and pharmacological treatment was excluded. In fact, the probability of the mass to be a thrombus or an endocarditic vegetation was low according to the full echocardiographic characterization with 2D and 3D examination. Surgical findings validated the hypothesis and histological analysis confirmed the tumoural nature of the mass.

Supplementary data

Supplementary data are available at European Journal of Echocardiography online.

References