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CASE REPORT
Cardiac tumors

Hemangioma of the left ventricle (RCD code: VI-1B.4)

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Abstract

Cardiac tumors are relatively rare, and their differential diagnosis is usually challenging. We present the case of a young man complaining of nonspecific chest pain, who was admitted to the Cardiology Department due to suspicion of a thrombus in the left ventricle (LV). Transthoracic echocardiography showed an additional mobile structure located in the apex of the LV. For further investigation cardiac magnetic resonance imaging was performed, which raised suspicion of a benign tumor. Ultimately the patient underwent uncomplicated cardiac surgery with total excision of the tumor. Histopathology examination revealed capillary hemangioma. JRCD 2017; 3 (4): 133–136

Key words: cardiac tumor, rare disease, echocardiography, magnetic resonance imaging, coronary angiography

Case report

A young man aged 32 was referred to our Center due to suspicion of a thrombus in the left ventricle (LV), based on ambulatory echocardiogram. The patient had no history of chronic diseases and had unremarkable family history. He had been complaining of nonspecific chest pain for a few months. Ambulatory electrocardiogram (ECG) was normal and physical examination did not reveal any abnormalities. Chest X-ray showed no significant pathologies. Transthoracic echocardiography (TTE) showed an additional, mobile structure in the LV apex, which at first, raised a suspicion of a thrombus and was the reason for referring the patient to cardiology department. On presentation the patient was in good general condition, hemodynamically stable, complaining of a stabbing chest pain of a several months duration. Baseline 12-lead ECG revealed sinus rhythm 60/bpm, normal cardiac axis, ST-segment elevation up to 1 mm in the inferior and lateral leads. Serum markers of cardiac necrosis were within normal range (high-sensitivity troponin 0.005 ng/ml [N <0.014 ng/ml] and creatine kinase (CK)-MB 6 U/L [N <24 U/L]). CK was 60 U/L [N <190 U/L], N-terminal pro-B-type natriuretic peptide [NT-proBNP] was 5 pg/ml [N <125.0 pg/ml], while the level of D-dimers was 125 ng/ml [N <500 ng/ml]. Other laboratory tests including blood cell morphology, electrolytes, serum creatinine level, aminotransferases were also within normal ranges.

TTE study showed an additional, spherical, balloting structure, measuring 1 × 1.2 cm, suspended on a fibrous band in the LV apex (Figure 1, 2). The structure had a smooth surface and uniform morphology. However, it became enhanced after intravenous echocardiography contrast injection (Figure 3). Dimensions, morphology and function of the cardiac chambers and valves were normal and small amount of fluid was found in the pericardium (4 mm behind the inferior wall).

Cardiac magnetic resonance (CMR) imaging performed which confirmed additional, mobile, spherical mass, sized 10 × 8 × 11 mm, suspended on a fibrous band, that was attached to the interventricular septum and lateral wall, in the upper one-third of the LV. The structure had isointense signal in steady-state free precession imaging and strong signal on T1-weighted, and T2-weighted images (Figure 4, 5). Moreover, it exhibited strong enhancement in LE and T1 Fat-Sat images, after gadolinium contrast injection (Figure 6). The surrounding walls of LV and the additional string displayed normal CMR signal. There were no visible late gadolinium enhancement (LGE) areas of the myocardium. The images of the structure obtained via CMR were atypical for thrombus, and suggested a tumor.

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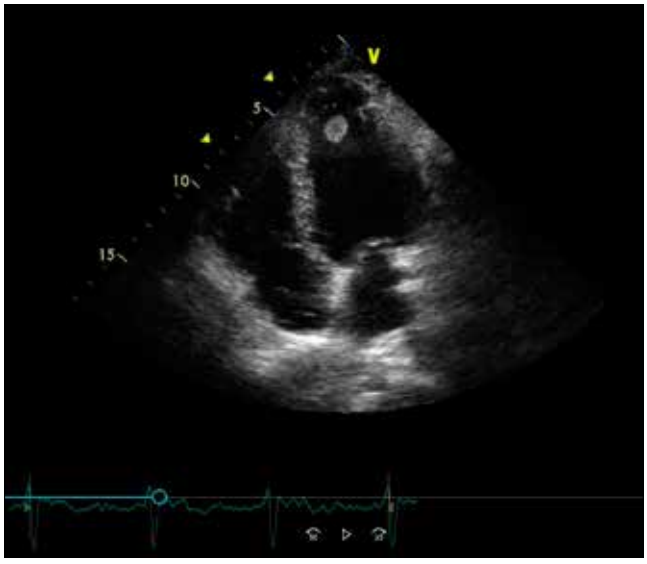


Figure 1. Transthoracic echocardiogram, apical 4-chamber view: an additional spherical structure 1x1,2 cm in the left ventricular apex

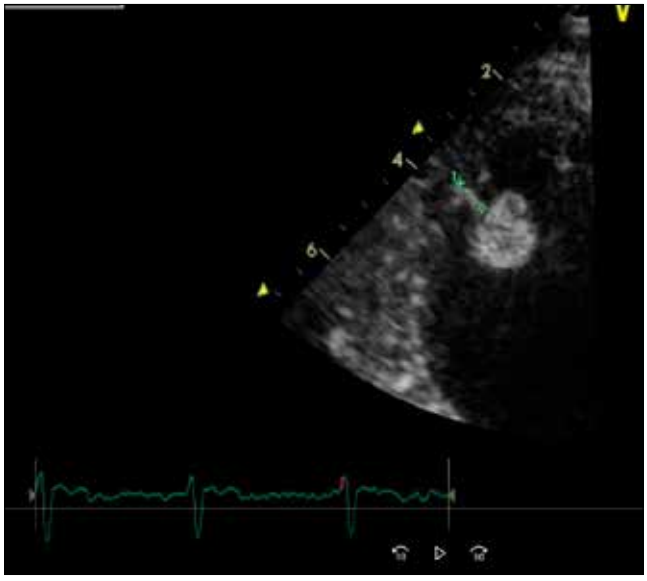


Figure 2. Transthoracic echocardiogram, zoom at the left ventricular apex: an additional structure attached to a fibrous band

The patient was consulted by a multidisciplinary Heart Team and was qualified for surgical excision of the tumor. Total excision of the mass was planned because of the considerable mobility of the tumor, risk of systemic embolization, as well as necessity for histopathological diagnosis. Coronary angiography was performed to exclude obstructive coronary artery disease before cardiac surgery. It showed normal coronary arteries and delayed vascular blush after contrast dye injection into the left coronary artery, indicating that a tumor may be supplied by this vessel. Delayed vascular blush persisted after contrast injection into the right coronary artery (Figure 7).

The operation was performed via median sternotomy. Surgical procedure included antero-lateral LV wall incision of the apical segments, allowing to visualize a pinkish and fragile mass suspended on a transversal chord visible. The tumor was completely excised along with the additional band. Recovery progressed without complications. Post-operative TTE prior to the discharge showed normal LV dimensions and function. Histopathological examination revealed proliferated capillary-size vascular channels, lined by attenuated endothelial cells, with hemorrhages and a thrombus in myxoid and fibrotic stroma. The characteristic morphological signs were typical for capillary hemangioma (Figure 8).

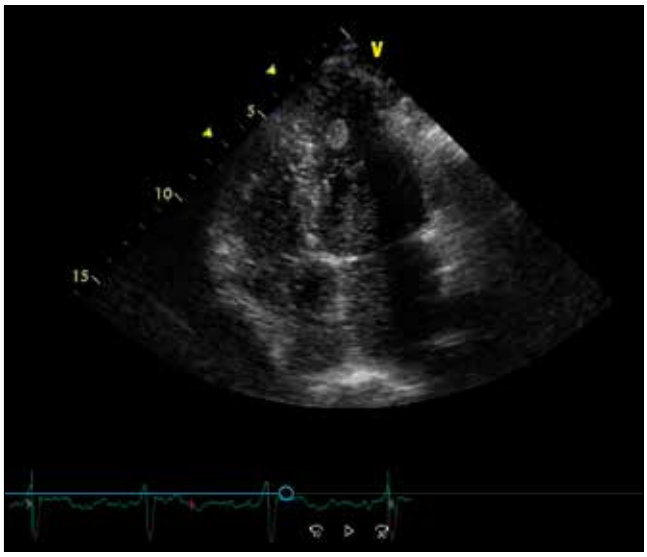


Figure 3. Transthoracic echocardiogram, apical 4-chamber view: tumor is enhanced after intravenous contrast agent injection

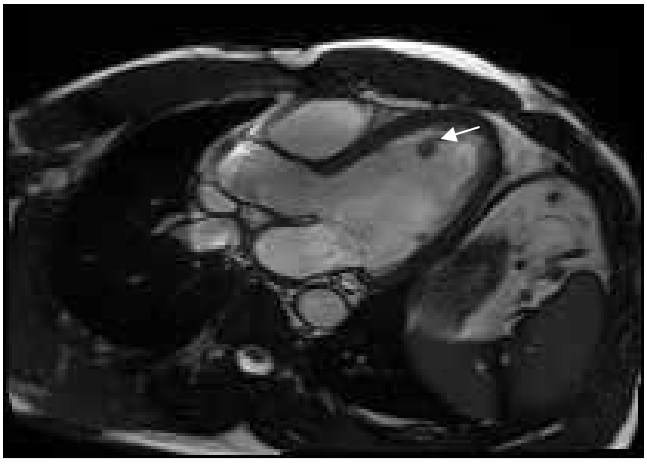


Figure 4. Magnetic resonance imaging, long axis of the left ventricle: isointense signal in steady-state free precession (SSFP); arrow indicates the tumor

Discussion

The prevalence of tumors originating from the heart and great vessels is about 0.02 % in all autopsies and among these, cardiovascular tumors in 70% are benign [1]. Definite classification of

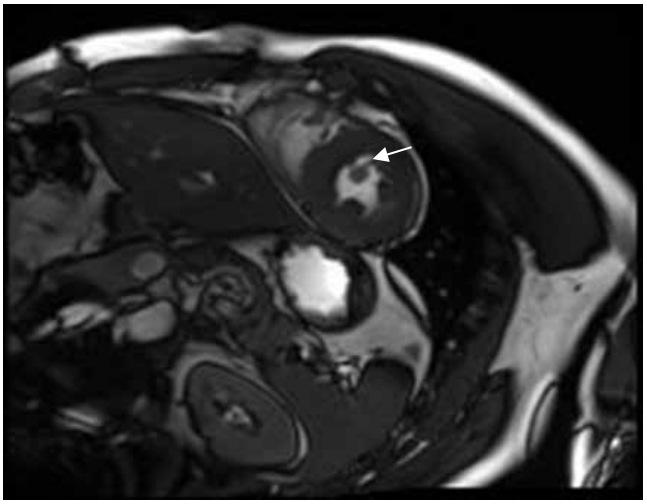
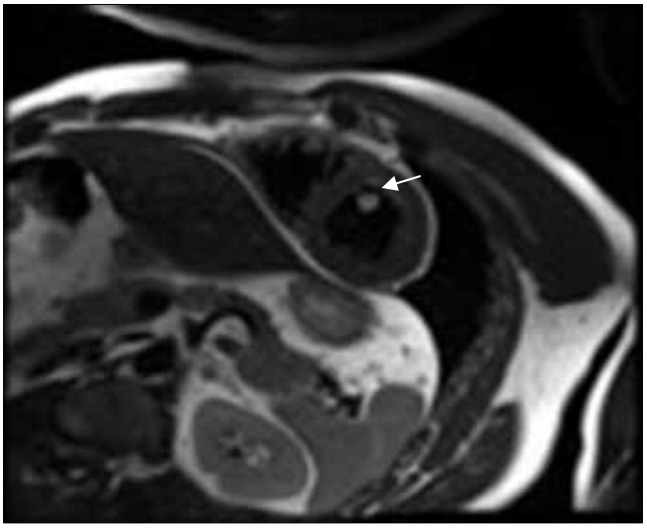


Figure 5. Magnetic resonance imaging, short axis of the left ventricle: the tumor is attached to a false tendon in the upper one third of the left ventricle (arrow)



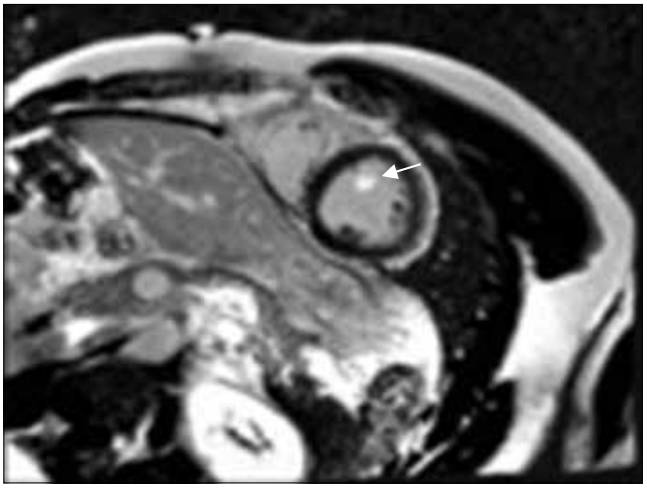


Figure 6. Magnetic resonance imaging, short axis of the left ventricle; the tumor with strong enhancement in LE (A) and T1 Fat-Sat (B) images, after contrast agent administration (arrows)

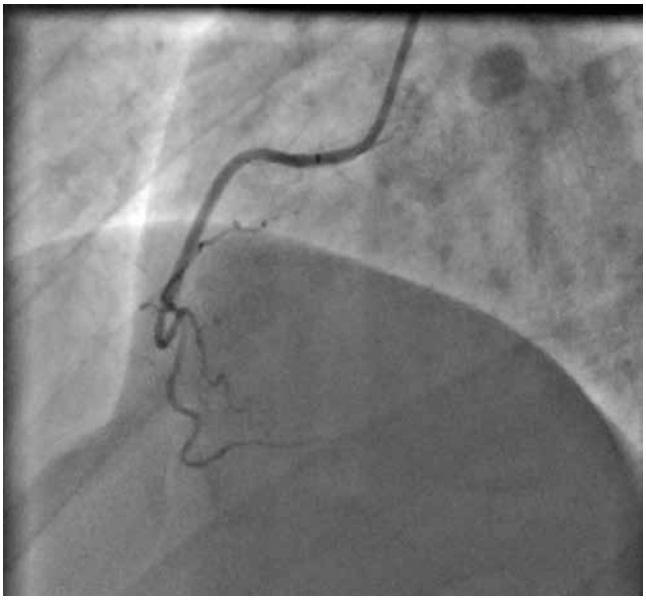
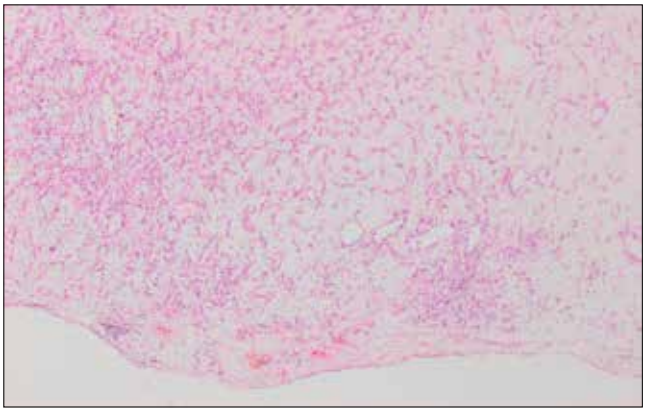


Figure 7. Coronary angiogram: properly contrasted right coronary artery; delayed vascular blush after contrast dye injection into the left coronary artery showing spherical vascular tumor that is fed by that vessel



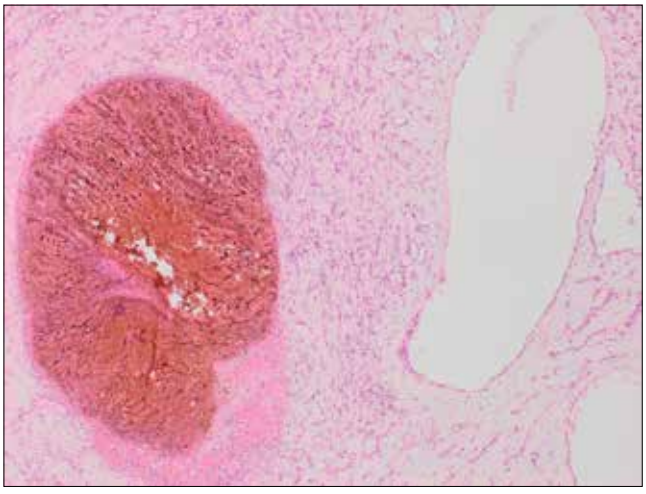


Figure 8. Histopathological analysis of the tumor. Capillary-size vascular channels, lined by attenuated endothelial cells (A) with hemorrhages and a thrombus in myxoid and fibrotic stroma (B)

tumors localized in confined spaces, such as heart and brain carries inherent risk of over-simplification. According to the Clinical classification of cardiovascular tumors and tumor-like lesions, established by Amano Jun et al., hemangioma takes the fourth place in the frequency of benign tumors (2.4–2.6%), after myxoma (34.0–43.1%), papillary fibroelastoma (11.4–17.7%) and lipoma (2.4–3.4%) [2]. On the other hand, according to the recently published classification of rare cardiovascular diseases, hemangioma is believed to be malignant tumor (RCD code: VI-1B.4) [3]. Hemangiomas originate from the endothelial cells and form extensive blood channels. They have an equal age distribution but higher incidence in female than male [1, 2]. Macroscopically they can be polypoid or sessile and histologically they are non-encapsulated tumors [4]. Hemangiomas of the heart may occur in the pericardium, myocardium or endocardium and in fact, they can originate from any cardiac chambers, also from intra-ventricular septum and very rarely from the mitral valve. Natural history of hemangiomas exhibit variable courses; they may enlarge, remain stationary or undergo involution [5, 6]. Hemangiomas are mainly asymptomatic but sometimes have various presentations, which depend on tumor localization, including atypical chest pain, shortness of breath, pericardial effusion, valvular obstruction, heart failure, systemic embolism, rarely as a syndrome of hemangiomas with thrombocytopenia, named Kasabach-Merritt syndrome [7]. Furthermore, there have been reported cases of sudden death due to arrhythmias and conduction disturbances and even cardiac rupture and tamponade [8–10].

Regarding to diagnostic schedule trans-thoracic echocardiography is the first line diagnostic tool which allows to visualize additional structures within the heart chambers. Standard 12–lead ECG usually does not reveal any changes. As it was previously noticed by M. Konieczyńska et al. noninvasive cardiac imaging including CMR or computed tomography (CT) play a crucial role in the diagnosis and subsequent management of patients with cardiac tumors [11]. Therefore, for further investigation CMR or CT are two complementary methods to assess the scope of myocardial and local invasion. Axial T2-weighted CMR shows a high signal mass due to extensive vascularity of the structure [12]. CT usually reveals, after an intravenous contrast injection, enhancement of the tumor, in Hounsfield units equivalent to that of the surrounding blood in the heart, suggesting the highly vascular nature of the tumor. Coronary angiography is helpful in the preoperative evaluation of coronary arteries, additionally it may reveal hemangioma by its characteristic vascular blush. Microscopic examination of the tissues provides unambiguous histological recognition. In most cases complete tumor excision is suggested to prevent serious complications of valvular, left or right ventricular outflow tract obstruction or systemic embolism. The surgery techniques depend on structure's localization in the heart and the surgeon experience. Hemangiomas have likely a good long-term prognosis and the rate of recurrence after surgical resection is rather small [13].

The patient was diagnosed with hemangioma of LV and, after multidisciplinary team evaluation, he underwent uneventful excision of the tumor. There were some clinical doubts earlier, if he should be operated, considering the fact of relatively large injury that the median sternotomy, antero – lateral left ventricular wall section and external cardiopulmonary circulation may do to the organism. On the other hand, the potential risk of systemic embolization was believed to exceed the operative risk and helped with the final decision. During 6 months of follow-up, the patient was in good clinical condition and there were no additional abnormalities on his echocardiogram.

Although there are no specific guidelines for screening of first-degree relatives, members of the patient's family underwent echocardiographic studies that excluded presence of any cardiac tumors.

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