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Leiomyosarcoma of the right atrium

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ABSTRACT

The principal method for early diagnostics of cardiac tumors is echocardiography. Transesophageal echocardiography allows to determine more accurately localization of the growth, structure and flexibility, and also the degree of obstruction of A-V-valves. We demonstrate a clinical case of leiomyosarcoma of the right atrium, its clinical manifestations were cardiac insufficiency, cardiac arrhythmia and pericardial effusion. Difficulties in diagnostics were connected with non-specific complaints due to tumor localization and infiltration with tumoral tissue into the cardiac structures. Our clinical case is of interest because of rarity of pathology, difficulty of intravital diagnostics of disease and interpretation of non-specific clinical picture.

Introduction

Primary malignant cardiac tumors are a rare pathology. Such tumors are characterized with metastasis [1]. Among primary malignant cardiac tumors angiosarcoma is the most frequent (30%), after that — rhabdosarcoma (20%). Primary cardiac leiomyosarcoma, according to different authors, composes less than 1% malignant tumors and less than 8% all malignant cardiac tumors [2, 3].

Urgent diagnostics of leiomyosarcoma is enough difficult task for functionalists and clinicians because of extremely unfavorable prognosis for survivability and objective difficulties for tumor verification. The tumor is characterized with rapid malignant growth and recurring after resection. Five-year survivability composes 25.4% [4, 5]. Early recognition allows to carry out radical surgical treatment that can increase the duration of patients' life in combination with chemotherapy and X-ray therapy.

Leiomyosarcoma grows from smooth muscle cells being located in retroperitoneum and ingrowing to the lower hollow vein. In case of cardiac localization the tumor is the most often found in the left atrium and ingrows to pulmonary veins provoking breathlessness, chest pain, pericardial effusion, damages in cardiac rhythm, signs of cardiac insufficiency. It occurs in all age groups, equally as often both in men and in women.

Difficulties in diagnostics are connected with non-specific complaints due to tumor localization and infiltration with tumor tissue of heart structures. Intravital diagnosis is the most often based on clinical data, echocardiography and more rarely, on data of computer tomography (CT) and magnetic resonance tomography (MRT).

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Aim of Research

It is necessary to analyze this clinical case being of interest because of rarity of pathology and hardness in diagnosis of disease.

Clinical Case (Materials and Methods)

Patient X., 64 years old, was admitted to the department of cardio surgery of Novosibirsk Regional Clinical Cardiologic Dispensary (NRCCD) on March 24, 2015 with complaints of dull, pressing pains in the chest, dyspnea on physical exertion.

The onset of disease was noted in November 2014 when it was appeared pressing pains behind the breastbone on physical exertion. With diagnosis "IHD. New onset of angina pectoris" the patient was directed to diagnostic coronary angiography (CAG) during which it was determined hemodynamically significant three-vessel disease of coronary arteries (stenosis of the right coronary artery — 80%, left marginal artery — 75%, diagonal artery — 70%), with regard to it in Railway Clinical Hospital (Novosibirsk) it was performed percutaneous coronary intervention (PCI) with stenting of stenosis of the right coronary artery (RCA) in the proximal part. As the second stage it was planning PCI in 3 months with stenting of stenosis of the anterior descending artery.

At the out-patient treatment stage the patient noted improvement of well-being, exercise tolerance, improving the quality of life. In January 2015 paroxysm of atrial fibrillation appeared for the first time — the patient was hospitalized to the State Clinical Hospital No. 1. Sinus rhythm was restored with drugs.

In February 2015 it was appeared recurrent pressing dull pains in the chest, bispnea on physical exertion. Paroxysm of atrial fibrillation was developed again that was controlled with drugs. However, in spite of continuation of drug therapy the patient noted progressive augmentation of dyspnea, sharp decrease of exercise tolerance. During echocardiography it was diagnosed expressed pericarditis with effusion with signs of cardiac tamponage. It was performed pericardiocentesis and evacuated 800 ml serohemorrhagic exudates. Conservative therapy was started, it included glucocorticoids but it was ineffective. Till March 20, 2015 during echocardiography-control it was noted accumulation of considerable amount of exudates in the pericardium cavity. The patient was consulted by phthisiologists, diagnosis of tuberculosis was not confirmed.

For additional examination and following treatment the patients was transferred to the department of cardiosurgery of NRCCD. On March 24, 2015 it was performed centesis and draining of pericardium. Summarily it was evacuated approximately 2000 ml of hemorrhagic exudates. In view of this it was made a guess about the presence of tumoral process in the pericardium cavity and in the heart. According to data of multi-spiral computer tomography (MSCT) any tumoral process and metastasing into other organs and systems were not revealed. For an update of diagnosis it was decided to perform transesophageal echocardiography (TEEchoCG) during which in the right atrium it was revealed a foreign tissue lesion occupying two thirds of the atrium (Figure1). The borders of the growth were irregular, flexible. The growth was not prolapsed into the cavity of the right ventricle. Cardiac valve was intact.

Repeated coronarography led to the following results: earlier implanted into RCA stent was without signs of restenosis. In the left coronary arterial district there was cascade hemodynamically significant lesion of the anterior descending artery (ADA) at the proximal and middle parts and hemodynamically significant stenosis of obtuse marginal artery more than 75%. Pathologic vascularization of the growth was in the right atrium from right coronary arterial district.

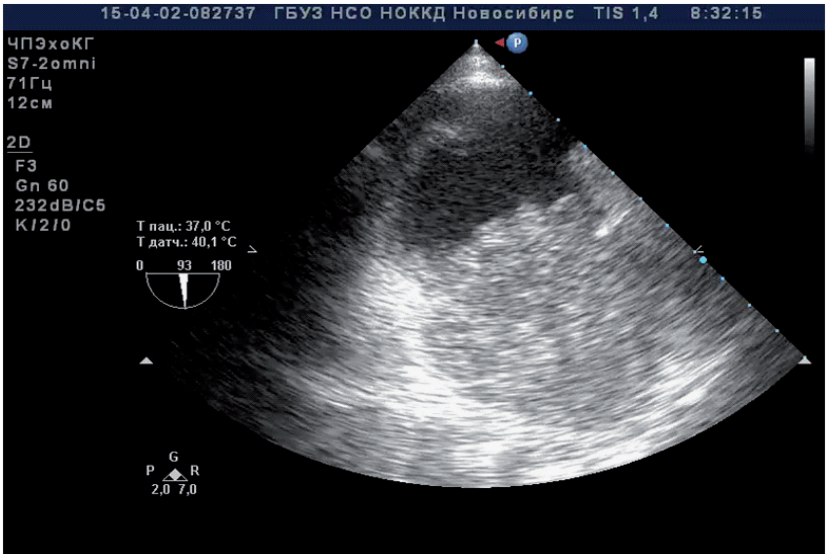

Figure 1. Growth in the right atrium in TEEchoCG

Taking into account combined pathology, great volume of the tumor, presence of chronic heart failure of the III functional class, recurrent damages of cardiac rhythm, it was stated indications for surgical resection of the growth in the right atrium and for aortocoronary bypass (ACB). April 15, 2015 it was carried out the following surgery: oncotomy of the right atrium and resection of a part of interatrial septum (Figure 2). Plastic repair of the right atrium was performed with xenopericardial patch. Heart bypass: left internal thoracic artery (ITA) with diagonal artery; autovein (ACB) with left marginal artery.

It was performed histological study that allowed to verify leiomyosarcoma of low degree of malignance (G1).

Because of the prevalence of the process to excise the tumor extremely was not possible. The patient was followed up by an oncologist. According to MSCT of the chest there were signs of fibrosis of basal segments of the left lung, blocked left-side hydrothorax. According to MRT of the lumbar part of the spine there were signs of secondary process of vertebrae bodies. Special therapy was not performed. The patient was dead in 4 months after surgical intervention.

Results and Discussion

For the period from 2008 to 2015 in Novosibirsk Regional Clinical Cardiologic Dispensary it was operated 12 patients with primary heart tumors: 91% (11 patients) were operated because of mixoma, 9% (1 patient) — because of leiomyosarcoma.

Intracardial leiomyosarcoma was revealed in 3–8% of sarcomata destroying the heart. Intracardial leiomyosarcoma is the most often localized in the left atrium and ingrowing pulmonary veins it is dressed up with symptoms of pulmonary hypertension, chronic heart failure, disorders in cardiac rhythm and conduction [1, 6]. Macroscopically the tumor looks like jelly masses, can be multiple. Due to the invasive growth in myocard the tumor can disturb interventricular septum till the formation of its defect [7, 8]. Right atrial localization of the tumor is a result from a growth of tumoral masses from the low hollow vein. Primary tumor of the right atrium occurs less often, in this case it is possible the fragmentation of tumoral masses and the development of thrombembolia of pulmonary artery.

In our observation there was both exophytic and intramural tumor growth in the right atrium without its prolapse into the cavity of the right ventricle and obstruction of tricuspid valve. In clinical picture the syndrome of cardiac failure is principal one.

Echocardiography is a valid method permitting to reveal a growth in size more than 3–5 mm. However, for soft-tissue structures more diagnostically accurate method is computer tomography and magnetic resonance tomography as echocardiography does not allow to see tumoral myocardial infiltration. By the way it is difficult to differentiate with EchoCG malignant growths from benign ones whereas in using of positron emission tomography (PET) it is possible with 90% response to verify benign and malignant tumors.

Necessary condition for the growth of tumor is intensive angiogenesis. In our patient it was noted pathologic vascularization of the growth in the right atrium from the right coronary atrial district. It was described the case of tumor which first manifestation was the elevation of the segment ST in V2–V4 that allowed to suspect of acute myocardial infarction. Coronaroangiography did not reveal hemodynamically significant stenosis but it revealed bypass of blood flow through the part of angiogenesis with development of “steal syndrome”. Echocardiographically it was revealed a tumor of the right ventricle with development to the pulmonary artery [9]. Therefore, cardioangiography can help in diagnosis. With that hypervascularized tumors are not always succeeded to locate in transthoracic EchoCG that was occurred in our patient. In this case it is necessary to perform transesophageal echocardiography.

Clinically it was revealed untypical localization of tumor — in the right atrium. The patient did not have any specific complaints that caused difficulty of non-invasive diagnostics of malignant cardiac tumor.

A B

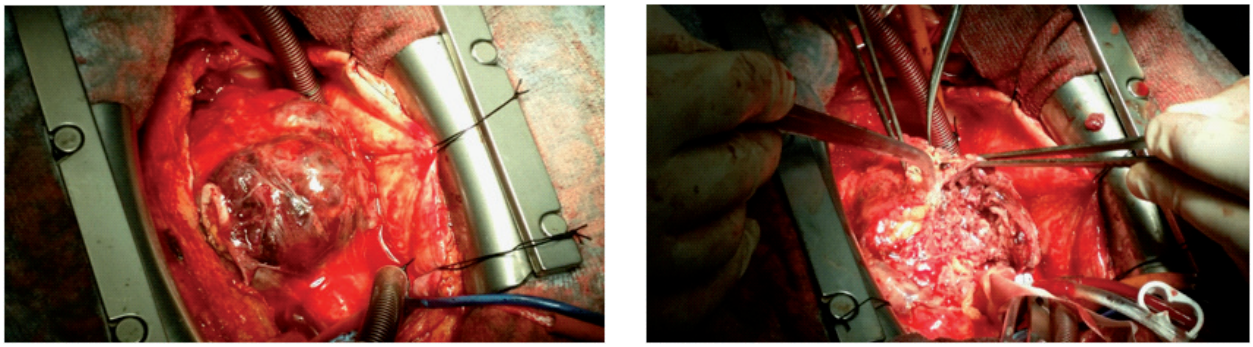

Figure 2. Intraoperative photos:
A — right atrium; B — tumoral burdens in the right atrium cavity

The patient was performed resection of the tumor in limits of health tissues. According to different authors [10, 11] the prognosis even after surgical treatment of leiomyosarcoma rests unfavorable. Leiomyosarcoma is low sensitive to X-ray therapy. The risk of development of such complications as myocarditis and pericarditis considerably exceeds a possible profit from X-ray therapy. Effectiveness of chemotherapy is not proven too.

According to data of F. Mayer et al. [12], more than 50% of patients with sarcomata have metastasis in lungs (35.7%), regional lymph nodes (14.2%) and liver (7.14%). Metastasing of primary cardiac tumors into bones occurs very rarely and has unfavorable prognosis. In our patient during postoperative period it was revealed distant metastasis in vertebrae bodies that considerably made worse our prognosis.

Conclusion

This studied case allows making following conclusions:

1. Primary malignant cardiac tumors are rarely occurred diseases and demonstrate objective difficulties in diagnostics because of the absence of specific complaints and difficulty in early diagnostics.

2. Echocardiography in such patients is a simple and available screening method of study allowing to evaluate: a) size of tumor, b) its location, c) interaction with heart structures, d) presence and obstruction degree of cardiac valves.

3. For final verification of tumor it is necessary to perform additional studies: computer tomography, magnetic resonance tomography with contrast substance, positron emission tomography.

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