

Myxomas

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Abstract

The Article discusses key aspects of the etiology, clinical picture, diagnosis and treatment of cardiac myxomas. Despite their histologically benign nature, these tumors pose a serious threat due to the risk of life-threatening complications, such as systemic embolism, valve obstruction and sudden cardiac death. The clinical picture is polymorphic and often imitates systemic inflammatory or rheumatic diseases, which complicates diagnosis. The main verification method is echocardiography, which allows determining the localization, size and mobility of the formation. The only method of radical treatment is timely surgical removal of the tumor, which is associated with a favorable prognosis. The work emphasizes the importance of differential diagnosis and the need for early surgical intervention to prevent complications. Special attention is paid to the differences in prognosis between sporadic and familial forms of the disease.

Keywords: cardiac myxomas; inflammatory; rheumatic diseases; complicates; diagnosis

Introduction

Early detection of cardiac neoplasms presents significant difficulties due to the specificity of metabolic processes and hemodynamics in the myocardium. Clinical manifestations of this pathology are polymorphic: from asymptomatic course at the initial stages to the presence of various non-specific complaints (shortness of breath, increased fatigue, tachycardia) in the absence of pathognomonic signs. In vivo diagnostics remains difficult, despite the use of modern imaging techniques, including echocardiography and magnetic resonance imaging. One of the most common primary intracardiac tumors is myxoma (according to ICD-10: D15.1), with an estimated incidence of 0.0017% to 0.28%. Histologically, it is benign. The neoplasm can be localized in any chamber of the heart, but the most typical localization is the cavity of the left atrium. Biatrial myxoma is a rare form of the disease characterized by damage to both atria with possible growth into the interatrial septum and/or ventricles. The pathology is diagnosed in all age groups, with a peak incidence between 30 and 60 years, and somewhat less often in male patients. Statistically, myxoma of the left atrium is detected significantly more often than that of the right atrium. Difficulties in visualizing the neoplasm are associated with the polymorphism of its density structure, the presence of intraluminal thrombi in the atria, as well as possible atypical localization on the atrioventricular valve cusps. As a result, this pathology is often an incidental finding during an examination for another reason or during a postmortem examination. Cardiac myxomas are sometimes combined with congenital heart defects. The size of myxomas varies from 1 to 12 cm in diameter, and the weight - from 0.6 to 80 g.

Main Part

The etiology of myxomas remains a subject of scientific debate. Potential trigger factors include genetic predisposition, previous cardiac trauma, and infection with Epstein-Barr, Coxsackie, and human papillomaviruses. Modern pathomorphological theories suggest that the tumor originates from endothelial cells or embryonic remnants with subsequent myxomatous degeneration. Histological analysis often reveals fibrous changes in the endocardial layer, atrioventricular valve structures, and basal sections of the atria. Fibrin deposits and proliferation of elastic and collagen fibers are characteristic. Severe tumor calcification can induce destruction of the valvular apparatus. Despite its histologically benign nature, the clinical course can be complicated by life-threatening conditions, including sudden cardiac death. Clinical manifestation is characterized by polymorphism, determined size, mobility and localization of neoplasia. Cardiac arrhythmia, thrombotic and thromboembolic complications are observed. The classic triad of symptoms includes: constitutional manifestations, embolic events, signs of intracardiac obstruction. Key clinical markers are: auscultatory murmurs that change with a change in body position, with an uncomplicated rheumatic history; therapy-resistant heart failure of rapid progression; the occurrence of embolism of peripheral vessels or pulmonary vessels while maintaining sinus rhythm, especially in young people; episodes of syncope, dyspnea, arterial hypotension and tachycardia (with obstruction of valve openings). Non-specific inflammatory markers: fever, anemia, accelerated ESR, increased C-reactive protein and interleukin-6. Additional symptoms include: joint pain in various locations; cyanotic network on the skin or livedo; Raynaud's syndrome (spasm of capillaries in the area of the fingertips, sometimes the ears, chin and nose, causing coldness, lack of sensitivity

and discoloration of the affected parts of the body); irregular heartbeat. A critical increase in the tumor size can cause obstruction of intracardiac blood flow. Most often, the formation is located in the left atrium, so it can completely or partially block the mitral valve or the pulmonary veins. This leads to the following symptoms: progressive dyspnea, cough, hemoptysis, rapid heartbeat, leg swelling, dizziness and loss of consciousness. When the myxoma is localized near the tricuspid valve, signs of right ventricular failure develop: edema, dyspnea, acrocyanosis, swelling of the jugular veins, hepatomegaly. It is important to note that small myxomas can be asymptomatic, complicating timely diagnosis. Extracardiac manifestations often imitate systemic vasculitis, collagenoses or chronic inflammatory processes. Tumor production of proinflammatory cytokines (especially interleukin-6) causes a systemic inflammatory response with cachexia, subfebrile condition, myalgia, arthralgia. These symptoms usually regress after radical tumor removal. Patients with constitutional symptoms have statistically larger tumor sizes, a tendency to multiple growth and recurrent course. Since myxomas are loose formations, some patients develop arterial embolism due to tumor fragmentation, which sometimes becomes the first manifestation of the disease. Thus, soft tissue polypoid myxomas more often give rise to embolism. Cerebral embolic events include ischemic strokes and transient ischemic attacks. The development of atrial fibrillation significantly potentiates the thromboembolic risk.

In this regard, timely verification of the cardiac source of embolism and radical surgical treatment as the optimal strategy for secondary prevention are of great importance.

Echocardiography (EchoCG) is recognized as the main method for verifying cardiac myxoma. Despite the higher diagnostic efficiency of transesophageal access, transthoracic two-dimensional echocardiography is usually sufficient for the initial diagnosis. The method allows determining the topography of the neoplasia, its linear parameters and degree of mobility. Differential diagnosis with a left atrial thrombus is of key importance. Thrombotic masses are usually localized in the posterobasal parts of the atrium and are characterized by a multilayered structure, while the presence of a vascular pedicle and high mobility are pathognomonic signs of myxoma. Doppler echocardiographic examination reveals hemodynamic disturbances corresponding to mitral stenosis and regurgitation. Transesophageal echocardiography has a higher resolution compared to the transthoracic technique, which allows visualization of small formations with a diameter of 1-3 mm. Angiocardiology occupies a significant place in the diagnostic algorithm. The pathognomonic radiological sign is the detection of a persistent rounded filling defect with smooth contours, which is flown around by a contrast agent. If echocardiography is insufficiently informative, computed tomography and magnetic resonance imaging methods are used. These methods allow: to evaluate the characteristics of pericardial structures, determine the presence of invasive growth in the main vessels and mediastinal structures, conduct differential diagnostics between myxomas, malignant neoplasms and intracardiac thrombi based on the analysis of density characteristics. Diagnostic difficulties arise with small tumor sizes, its atypical localization or unclear indication of the location.

Myxomas should be differentiated from heart valve diseases, cardiomegaly, septic endocarditis, and heart rhythm disorders. This heart pathology should be suspected in systemic and pulmonary embolism, periodic syncopal states, and heart failure. Myxomas can lead to complications such as: chronic heart failure, sudden death, cardiac arrhythmia, infectious disease, embolization, cardiac rupture, and myocardial infarction. The only method of treating cardiac myxomas is surgical intervention. Surgical treatment is indicated in the shortest possible time after diagnosis verification, due to the high risk of developing life-threatening complications. Verification of the diagnosis requires mandatory histological examination of all removed emboli. Modern cardiac surgeries for myxomas are classified as routine, with a favorable prognosis for survival and maintaining working capacity. Complete surgical sanitation is achieved. Refusal of surgical treatment is associated with an unfavorable prognosis. Family members with documented cases of myxomas are subject to mandatory screening, since embolization is the most significant complication, especially in familial forms of the disease. The duration of the clinical course is determined by the time of diagnosis. In sporadic cases, a favorable prognosis is characteristic with a 1% risk of recurrence, while in familial forms, up to 10% risk of relapse or the appearance of a neoplasm of a different localization is noted. Sudden mortality, reaching 15%, is mainly due to systemic or coronary embolism or acute heart failure due to atrioventricular valve obstruction.

As a clinical example, our own case of observation of a patient with a left atrial myxoma is presented. Patient A., 44 years old, nurse. Height 167, weight 70 kg. No bad habits. During a medical examination on 30.04.2019, atrial fibrillation was detected for the first time on the electrocardiogram (ECG), due to which the patient was taken to the hospital. The patient complained of weakness, shortness of breath during physical exertion (climbing to the 2nd floor), palpitations, associated with detraining, and was not examined. According to the patient, rhythm disturbances appeared over the past 3-4 months. Concomitant diseases: chronic venous insufficiency, varicose veins of the lower extremities, compensation stage. In the anamnesis, it is noteworthy that over the past 3 months, the patient noted the appearance of short-term subfebrile temperature, in the general blood test (CBC) an increase in ESR to 25 mm / hour, did not consult a doctor, took antipyretics. During an examination in the hospital during echocardiography according to the standard method from 05/17/2019, a volumetric round hyperechoic mobile formation (myxoma) of an oval shape, measuring 3.6 x 2.3 cm, is visualized. The tumor has a stalk, is fixed in the area of the oval fossa, mobile, with prolapse through the mitral valve into the cavity of the left ventricle, fixed on a stalk in the projection of the open oval window (figure). Ejection fraction is 65%, right atrium 42/49 mm, left atrium 51/55 mm, right ventricle 25/41 mm, left ventricle: EDS/ESR 43/23 cm, EDV/ESR 81/19 ml, IVS 16/9 mm, LVP 10/15 mm, aorta 32 mm, aortic valve is tricuspid, gradient 2.98, orifice area 3.3 mm, mitral valve cusps are unchanged. Tricuspid valve - grade 1 regurgitation. Mean pulmonary artery pressure is 31 mm Hg. The patient was hospitalized for surgical treatment with a diagnosis of left atrial myxoma, permanent atrial fibrillation, type 2A.

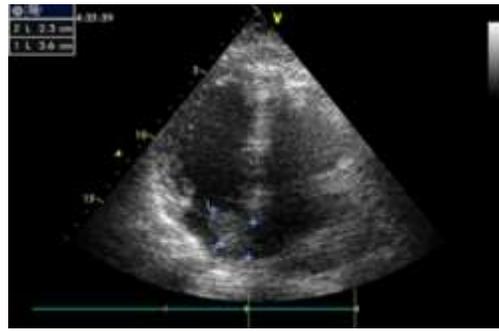


Figure: Myxoma of the left atrium

On ECG (upon admission): atrial fibrillation with a heart rate of 96 beats/min, normal position of the electrical axis of the heart, P-Q - 0.16 s, QRS - 0.08 s, QRST - 0.36 s. In the complete blood count on 05/14/2019: Hb 100 g/l, Erythrocytes 2.96x10¹² g/l, Leukocytes 13.5x10⁹ g/l, Platelets 180x10⁹ g/l, ESR 40 mm/h.

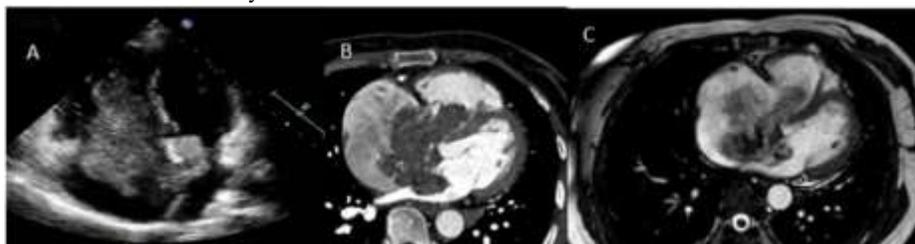
Biochemical tests and general urine analysis are normal. On May 30, 2019, the patient underwent surgical removal of the myxoma and ligation of the left atrial appendage. The picture of myxoma was histologically confirmed. The wound healed by primary intention. After surgical removal of the myxoma, the patient continued to have a permanent form of AF. 08/05/2019, the patient is hospitalized for cryoablation of the pulmonary vein orifices. After successful surgical treatment, the patient took amiodarone and oral anticoagulants (rivaroxaban) for a month. After a follow-up examination a month later, the drugs were discontinued. Three months after discharge from the hospital, the patient underwent a standard clinical and diagnostic examination, including daily Holter ECG monitoring (HM ECG). According to the results of HM ECG, no heart rhythm disturbances were detected. Sinus rhythm with an average frequency of 74 beats / min. Currently, the woman feels well, has no complaints. The patient's condition is satisfactory, her ability to work has been fully restored. Sinus rhythm is recorded on the ECG.

Conclusion. The presented clinical case is an example of accidental detection of myxoma in connection with the examination of a patient for atrial fibrillation. The features of the course of the disease in this patient were low-symptom dyspnea, weakness, palpitations, which did not bother the patient. The woman did not consider it necessary to consult a doctor

and explained her condition by asthenia. It should be noted that the appearance of dyspnea in young patients without concomitant diseases requires a detailed examination. In this case, the developed atrial fibrillation can be considered a secondary disease that developed against the background of myxoma of the left atrium. As mentioned above, the presence of a tumor can lead to the formation of chronic inflammation, which maintains the concentration of cytokines in the blood. This is how we explain the episodes of subfebrile temperature, increased ESR and leukocytosis in our patient. The appearance of atrial fibrillation became the reason for referring the patient for an in-depth examination, in connection with which the correct diagnosis was made. Thus, despite the increased diagnostic capabilities, the disease is often diagnosed late, which can be fatal for the patient. It is important to remember about cardiac tumors in order not to lose clinical alertness when assessing the clinical picture of the disease and objective data, about timely referral of patients for examination to detect the tumor and treatment.

2. A clinical case from China, published in the Asian Journal of Surgery.

A 53-year-old female patient with no history of chronic diseases. She was admitted to the emergency department with complaints of tachycardia and dizziness. Laboratory examination: all parameters are within normal limits. ECG: tachycardia. According to ECHO-CG (see Figure. A), CT angiography (see Figure.), MRI (see Figure. C): a space-occupying biatrial lesion with invasion into the right ventricle (biatrial myxoma?)



Cardiac catheterization and endomyocardial biopsy were performed. Histological conclusion: myxoma. Treatment: surgical removal of myxoma.

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