Subtotal occlusion of the left ventricular outflow tract in a young woman

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Primary cardiac tumors are rare intracavitary structures preferentially affecting the left atrium. Available data show that 75% of cardiac tumors are benign with most commonly observed myxoma and cardiac papillary fibroelastoma which may lead to embolic complications, left ventricular inflow obstruction, or may mimic infective endocarditis.[1,2] Undifferentiated sarcomas of the heart are extremely rare. Preoperative diagnosis of cardiac sarcoma of the heart is challenging as it may be mistaken for benign tumors.

We report a case of a 43-year-old woman without a prior history of chronic diseases who was referred to our department for diagnostic evaluation after the first episode of syncope resulting with facial trauma. She had heart palpitations and fatigue on admission. Physical examination revealed systolic murmur at the right upper sternal border. Subsequent transthoracic echocardiography showed a large mass in the left ventricle (LV) attached to the apical segments of the lateral and inferior wall that divided into 2 parts. The smaller one reached the mitral valve, and the larger caused subtotal occlusion of the left ventricular outflow tract and ended in the ascending aorta resulting with V max of 4.25 m/s and max/mean pressure gradient of 68/38 mm Hg (FIGURE 1A–1C, Supplementary material, *Videos S1* and *S2*). No abnormalities of the aortic valve leaflets were observed. Although contractility of the segments that communicated with the mass was diminished, the global contractility of the LV remained preserved with left ventricular ejection fraction of 50%. Complementary cardiac computed tomography confirmed a large irregular structure (51 × 40 × 100 mm) protruding to the ascending aorta (FIGURE 1D). The patient was immediately transferred to the cardiac surgery department, and subsequently, tumorectomy was performed (FIGURE 1E and 1F). The pathological analysis revealed undifferentiated sarcoma. In both postoperative transthoracic echocardiography and chest computed tomography with contrast, a hyperechogenic residual mass in the apex with the diameter of 55 × 22 mm was observed. Unfortunately, due to the rapid progression, advanced stage of sarcoma, and unsuccessful initial chemotherapy, the patient was discharged to hospice care and died 2 months after the initial diagnosis.

Undifferentiated sarcomas of the heart are extremely rare whereas cardiac metastases are more frequently observed in clinical practice than primary tumors, often remain undiscovered until the autopsy, and rarely play a dominating role in the clinical presentation.[1,3] Thrombus and vegetation should be primarily considered in the differential diagnosis of cardiac masses.[3] Surgical tumorectomy in sarcomas affecting the heart can be performed only in selected cases of life-threatening intracavitary solitary tumors but the complete resection remains challenging, operation is palliative in the majority of cases, and the postoperative mortality still remains high.[1,4,5] According to Guan et al[5] the tumor size is an independent prognostic factor for overall- and cancer-specific survival. Moreover, surgery can improve outcomes in patients with primary cardiac tumor size larger than 4 cm.[5]

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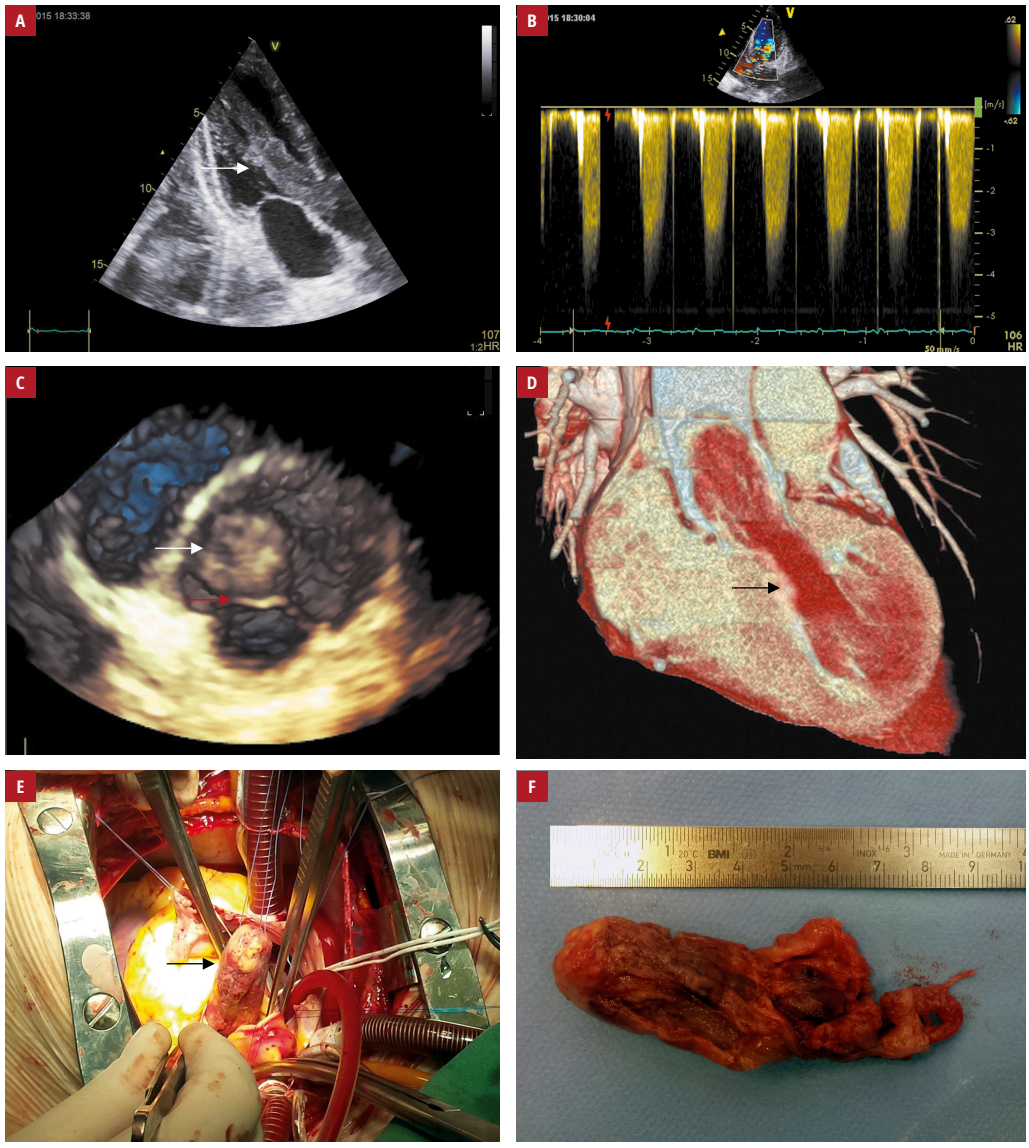

FIGURE 1 **A** – transthoracic echocardiography; the apical 3-chamber view showing a large mass in the left ventricle attached to the apical left ventricular segments (arrow); **B** – systolic continuous-wave Doppler spectrum through the aortic valve; **C** – 3-dimensional echocardiography showing the tumor causing subtotal occlusion of the left ventricular outflow tract (white arrow); red arrow indicates the anterior mitral leaflet; **D** – computed tomography of the heart confirmed a large irregular structure protruding to ascending aorta (arrow); **E**, **F** – tumorectomy (arrow)

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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