Journal of Rare Cardiovascular Diseases 2016; 2 (8): 263–265
www.jrcd.eu

CASE REPORT
Cardiac tumors

Blood cyst attached to the anterior leaflet of the mitral valve (RCD code: VI-1A.0)

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Abstract

We report a case of a 15 year old boy with a cyst attached to the anterior leaflet of mitral valve. At the age of 6 he was submitted for a cardiac evaluation due to a heart murmur. On physical exam a soft systolic murmur with loudness of 2–3/6 in Levine scale was found without symptoms of congestion. Electrocardiogram and chest X ray were within normal range. Cardiac echo study (ECHO) revealed a single balloon-like anomaly with small compartments inside. The tumor was localized on the ventricular surface of the anterior leaflet of mitral valve. Initially inflow and outflow tracts in colour and pulse wave Doppler were normal without any disturbances. During follow up the patient was asymptomatic without episodes of syncope, congestive heart failure or thromboembolic episodes. He was not treated pharmacologically. At the age of 15, in a subsequent ECHO the size of the cyst was up to 20 mm. The inflow was normal without signs of stenosis, while the left ventricle outflow tract revealed mild dynamic stenosis at the end of systole with maximum pressure gradient of 15 mmHg. The child was qualified for surgery, however the parents did not agree for operation due to the high risk of the procedure and high likelihood of a need for mitral prosthesis implantation requiring lifelong anticoagulation. JRCD 2016; 2 (8): 263–265

Key words: rare disease, echocardiography, cardiac tumor, hamartoma, left ventricle outflow tract obstruction

Case presentation

We present a case of a 15-year-old boy with a cyst attached to the anterior leaflet of the mitral valve. At the age of 6 he was submitted for cardiac evaluation due to a heart murmur. On physical examination a soft, systolic murmur with loudness of 2–3/6 in Levine scale was found, best audible in the third, left intercostal space with normal heart sounds. The loudness of the murmur did not change while squatting or on Valsalva maneuver. Electrocardiogram (ECG) showed regular sinus rhythm, normal axis, no hypertrophy of the ventricles. In chest X-ray the cardio – thoracic ratio was 0.5. Laboratory tests were within normal limits. Transthoracic echocardiography (TTE) revealed a single balloon – like cyst located on the ventricular surface of the anterior mitral leaflet with a diameter of 10 mm. Initially, evaluation of inflow and outflow tracts on colour and pulse wave Doppler were within normal limits, without any disturbances. The cyst was divided into 4 small compartments. During 12 years of follow-up the patient was asymptomatic, without episodes of syncope, with good physical capacity. The boy did not present symptoms of congestive heart failure or thromboembolic episodes. He did not require any specific pharmacological treatment. At the age of 15, in the subsequent TTE study the cyst diameter reached 20 mm (Figure 1). The inflow was normal without symptoms of stenosis, while the left ventricle outflow tract (LVOT) was mildly obstructed with pressure gradient of 15 mmHg (Figures 2, 3). The final diagnosis of the blood cyst was established on the basis of the echocardiographic morphology of the tumor, our experience and references. The child was qualified for surgery due to the LVOT obstruction. However, the parents did not agree for the operation due to the high risk of the procedure and the necessity for mitral prosthesis implantation requiring life-long anticoagulation. We started treatment with aspirin as a prophylaxis of thromboembolic episodes.

Review of literature

A tumor is an abnormal mass of tissue with different shape and origin. The most common are inflammatory tumors which disappear as the process retreat. Another type of tumors are neoplasms

Conflict of interest: none declared. Submitted: March 14, 2016. Accepted: August 22, 2016

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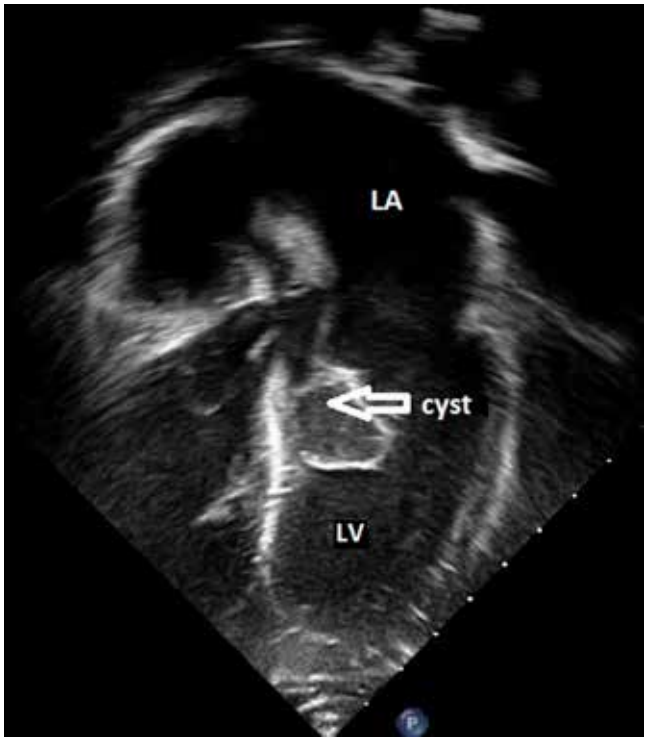

Figure 1. Transthoracic echocardiogram – apical 5 chamber view. "Balloon"-like cyst (white arrow) attached to the anterior mitral leaflet

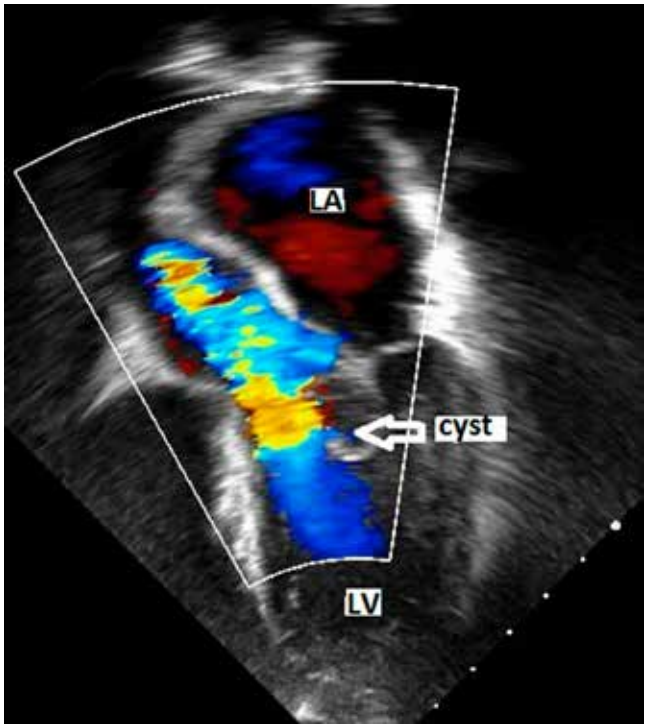

Figure 2. Transthoracic echocardiogram – apical 5 chamber view. Left ventricle outflow tract obstruction due to the blood cyst (white arrow) attached to the anterior mitral leaflet

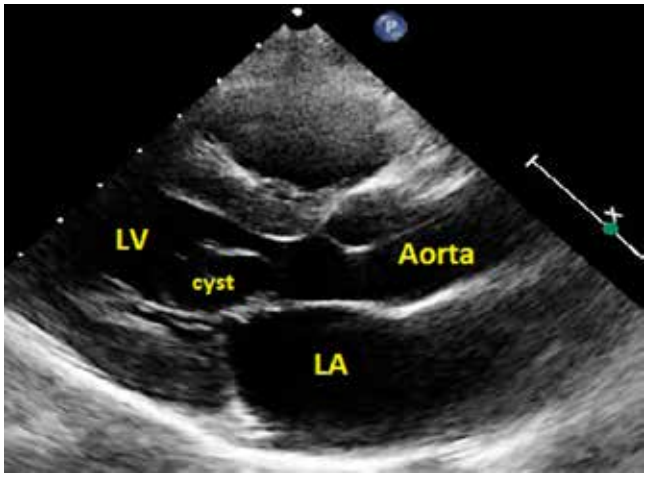

Figure 3. Transthoracic echocardiogram – parasternal long axis view. Blood cyst obstructing left ventricle outflow tract

with benign or malignant character. Benign tumors do not produce metastases, however they can be potentially malignant due to location in the heart leading to flow obstructions or intractable arrhythmias and eventually to cardiac failure [1].

Cardiac neoplasms in children are rare, mostly primary with a prevalence of approximately 1 in 10 000 autopsies. Benign tumors are found 3 times more frequently than malignant. The most common type of benign tumor is rhabdomyoma, which can be multiple with a possibility for a spontaneous resolution. Rhambomyomas are hamartomas associated very often (in about 50–80% of patients) with tuberous sclerosis. Other benign neoplasms include myxomas, fibromas, teratomas, angiomas and lipomas. Among malignant tumors sarcomas are the most common (angiosarcoma, fibrosarcoma, rhabdomyosarcoma, liposarcoma, myxosarcoma) followed by teratoma malignum giving widespread metastases [1].

Cardiac tumors should be distinguished from cysts, which can be found on atrioventricular valves mostly in neonatal period. They show the predisposition to spontaneous disappearance. They can be diagnosed incidentally in older patients on TTE, which is usually performed due to a heart murmur, episodes of syncope or complaints of easy fatigue [1]. The cysts resemble balloon – like tumors with divisions or small cavities inside. The cysts are usually single, small, fibrotic structures connected to atrioventricular valves (mostly mitral, rarely tricuspid) [2]. In rare cases they may present as enormous huge masses obstructing inflow and / or outflow with symptoms of congestion [3]. Etiology of cysts is complex including inherited or acquired types. During embryological development of atrioventricular valves lacunas within endothelium may develop. In case of disturbed flow they may grow and turn into larger cysts. Acquired cysts are iatrogenic and may develop after cardiac catheterization, cardiosurgical operations or inflammatory process. Bleedings from small vessels on the surface of atrioventricular valves may lead to hematomas formation which often resolve spontaneously [2]. Clinical course of a cyst is determined by its size and location. Patients stay asymptomatic for a long time and the initial symptoms may not be specific and include easy fatigue, dizziness, arrhythmias, syncope, stroke or even sudden death. Right-sided cysts may lead to obstruction of tricuspid valve with cardiac failure,

congestion in systemic veins with jugular vein dilatation, hepatomegaly, peripheral oedema, thrombus formation and pulmonary embolism. On the other hand, obstruction of the mitral valve or left ventricle outlet tract may lead to congestion in the lungs with dyspnoea on exertion, easy fatigue, syncope or thromboembolic cerebrovascular episodes [4]. Cysts may be found incidentally on TTE studies in patients with heart murmurs. Large cysts may cause progressive degeneration of atrioventricular valves with stenosis and / or regurgitation [5]. Asymptomatic patients are usually not treated pharmacologically or surgically. However, these patients require regular follow-ups with repeated TTE evaluations of the cyst size, presence of obstruction and functioning of atrioventricular valves. Rarely cysts may predispose to arrhythmias requiring initiation of antiarrhythmic agents. Turbulent flow and congestion due to obstruction may lead to thrombus formation and episodes of emboli (especially strokes). Thromboembolic prevention in asymptomatic patients is a controversial issue [6]. However, in case of high risk for thromboembolic complications or in secondary prophylaxis some patients may require treatment with antiplatelet agents e.g. aspirin with a dose of 3-5mg per kg. In patients with obstructive flow, congestion or episodes of emboli we recommend cardiosurgery with the cyst exision and, if necessary, mitral prosthetic valve implantation.

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