Heart palpitations as an early presentation of a heart tumor

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A 28-year-old Caucasian woman complaining of heart palpitations was admitted to a cardiac department to undergo a detailed diagnostic workup for an incidental mass found in the heart. A physical examination revealed a systolic murmur (3/6 in the Levine scale) over the pulmonary valve. Laboratory tests and past medical history were unremarkable. A 2-dimensional transthoracic echocardiography revealed a mass (33 mm × 57 mm) in the right ventricle obstructing its outflow tract (FIGURE 1A–1D). Cardiac magnetic resonance imaging demonstrated a tumor in the middle mediastinum located between aorta and pulmonary trunk, tightly connected to the upper wall of the left ventricle and intraventricular septum and bulging into the right

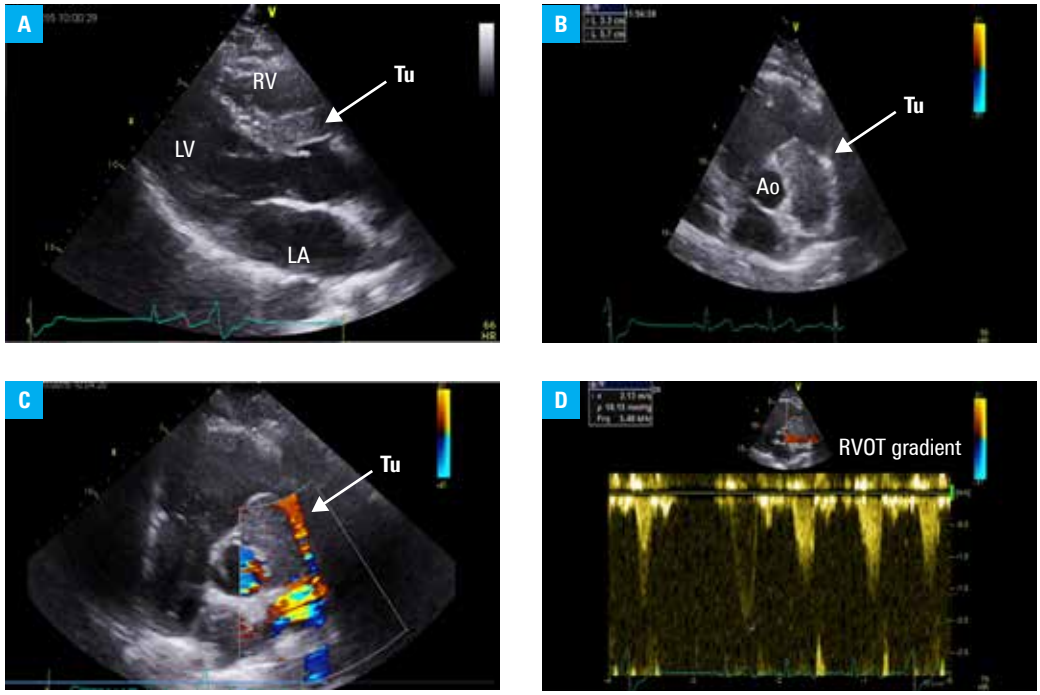

FIGURE 1 **A** – thickening of the intraventricular septum due to tumor infiltration (a transthoracic echocardiographic parasternal long-axis view); **B** – measurement of cardiac mass (33 mm × 57 mm) in the right ventricular outflow tract (RVOT) at the level of the aortic valve (transthoracic echocardiographic parasternal short-axis view); **C** – transthoracic echocardiographic parasternal short-axis view color Doppler mode; **D** – pressure gradient in the RVOT (transthoracic echocardiographic parasternal short-axis view pulse-wave mode); **E** – cardiac magnetic resonance (Steady State Free Precession [SSFP]) image demonstrating a hypointense mass between the aorta and pulmonary trunk; **F** – cardiac magnetic resonance (SSFP) image demonstrating a well-defined hypointense mediastinal mass; **G** – cardiac magnetic resonance (SSFP) image demonstrating a tumor obstructing the RVOT; **H–I** – selective angiography of the left coronary artery demonstrating high vascular supply to the tumor (arrows)

Abbreviations: Ao, aorta; LA, left atrium; LV, left ventricle; RV, right ventricle; Tu, tumor

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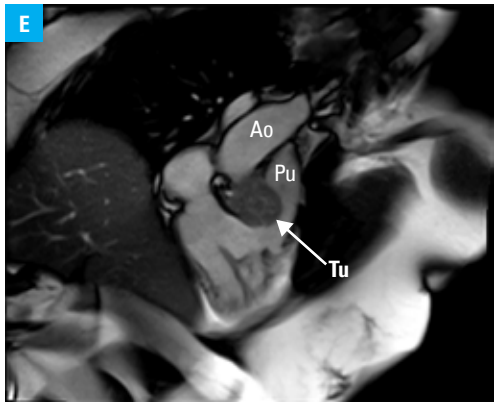

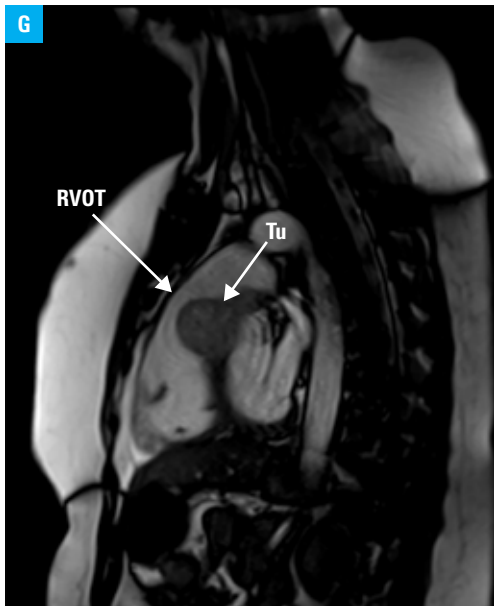

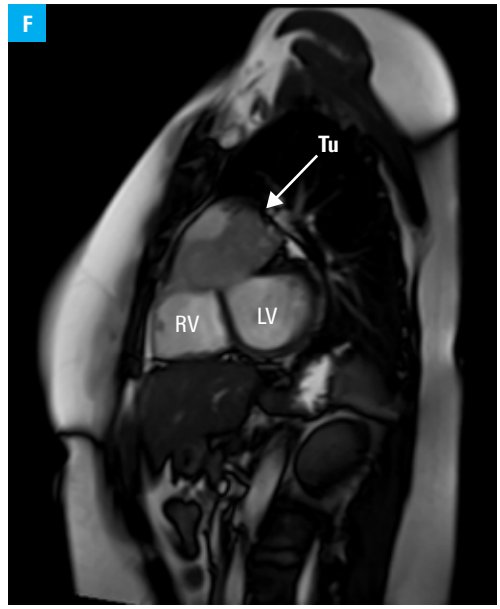

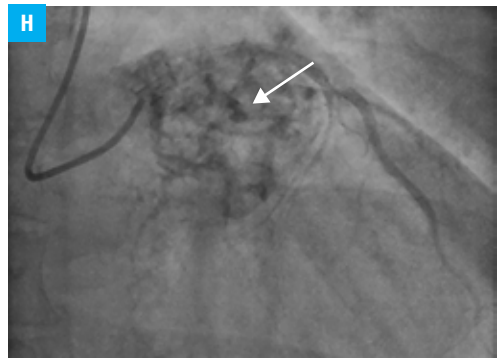

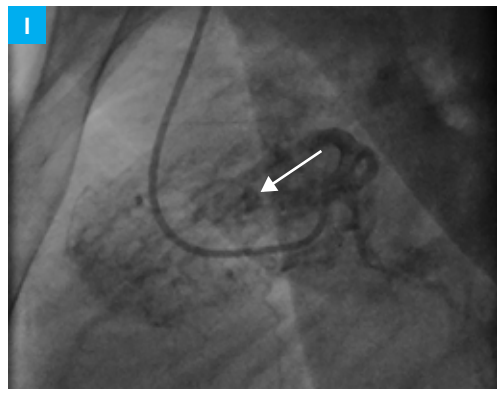

ventricular outflow tract (FIGURE 1E–1G). A selective coronary angiography showed high vascular supply from the left coronary artery to the mass (FIGURE 1H and 1I). A computed tomography of the abdomen, pelvis, and chest excluded any additional tumors or adrenal gland involvement. A positron emission tomography–computed tomography scan showed no focal uptake. Surgical approach was recommended for definite diagnosis and management of the mass. Operative findings following cardiopulmonary bypass confirmed the presence of the tumor in the right ventricular outflow tract, infiltrating the intraventricular septum. The total removal was unfeasible because of the size and location of the tumor, and cardiac surgeons decided on palliative approach (obstruction relief, biopsy). In an immunohistochemical examination, biopsy samples proved positive for chromogranin and synaptophysin as well as sustentacular cells for S-100 protein staining, which confirmed the diagnosis of paraganglioma. The postoperative course was complicated by episodes of sustained ventricular tachycardia, resulting in administration of antiarrhythmic therapy and implantation of an implantable cardioverter defibrillator (ICD).

A 3-year follow-up has shown a stable size of the tumor in echocardiographic examinations. No arrhythmic episodes were retrieved from ICD interrogation. However, the patient is considered a candidate for orthotopic cardiac transplantation.

Paraganglioma is a neuroendocrine tumor originating from extraadrenal tissue usually located in the abdomen. This report describes primary cardiac paranganglioma occurring only in 0.001% to 0.003% of the overall population, with female predominance.[1] Cardiac paragangliomas are typically benign tumors, generally found in the left atrium.[2] Clinical presentation of this neoplasm is often intricate as there is a wide variety of symptoms depending on its functional status (secretion of catecholamines), as well as the size and location in the heart.[3] In

the differential diagnosis, tumors with rich vascular supply should be considered. The diagnosis of cardiac masses relies on multiple imaging techniques, including echocardiography, cardiovascular magnetic resonance imaging, and cardiac computed tomography. This case highlights also the importance of tissue diagnosis in the proper management of the patient with heart tumor.[4] The gold standard treatment for cardiac paragangliomas is surgical approach: complete removal and reconstruction of the involved structures, which however may be impossible due to extensive infiltration of the heart. In such a case, heart transplantation remains the only option for symptomatic cardiac paraganglioma, but long-term prognosis of this approach is unknown.[5]

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