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A CLINICAL CASE OF PRIMARY MALIGNANT FIBROUS HISTIOCYTOMA OF THE HEART

Sarcomas are common malignant tumors of the heart. Malignant fibrous histiocytoma is one of the rare forms of heart sarcomas, morphologically diagnosed by cytological and immunohistochemical studies.

The article presents a clinical case of primary malignant fibrous histiocytoma of the heart from the practice of the cardiology department of the Yakutsk Republican Clinical Hospital. The tumor was located in the right ventricle, which is rare and the clinical picture was dominated by symptoms of chronic pulmonary embolism with hypertension. On echocardiography and computed tomography, the volumetric lesion was regarded as a thrombus. Autopsy and histological examination revealed malignant fibrous histiocytoma of the heart. The complexity of the diagnosis was associated with the nonspecific polymorphic clinical picture and the different structure of the neoplasm itself.

Keywords: cardiac tumors, malignant fibrous histiocytoma, pulmonary thromboembolism, chronic thromboembolic pulmonary hypertension, pulmonary trunk, right ventricle, clinical case, malignant neoplasms, diagnosis, clinical case.

Introduction: primary cardiac tumors are very rare, compared with secondary ones, of all autopsies they are detected in approximately 0.001-0.28% [1]. The reason for the rare development of cardiac tumors is believed to be that there are features in myocardial metabolism, such as rapid blood flow inside the heart and limited lymphatic connections in the heart [2].

Benign, among primary cardiac tumors are three times more common than malignant. Cardiac sarcomas are malignant tumors that often develop at the age of 30-50 years, on average at 40 years [1,2]. In men, they occur with a frequency of 65-75%, respectively more than in women [6].

The clinical picture depends on the localization and on the type of tumor; it is manifest by symptoms not only from the heart, but also from other systems and organs. Only in 3-10% of cases they are detected by clinical signs [3,6]. They can usually resemble signs of common heart disease: a variety of chest pains, syncope, arrhythmias, heart murmurs, conduction disturbances, pericardial effusion, or cardiac tamponade. The nature of the symptoms observed in cardiac tumors is most closely related to the localization of the tumor [7,8]. Circulatory failure is the most common manifestation of primary cardiac tumors. It occurs when the tumor is large and is caused by the mobility of the tumor in the heart chamber, causing disturbances in the flow or outflow of blood. If the myocardium is damaged by a tumor, its contractile function may be impaired, which also leads to congestive heart failure [6].

From obstruction by a tumor of the atrioventricular opening in 30% of patients, sudden death occurs, and in the rest, more often the cause of death is progressive heart failure and embolic complications [4,7].

Detection of a volumetric neoplasm in the cavity or layers of the heart and positive biopsy results are the main diagnostic signs of cardiac tumors [9].

Malignant fibrous histiocytoma is characterized by intracavitary growth, accompanied by obstructive and constitutional symptoms. The most common tumor localization is the left atrium, but the tumor can also grow in the right atrium and right ventricle [4,5]. Since most malignant primary cardiac tumors have a crumbling intracavitary surface, blood clots can form on uneven surfaces. Accordingly, tumor embolism of the pulmonary artery, as well as obstruction of the left heart, can lead to pulmonary hypertension [8,9]. In 11-24% of cases, malignant fibrous histiocytoma is detected among sarcomas [10]. Metastasis to other internal organs is rare.

The main radical treatment method is surgical removal of the primary tumor [8].

The prognosis for malignant neoplasms of the heart is unfavorable, patients die within 6 - 12 months after the onset of the first clinical symptoms [7, 10].

Description of the clinical case: we present a clinical case from the practice of the cardiology department of the Yakutsk Republican Clinical Hospital.

Patient Z., 46 years old, a resident of the village, upon admission complained of shortness of breath at the slightest physical exertion, weakness, loss of consciousness.

From the anamnesis it is known that from time to time he was bothered by compressive pains in the region of the heart, which healed on their own. He did not seek medical help. Later, the pain began to appear more often, shortness of breath during physical exertion began to disturb. I went to the local hospital. Auscultation revealed a murmur in the heart, diagnosed with a heart defect. To clarify the diagnosis, he was sent to the Yakutsk Republican Cardiological Dispensary, where, after examination, he was diagnosed: chronic pulmonary embolism. Secondary pulmonary hypertension of 1 degree. CCPF 2 A, FC II. Right ventricular thrombus. An extract was sent to the Federal State Budgetary Institution National Medical Research Center named after Academician E.N. Meshalkin, Ministry of Health of the Russian Federation, Novosibirsk. On the recommendation, he took warfarin under INR control, amlodipine 2.5 mg. In connection with the deterioration of his condition within 6 months, in the form of an increase in shortness of breath, which began to appear at the slightest physical exertion, he was sent again to the Yakutsk Republican Cardiological Dispensary. During physical exertion - climbing on foot to the 8th floor (the elevator did not work), fainted. Delivered by ambulance to the cardiology department of the Yakutsk Republican Clinical Hospital.

Upon admission: the general condition

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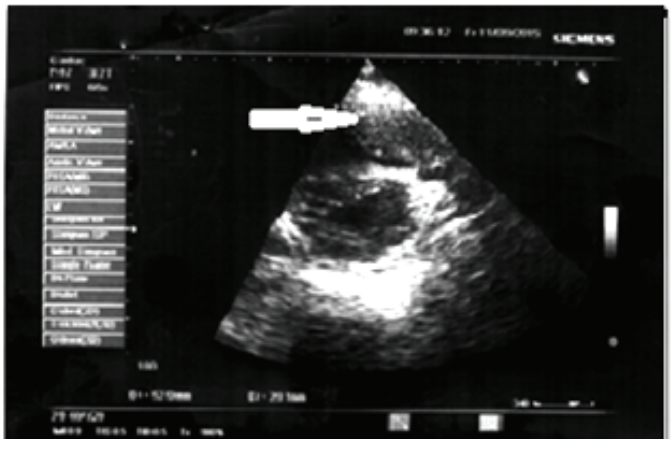

Fig.1. Echocardiography with formation in the right ventricle

is severe. Consciousness is clear. Active position. The skin is clean, pale. Rashes, no hemorrhages. Skin turgor is preserved. Visible mucous membranes are clean and pale. Nasal breathing is free. On auscultation in the lungs, breathing is hard, no wheezing. Respiratory rate - 20 per min. SpO2 - 94%. Heart sounds are weakened, the rhythm is correct. Diastolic murmur over the tricuspid valve, accent of the second tone over the pulmonary artery. BP - 100/80 mm Hg, heart rate - 96 per min. The abdomen is soft and painless on palpation. The liver is not enlarged, painless. The chair is not broken. Free, painless urination. The urine is light. No peripheral edema.

Laboratory tests: a general blood test and a general urine test without pathological changes, in the biochemical blood test, an increase in CRPhs was noted - 14.3 mg / l, creatinine - 146 mmol / l; D-dimer was within the normal range of 186 ng / ml, INR over time from 1.6 to 3.2.

Instrumental studies: chest X-ray - in the lungs without focal and infiltrative changes.

ECG shows sinus tachycardia 109 beats per minute. Incomplete right bundle branch block. Signs of chronic cor pulmonale. Pulmonary embolism.

Echocardiography: aorta 36 mm, not dilated, not changed. The leaflets of the aortic valve are thin. The left atrium is not dilated (28x35mm), the left ventricle is not dilated (EDD - 39mm, ESD 23mm), the right ventricle is expanded (40mm), the right atrium is expanded (58x53 mm). Pulmonary trunk 21 mm. The valves of the pulmonary trunk are compacted with fibrosis, deformed. Pressure gradient 7.3 mm Hg. The leaflets of the tricuspid valve are thin, regurgitation of the II-III degree. On the anterior wall of the right ventricle, there is a fixed hyperechoic formation with a diameter of 52.9x29 mm, color doppler mapping "0" of blood flow. A blood clot from the right ventricle descends to the valve leaflet, partially covering it. Fluid in the pericardium. EF 72% satisfactory (Fig. 1).

Computed tomography of the chest and mediastinum with a bolus injection of contrast medium - CT for signs of a thrombus in the right ventricle, pulmonary trunk, partly in the left pulmonary artery. Thromboembolism of small branches of the pulmonary artery (small wedge-shaped foci with indistinct contours along the S1-2 projection of the left lung and S8 of the right lung) is not excluded. Parietal calcifications on the pulmonary trunk. CT - signs of hydropericardium. Diffuse parenchymal changes in the liver.

In the department, the patient's condition remained consistently grave. Received treatment: warfarin 5 mg under INR control, amlodipine 2.5 mg, inspra 25 mg, digoxin 0.125 mg, sildenophil 75 mg per day. In the department he was consulted by an angiosurgeon, diagnosed with thrombosis of the right ventricle, pulmonary trunk, left pulmonary artery and by a cardiac surgeon, diagnosed with pulmonary embolism. Thrombosis of the pulmonary trunk, left pulmonary artery. Right ventricular thrombus. High-grade pulmonary hypertension. It was recommended to send the patient to federal centers of cardiovascular surgery when stabilizing

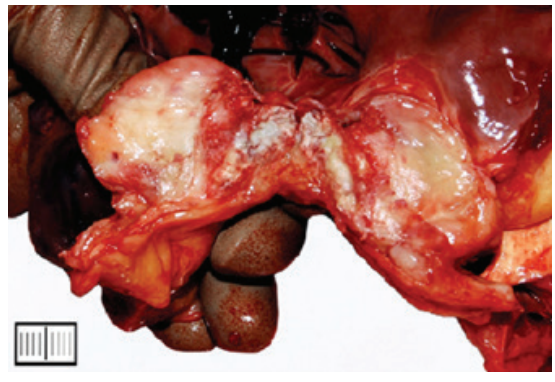

Fig. 2 Malignant fibrous histiocytoma in the right ventricle.

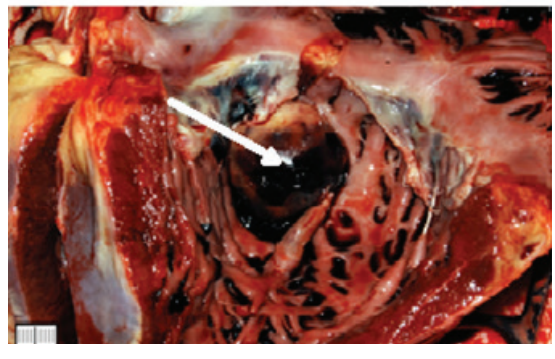

Fig. 3 Thrombus on the tumor

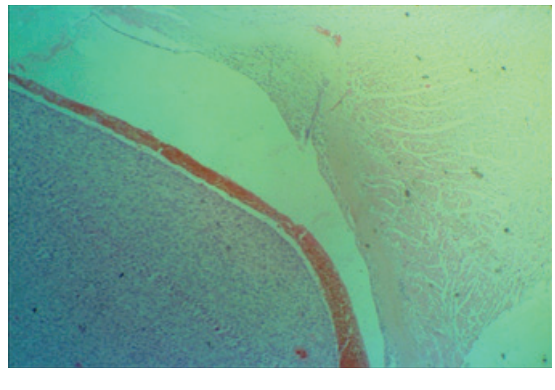

Fig. 4 Malignant fibrous histiocytoma of the right ventricle.

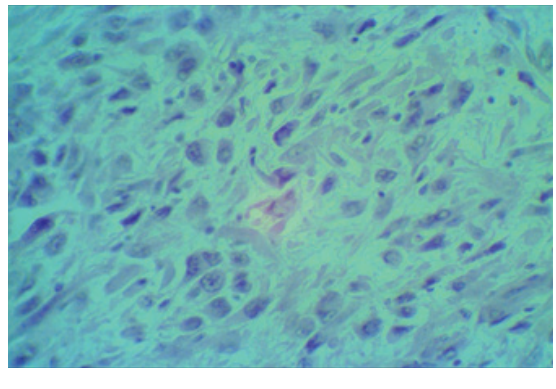

Fig. 5 Germination of malignant fibrous histiocytoma into the myocardium

the patient's condition due to the lack of the possibility of performing surgical treatment in full in the conditions of the cardiac surgery department of the State Autonomous Institution of the Republic of Sakha (Yakutia) "Republican Hospital No. 1 - National Center of Medicine". By the decision of the council, an extract was prepared and sent to the Federal State Budgetary Institution National Medical Research Center of Cardiology of the Ministry of Health of Russia, Moscow. Based on complaints, anamnesis data, laboratory and instrumental studies, a clinical diagnosis was made: recurrent pulmonary embolism. Right ventricular thrombus. Chronic thromboembolic pulmonary hypertension of severe severity. Complication of the underlying disease: chronic cor pulmonale. CHF 2A FC III NYHA. Lung infarctions S1-2 on the left, S8 on the right. Syncope condition. Cardiogenic liver fibrosis. Concomitant diseases: chronic bronchitis, without exacerbation

In the department, his condition worsened, shortness of breath increased and there was an episode of syncope, in connection with which he was transferred to the intensive care unit. Due to the progression of heart and respiratory failure, the patient died.

Autopsy and histological examination revealed malignant fibrous histiocytoma of the anterior wall of the right ventricle (at the outlet of the right ventricle) and proximal pulmonary artery with invasion into the myocardium and posterior left pulmonary artery valve with parietal thrombosis (Fig. 2,3,4,5). The cause of death was pulmonary embolism.

Conclusion: tumors that grow in the heart cavity need differentiation between themselves and other masses in the heart cavity, for example, with blood clots. In this clinical case, echocardiography and computed tomography were performed, where the mass was regarded as a thrombus. Autopsy and histological examination revealed malignant fibrous histiocytoma of the heart.

Obviously, the discrepancy in the diagnosis was due to the difficulty of diagnosing primary cardiac tumors associated with a nonspecific polymorphic clinical picture and a different structure of the neoplasm itself.

The described clinical case is an example of a pathology with a rare localization of a heart tumor. The presence of masses with a clinic for thromboembolism and pericardial effusion should be alarming in terms of oncology with an expanded patient examination plan.

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