DOI: 10.20514/2226-6704-2022-12-5-389-393
EDN: VVORKA
УДК 616.125.2-006.325-036.1-07

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КЛИНИЧЕСКОЕ НАБЛЮДЕНИЕ БЕССИМПТОМНОЙ МИКСОМЫ ЛЕВОГО ПРЕДСЕРДИЯ

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Clinical Observation of Asymptomatic Left Atrial Myxoma

Резюме

Миксомы являются наиболее распространенным типом первичных доброкачественных опухолей сердца у взрослых, частота которых в популяции по данным аутопсий составляет около 0,2 %. Миксомы развиваются из мультипотентной мезенхимы и обычно представляют собой недифференцированное предсердное образование, имеющее ножку и прикрепленное к овальной ямке на левой стороне межпредсердной перегородки. Частое бессимптомное течение заболевания затрудняет своевременную диагностику и лечение. Представленное клиническое наблюдение демонстрирует случайное выявление миксомы левого предсердия у пациентки 68 лет с последующим успешным оперативным вмешательством.

Ключевые слова: *миксома сердца, диагностика, клиническое наблюдение, бессимптомное течение*

Конфликт интересов

Авторы заявляют, что данная работа, её тема, предмет и содержание не затрагивают конкурирующих интересов

Источники финансирования

Авторы заявляют об отсутствии финансирования при проведении исследования

Статья получена 27.04.2022 г.
Принята к публикации 08.06.2022 г.

Для цитирования: Сергеева В.А., Толстов С.Н., Сычкова Е.Д. КЛИНИЧЕСКОЕ НАБЛЮДЕНИЕ БЕССИМПТОМНОЙ МИКСОМЫ ЛЕВОГО ПРЕДСЕРДИЯ. Архивъ внутренней медицины. 2022; 12(5): 389-393. DOI: 10.20514/2226-6704-2022-12-5-389-393 EDN: VVORKA

Abstract

Myxomas are the most common type of primary benign cardiac tumor in adults, with an incidence of about 0.2 % in the population at autopsy. Myxomas develop from multipotent mesenchyme and are usually an undifferentiated, pedunculated atrial mass attached to a fossa ovale on the left side of the atrial septum. Frequent asymptomatic course of the disease complicates timely diagnosis and treatment. The presented clinical observation demonstrates the accidental detection of left atrial myxoma in a 68-year-old patient with subsequent successful surgical intervention.

Key words: *cardiac myxoma, diagnosis, clinical observation, asymptomatic course*

Conflict of interests

The authors declare no conflict of interests

Sources of funding

The authors declare no funding for this study

Article received on 27.04.2022
Accepted for publication on 08.06.2022

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For citation: Sergeeva V.A., Tolstov S.N., Sychkova E.D. Clinical Observation of Asymptomatic Left Atrial Myxoma. The Russian Archives of Internal Medicine. 2022; 12(5): 389-393. DOI: 10.20514/2226-6704-2022-12-5-389-393 EDN: VVORKA

BP — blood pressure, CT — computed tomography, ECHO-CG — echocardiography, LA — left atrium, MRI — magnetic resonance imaging

Introduction

Until the middle of the 20th century, intravital diagnosis of space-occupying lesions in heart was scarcely performed due to the lack of the required instrumental examination methods. With the implementation of angiocardiography in 1951, the ability to detect cardiac tumors in patients has increased significantly. However, surgical treatment of space-occupying intracardiac lesions consistently failed [1]. A breakthrough event was the development of heart-lung machine and the successful surgical removal of cardiac myxoma in 1954 performed by a Swedish cardiac surgeon Clarence Crafoord [2]. A new milestone in the diagnosis of cardiac tumors was the implementation of echocardiography into clinical practice in 1959 [3]. It is still the basic method in the diagnosis of any cardiac pathology. Comprehensive information about the size, position and the suspected nature of an intracardiac mass can be obtained using computed tomography (CT) and magnetic resonance imaging (MRI) of heart with contrast enhancement.

Myxomas are the most prevalent primary benign cardiac tumors that account for up to 80 % of all diagnosed intracardiac neoplasms [4]. Myxomas are most often located in left atrium (LA) (up to 75 %); about 2-3 times as frequently in women, in the age group of 30 to 60 years [5]. In most cases, this disease is detected during occasional examination, however, with no timely treatment, intracardiac hemodynamics may be disturbed with the development of progressive heart failure and embolic complications that can result in disability and death of the patient.

Etiology of this disease is not completely understood. It is assumed that herpes simplex virus type I is a trigger; it can cause chronic endocarditis and neoplastic transformation, since antigens and DNA of herpes simplex virus type I are detected in 70 % of patients with myxoma [6]. Cardiac myxomas are also found in patients with a mutation in PRKAR1A gene that is located on the long arm of chromosome 17 (17q23-q24) as part of a rare hereditary neoplastic syndrome with an autosomal dominant mode of inheritance, the so-called Carney complex [7]. The clinical signs of this genetic pathology also include spotty skin pigmentation and myxomas, active tumors of endocrine system (nodular hyperplasia of adrenal cortex, breast fibroadenoma, testicular tumors, pituitary tumors with gigantism or acromegaly) and the sheaths of nerve trunks. About 10 % of all heart myxomas are a sign of Carney complex and are considered to be the main clinical criteria of this disease; they are found in 30-40 % of cases of this pathology. However, to diagnose Carney complex, two or more major clinical criteria, or one major and one additional (hereditary) factor are required [7].

The size, shape and weight of a myxoma can vary over a wide range. In various clinical observations report on tumors from a few millimeters to 16 centimeters in diameter and from 2 to 250 grams in weight [5]. World Health Organization defines a cardiac myxoma as a neoplasm consisting of stellate or plump, cytologically soft mesenchymal cells located in the myxoid stroma. Cardiac myxoma cells often form rings, nests, and linear syncytia that originate from vascular structures. Fibrosis, calcification and organized thrombosis are common; however, mitoses are scarcely found. A myxoma usually has a pedicle and is attached to the oval fossa on the left side of interatrial septum [8].

Regardless of the presence or absence of clinical signs of cardiac myxoma, the only method of treatment is the surgical removal of this tumor. Surgery should be performed by a skilled cardiac surgeon, since incomplete removal of a myxoma may result in its relapse [5]. Following the surgical treatment, regular echocardiography is recommended to monitor the patient's condition. According to the literature, sporadic myxoma relapse develops in about 3 % of tumors [5]. It may occur months or years after the first surgery. Ricardo Oliveira et al. [9] presented a study where 19 patients underwent surgery for cardiac myxoma; on average, 2 relapses were diagnosed over a period of 5.2 ± 3.7 years (relapse rate was 10.5 %).

We present our own clinical experience of follow-up a patient with asymptomatic cardiac myxoma; the neoplasm was found during computed tomography of thoracic organs as part of examination for COVID-19.

Case report

In June 2021, patient M., female, 68, a resident of the Saratov region (Ershov) had malaise, weakness, fever, sore throat, palpitations. She visited a therapist at a local clinic to exclude novel coronavirus infection. To diagnose SARS-CoV-2, nasopharyngeal swabs were taken. Positive PCR result confirmed COVID-19. Outpatient treatment was provided. The patient underwent chest CT to exclude lung damage. No pulmonary parenchyma infiltration was found, however, the change in the size of mediastinum was observed. The mediastinum was expanded due to the dilatation of heart chambers, mainly of LA to 5 cm with decreased density of X-ray radiation

and small linear high-density inclusions. A routine additional examination in the Regional Cardiac Surgery Center was recommended after the recovery from COVID-19. The patient had no apparent cardiovascular clinical signs. For about two years, she has high blood pressure (BP) up to 150 and 100 mm Hg maximum; she regularly receives antihypertensive therapy (indapamide + bisoprolol), and complies with the therapy. Therapy-associated BP is 120 and 80 mm Hg. Besides, she regularly receives acetylsalicylic acid (ASA) and atorvastatin. After the novel coronavirus infection, she had dyspnea at moderate exercise and asthenia. The patient was hospitalized to the Regional Cardiac Surgery Center in August 2021 for further examination. At admission, the patient had almost no complaints. From past medical history: the patient grew and developed according to her age. Comorbidities: chronic non-obstructive bronchitis, chronic erosive gastritis (not tested for Helicobacter pylori), varicose veins disease. Past surgeries — cholecystectomy in 2005, epidemiological and allergic anamnesis within normal, bad habits — denies. Upon admission, negative PCR for SARS-CoV-2.

Physical examination

Satisfactory condition. Active position. Clear consciousness. Height 165 cm, weight 85 kg. Body mass index 31.22 kg/m^2 — class 1 obesity. Respiratory organs: respiratory rate 18 per minute, vesicular respiration is heard in all lung fields. No adventitious breath sounds. Circulatory organs: apex beat is felt in 5 intercostal space along the midclavicular line. On percussion, borders of relative cardiac dullness are expanded along the upper border to the 2nd rib (enlarged LA). On auscultation, muffled rhythmic heart sounds. Heart rate coincides with the pulse and is 67 bpm. BP in both brachial arteries is 110 and 70 mm Hg.

Results of laboratory tests and instrumental examinations

Results of laboratory tests revealed no significant deviations. Acute phase indicators were within normal. Due to the constant intake of 20 mg of atorvastatin per day, total cholesterol was 3.2 mmol/L, and low-density lipoprotein cholesterol — to 1.6 mmol/L.

Results of electrocardiography is presented in **Fig. 1.** Conclusion: PQ 0.2 s, QRS 0.08 s, QT 0.4 s, sinus rhythm with heart rate of 92 per minute. Vertical QRS axis. Slowing of atrioventricular conduction. Enlarged atria. Left ventricular hypertrophy.

During inpatient examination, transthoracic echocardiography (ECHO-CG) was performed; it revealed increased LA (ESD 57 mm; normal range 27-38 mm). Mass lesion in LA, large (59 x 48 x 30 mm), almost filling its entire cavity. Mass lesion of homogeneous, hypoechoic structure, with single calcifications, sharp smooth contours, poorly mobile with blood flow, however, with no signs of mitral intracardiac obstruction. This mass is supposed to be attached to the interatrial septum with a broad pedicle. No significant cardiac valve dysfunction was found. The size of LV cavity and its contractility were within normal. Left ventricular myocardial mass index 81 g/m^2 (normal range 44 — 88 g/m^2). Inferior vena cava is of normal size and with normal respiratory variation. No effusion in pericardial cavity.

To detect the degree of atherosclerotic lesions of vascular bed and to exclude thrombotic lesions of the deep veins of lower limbs, coronary angiography and duplex scanning of brachiocephalic arteries and of the vessels of lower limbs were performed. There were no indications for coronary revascularization. Early signs of atherosclerosis were found in brachiocephalic arteries. Varicose veins of lower limbs were observed. Deep veins were patent. There were no signs of valvular insufficiency or deep vein thrombosis of lower limbs.

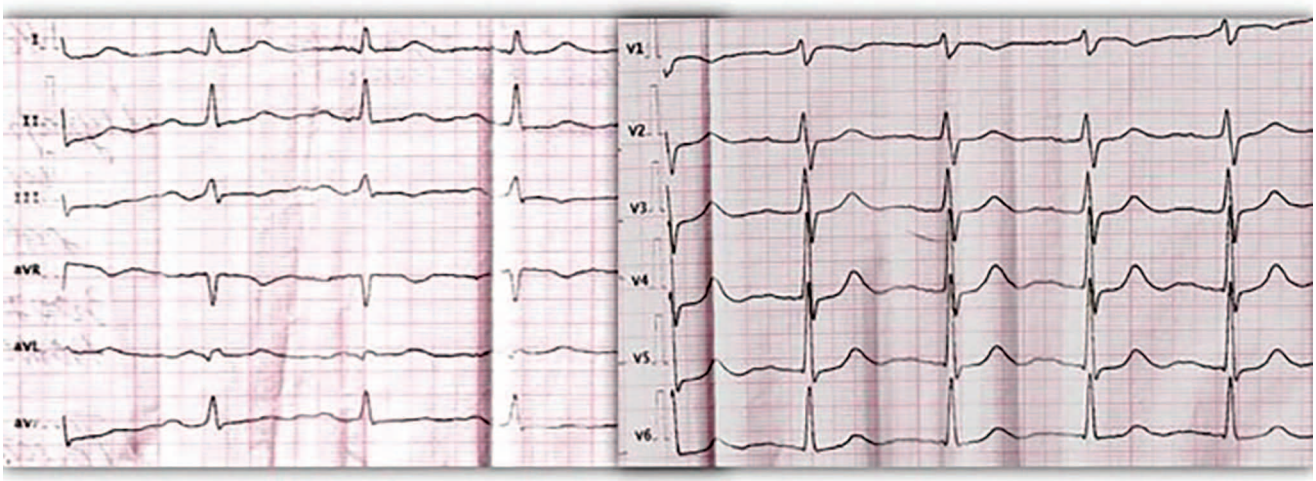

Figure 1. *ECG of patient M., 68 y.o.*

Minor atherosclerotic changes in the arteries of lower limbs were found. Results of the ultrasound examination of abdominal organs revealed steatohepatosis and non-uniformity of the contours of pancreas.

In order to visualize a mass lesion in LA, CT with IV bolus contrast enhancement (Ultravist) was performed (**Fig. 2**). According to CT data, a contrasting defect was visualized in LA; it was caused by a soft-tissue mass lesion with uneven tuberous contours, of inhomogeneous structure (due to inclusions of small calcifications). The size of this mass lesion was up to 60 x 55 x 61 mm (mediolateral, sagittal, vertical), volume — 117 mL, it occupied almost the entire LA cavity. The mass lesion described is broadly adjacent to the middle third of interatrial septum; it spreads in the mouth of right superior pulmonary vein with no significant stenosis of lumen; it does not accumulate contrast agent. LA volume with auricula was 225 mL. Conclusion: LA myxoma.

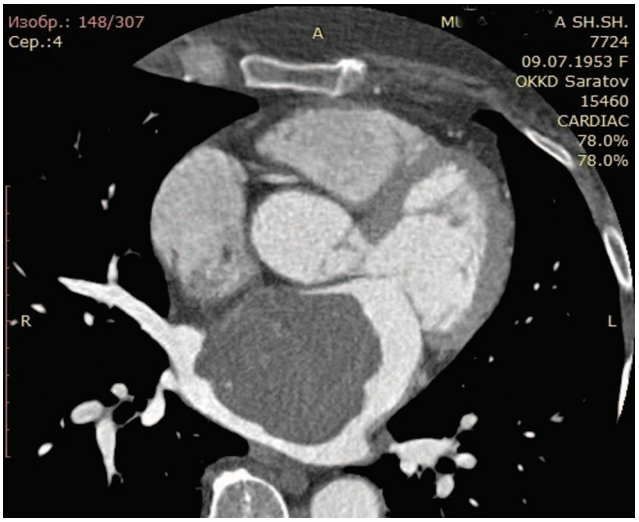

Figure 2. *Cardiac computed tomography with contrast of patient M., 68 y.o.*

Treatment

On September 3, 2021, the patient underwent routine surgical treatment: removal of LA mass, atrial septal defect closure with xenopericardium patch in conditions of artificial circulation and pharmaco-cold cardioplegic protection. The mass removed — myxoma (no histological signs of malignancy found) is presented on **Fig. 3**. In the early postoperative period, the patient had a rhythm disorder — paroxysm of atrial fibrillation with a rate of 65–170 bpm. Successful pharmacological cardioversion with amodarone was performed. Control CT and ECHO-CG revealed no intracardiac hemodynamic disorders. The patient was discharged for further rehabilitation at the outpatient stage with sinus rhythm, stable hemodynamics, in a satisfactory condition on September 15, 2021. 6 months after the surgery the patient confirmed stable and satisfactory condition during a telephone call. She has a usual lifestyle. She had no irregular heart function. She adhered to all recommendations.

Discussion

In about 20 % of cases, cardiac myxomas are characterized by an asymptomatic course and slow progression what complicates the early diagnosis of this disease [8].

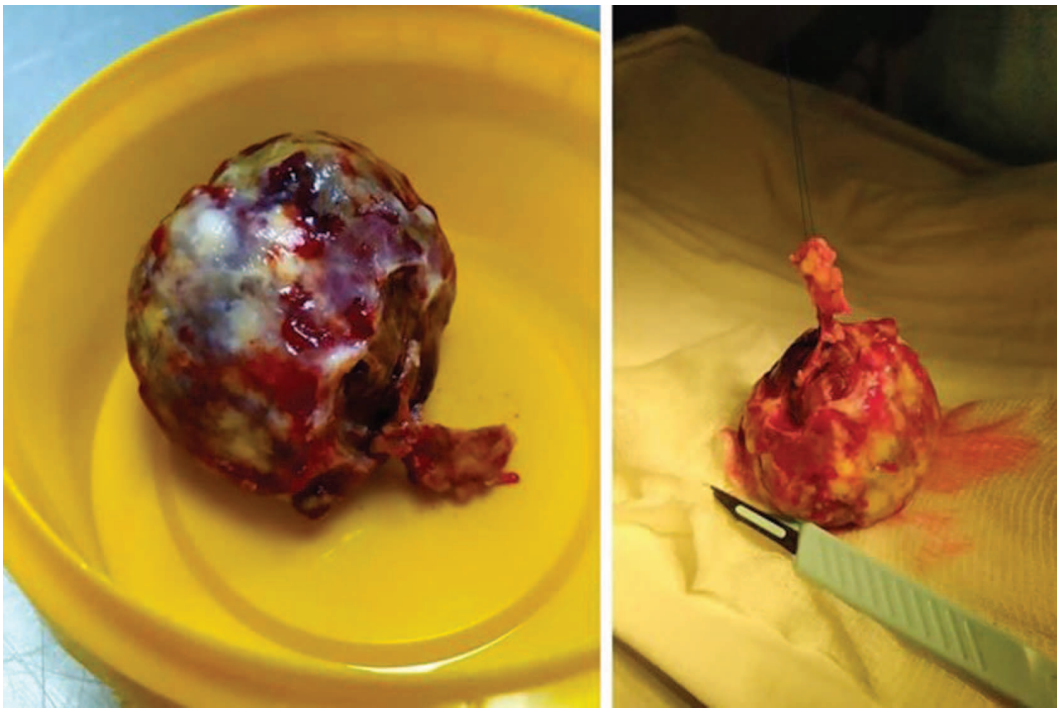

Figure 3. *Intracardiac tumor removed during surgery — myxoma*

In the above case report, a mass lesion was found incidentally, during the examination of lungs as part of examination for COVID-19. Despite the large size of the intracardiac mass, there were no clinical signs. According to many literature sources, the asymptomatic course of this process depends not so much on the size of mass lesion, but to a greater extent, on the presence of intracardiac hemodynamic disorders. In regard to LA myxomas, on the presence of mitral obstruction [5, 10]. Clinical signs include hemodynamic signs (signs of heart failure, arrhythmias, sudden cardiac death), signs of systemic embolism (peripheral vascular embolism, transient ischemic attacks or strokes) and constitutional signs (fever, weight loss, arthralgia, asthenia) [10]. The onset of these symptoms, of course, leads to active diagnostic search and diagnosis.

Cardiac myxoma does not belong to diseases that are hard to diagnose. To find a mass lesion, one should have any available imaging method for examination of the heart (ECHO-CG, CT, MRI).

Despite the asymptomatic course, surgical treatment is the "gold standard" for the management of myxoma, since complications associated with further tumor growth can be fatal for a patient.

Conclusion

Chest CT for the novel coronavirus infection contributed to the timely diagnosis of asymptomatic LA myxoma, and the successful surgery made it possible to avoid severe cardiovascular complications. Despite the outpatient follow-up for arterial hypertension, no ECHO-CG was performed over several years. Timely performed, this routine instrumental examination could contribute to the earlier detection and surgical management of cardiac myxoma.

Вклад авторов:

Все авторы внесли существенный вклад в подготовку работы, прочли и одобрили финальную версию статьи перед публикацией

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Сычкова Е. Д.: написание, редактирование текста и утверждение финального варианта статьи

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All the authors contributed significantly to the study and the article, read and approved the final version of the article before publication

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