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MYXOMA OF THE HEART: MORPHOLOGY, DIAGNOSIS, SURGICAL TREATMENT. A CLINICAL CASE OF VILLOUS MYXOMA OF THE LEFT ATRIUM

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Abstract. Myxoma is a tumor, which is the most common nosological form among the primary neoplasms of the heart, usually characterized by its goodness, but with frequent manifestations of relapses. Myxomas account for about half of all primary neoplasms of the heart, most of them occur in the left atrium and only occasionally attach to the mitral valve. These neoplasms may be manifested by a combination of obstruction of blood flow, systemic embolization and constitutional symptoms. Gender differences in the structure of epidemiology of the heart mix indicate the following: the disease is more common mainly in women than in men. Morphologically, myxomas of the heart are very diverse and heterogeneous. The article describes in detail the villous myxomas of the heart, which most often appear in the structure of this pathology. Data on localization, macroscopic and microscopic structure are given. We also present a clinical case of the Department of Cardiac Surgery of the Leningrad Regional clinical hospital. Also, this review presents a clinical case of villous myxoma detected (during echocardiography) and surgically resected, in an 80-year-old patient, in the Department of Cardiac Surgery of the Leningrad Regional clinical hospital. The tumor was surgically resected and histologically classified as a villous myxoma.

Key words: heart; myxoma of the heart; myxoma villosa; primary heart tumors; benign heart tumors; tumor morphology; diagnosis of tumors; surgical treatment of tumors; clinical case.

МИКСОМЫ СЕРДЦА: МОРФОЛОГИЯ, ДИАГНОСТИКА, ОПЕРАТИВНОЕ ЛЕЧЕНИЕ. КЛИНИЧЕСКИЙ СЛУЧАЙ ВИЛЛЕЗНОЙ МИКСОМЫ ЛЕВОГО ПРЕДСЕРДИЯ

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Резюме. Миксома — опухоль, представляющая собой самую распространенную нозологическую форму среди первичных новообразований сердца, обычно характеризующуюся своей доброкачественностью, но с нередкими

проявлениями рецидивов. Миксомы составляют примерно половину всех первичных новообразований сердца, большинство из них встречаются в левом предсердии и лишь изредка прикрепляются к митральному клапану. Эти новообразования могут проявляться сочетанием обструкции кровотока, системной эмболизацией и конституциональными симптомами. Гендерные различия в структуре эпидемиологии миксом сердца говорят о следующем: заболевание чаще встречается преимущественно у женщин, нежели у мужчин. Морфологически миксомы сердца очень разнообразны и неоднородны. В статье подробно описаны виллезные миксомы сердца, которые наиболее часто фигурируют в структуре данной патологии. Приведены данные о локализации, макроскопическом и микроскопическом строении. Мы также представляем клинический случай отделения кардиохирургии Ленинградской областной клинической больницы. В данном обзоре также представлен клинический случай виллезной миксомы выявленной (при проведении ЧПЭхоКГ) и хирургически рецезированной, у пациентки 80 лет, в отделении кардиохирургии Ленинградской областной клинической больницы. Опухоль была хирургически резецирована и гистологически классифицирована как виллезная миксома.

Ключевые слова: сердце; миксома сердца; виллезные миксомы; первичные опухоли сердца; доброкачественные опухоли сердца; морфология опухолей; диагностика опухолей; хирургическое лечение опухолей; клинический случай.

INTRODUCTION

Myxoma is the most common nosological form among the primary cardiac neoplasms (Table 1), usually characterised by its benign nature but with frequent recurrences [7, 10, 35].

At least 14 different names of primary cardiac tumours are known, but not all of them occur in young children.

In their overall presentation, cardiac tumours are rare, with an incidence of only 0.027–0.05 % among newborns in the first large series of observations [22].

The low percentage of occurrence and, first of all, detectability of cardiac tumours, including cardiac myxomas, was associated with hemodynamically insignificant influence of the latter on the general hemodynamics and, thus, did not manifest itself clinically. But in the era of introduction of screening and highly specific diagnostic methods: echocardiography, computer tomography, magnetic resonance tomography — the epidemiological significance, the specific weight of diagnoses increased in relation to primary data many times.

In the early 2000s, however, and at present, myxomas represent about 30-80% of all primary cardiac tumours [9, 23, 30, 32, 38], and in the 2020s, myxomas remain the most common primary cardiac tumour [30].

More than three quarters of primary cardiac neoplasms are benign, with myxomas and rhabdomyomas being the most common cardiac tumours seen in adults and children (Table 2). Primary malignant cardiac tumours are extremely rare, whereas metastatic lesions can be observed in approximately 8% of patients dying from oncology [5, 29].

More than 75% of myxomas arise in the left atrium, either in the region of the left atrioventricular (mitral) foramen (annulus) or at the oval fossa border of the interatrial septum; 20% arise from the right atrial wall, and 5% originate from both the atrium and ventricle [30].

The majority (75–86 %) of myxomas are localised in the left atrium (left atrial myxoma was first described in 1845 [26]) (Fig. 1), about 70 % of them are located in the oval fossa (Fig. 2), less frequently in the posterior and anterior

Table 1

McAllister H.A. and Fenoglio J.J. classification of primary cardiac tumours., n = 444 (100%) (Atlas of Tumor Pathology, 1978)

Benign cardiac tumors			Malignant cardiac tumors		
the type of tumour	Amount		The type of tumour	Amount	
	abs.	%		abs.	%
Myxoma	130	29,3	Angiosarcoma	39	8,8
Lipoma	45	10,1	Rhabdomyosarcoma	26	5,9
Papillary fibroelastoma	42	9,5	Malignant mesothelioma	19	4,3
Rhabdomyoma	36	8,1	Fibrosarcoma	14	3,2
Fibroma	17	3,8	Malignant lymphoma	7	1,6
Hemangioma	15	3,4	Extracellular osteosarcoma	5	1,1
Teratoma	14	3,2	Neurogenic sarcoma	4	0,9
Tumors of the atrioventricular nodal region	12	2,7	Malignant teratoma	4	0,9
Granular cell tumor	3	0,7	Timoma	4	0,9
Neurofibroma	3	2,7	Leiomyosarcoma	1	0,2
Lymphangioma	2	0,5	Liposarcoma	1	0,2
			Synovial sarcoma	1	0,2
TOTAL	319	72	TOTAL	125	28

Table 2

The incidence of cardiac tumours in children

Indicators	Total number of observations, abs. (%)					
	Takach T. et al., 1996[1]	Salle D. et al., 1999[2]	Becker A.E. 2000[3]	Freedom R.M.et.al., 2000[4]	Stiller B. et al., 2001[5]	Всего
Total number of observations	40	22	76(55)[6]	56	267	220
Benign tumours, including:	37 (92)	21 (95)	63 (83)	56 (100)	26 (100)	203 (92)
Rhabdomyoma	–	11	29 (27)	44	29	113(63)[7]
Fibroma	–	1	18 (13)	6	9	34(18,9)[8]
Myxoma	–	1	6 (2)	–	–	7(3,9)[8]
Hemangioma	–	1	4 (3)	–	2	7(3,9)[8]
Teratoma	–	2	1 (1)	1	2	6(3,3)[8]
Purkinje cell tumour	–	–	4 (4)	–	–	4(2,2)[8]
AV node mesothelioma	–	1	2 (1)	–	–	3(1,7)[8]
Lipoma	–	–	1 (0)	1	–	2(1,1)[8]
Multicystic hamartoma	–	–	–	1	–	1(0,5)[8]
Unclassified	–	4	–	3	–	7(3,9)[8]
Malignant tumours	3 (7,5)	1 (4,5)	13 (17)	–	–	17 (8)
Average incidence per year	1,14	1,22	4,47	3,11	2,6	2,3
Average incidence per period	1,2		3,4			

Note. [1] — 35 years of follow-up; [2] — 18 years of follow-up; [3] — 17 years of follow-up; [4] — 18 years of follow-up; [5] — 10 years of follow-up; [6] — number of patients younger than 1 year in parentheses; [7] — number of patients differs from number of tumours; [8] — among the four penultimate columns [12].

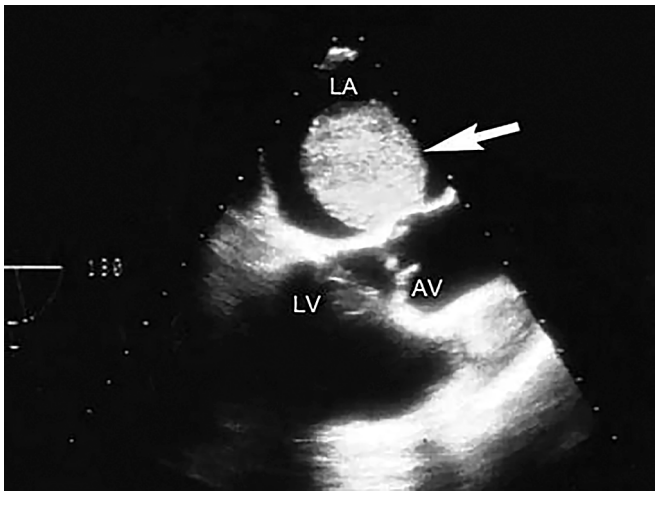

Fig. 1. Transesophageal echocardiography. Left atrial myxoma arising from the lower part of the interatrial septum: LV — left ventricle; LA — left atrium; AV — aortic valve [33]

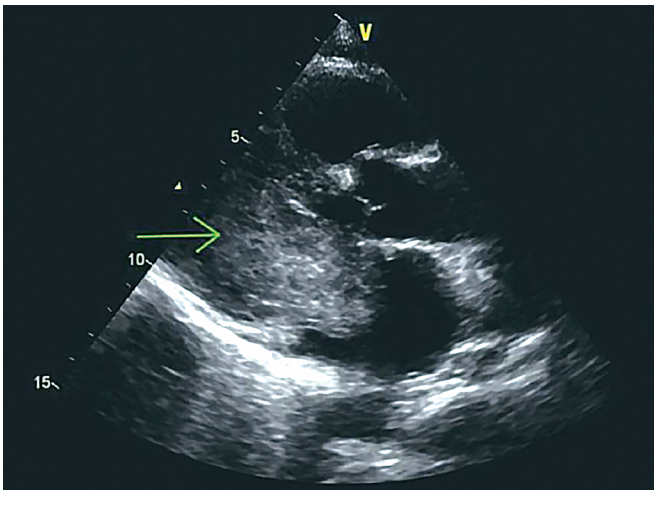

Fig. 2. Transthoracic echocardiography (long axis view) showed severe prolapse of left atrial myxoma (an arrow) into the left ventricle in a 56-year-old patient with fever of unknown origin [41]

wall of the atrium and in its auricle, 10–20 % in the right atrium (Fig. 3), often in the region of the oval fossa and Koch's triangle (Fig. 4) or on the eustachian valve or between the latter and the mouth of the coronary sinus. There are also descriptions of cardiac myxomas on the structures of the mitral, tricuspid valves, in the area of the pulmonary vein apertures [17, 25].

Complications from the sprouting of this type of cardiac tumours appear as a triad:

1. Obstruction (Fig. 5, *a*, *b*).

2. Embolisation (in a recent article, physicians from the Department of Neurology, Yangpu Hospital Tongji University School of Medicine, Shanghai, China, published a case of middle cerebral artery thromboembolism in a patient with

cardiac myxoma, which led to cerebral artery embolism with subsequent development of arterial aneurysm; it is noted that thrombolysis in such patients is an adequate therapy leading to restoration of blood flow and improvement of the patient's prognosis [3, 8, 28]).

3. Individualisation of the clinical picture (symptoms of intoxication: fever (Fig. 6), decreased appetite, malaise, arthralgia; weight loss; skin symptoms; aggravation of chronic diseases, their decompensation).

Gender differences in the structure of epidemiology of cardiac myxomas show the following: the disease is more frequent mainly in women than in men. The peak of morbidity in women is at 40–60 years of life, with tumour localisation in the ventricles, the age of patients is much younger [42].

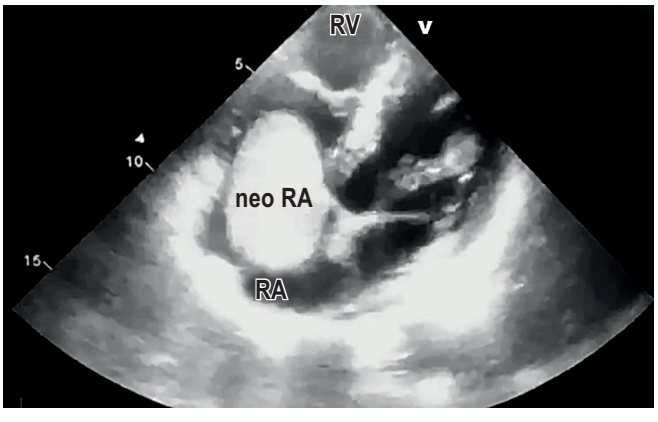

Fig. 3. **Hyperechogenic homogeneous homogeneous oval-shaped mass with smooth contour, vascularised, is visualised in the right atrial cavity. The diameter is 4.0 × 6.0 cm. The mass prolapses into the cavity of the right ventricle (RV) with the formation of obstruction of the RV inflow pathway. RA — right atrium. (Observation of the Leningrad Regional Clinical Hospital, 2018)**

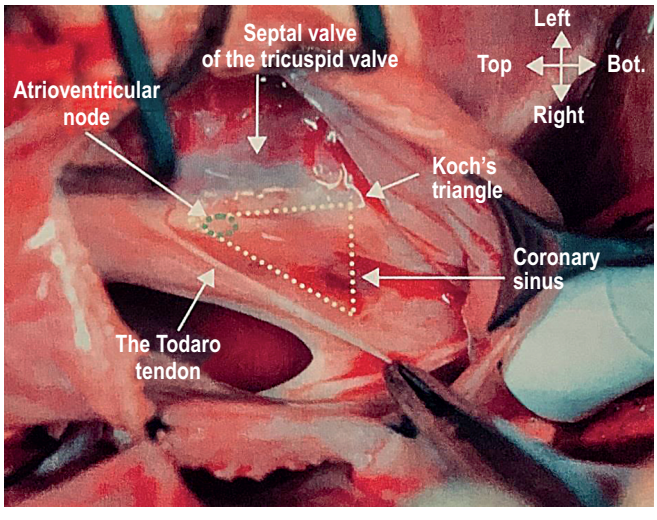

Fig. 4. **Intraoperative view of the heart with open oval window from the right atriotomy from the surgeon's side. Boundaries of Koch's triangle (yellow dashed line). The Todaro tendon is represented by the hypotenuse of Koch's triangle, located between the eustachian valve (the Todaro tendon is pulled up with forceps for better visualisation) and the attachment of the septal leaflet of the tricuspid valve [1]**

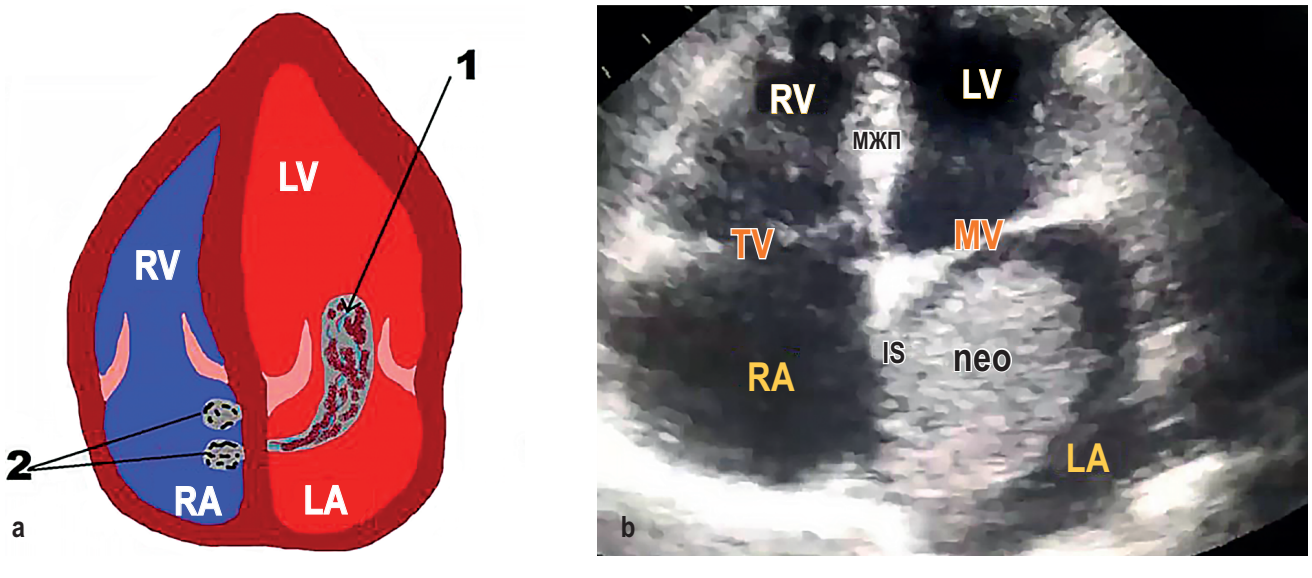

Fig. 5. **Left atrial (LA) myxoma elongated in length and flotating into the left ventricle (LV), small right atrial (RA) myxomas (a); transthoracic echocardiography of a patient with complaints of palpitations (b). An oval-shaped (neo), hyperechogenic, on a pedicle — mobile with a site of attachment to the middle third of the interatrial septum (IS) in the cavity of the LV. The diameter is 6.3 × 6.5 cm. The mass prolapsed into the left ventricular cavity (LV) with the formation of LV obstruction. MV — mitral valve; TV — tricuspid valve (Observation of the Leningrad Regional Clinical Hospital, 2017)**

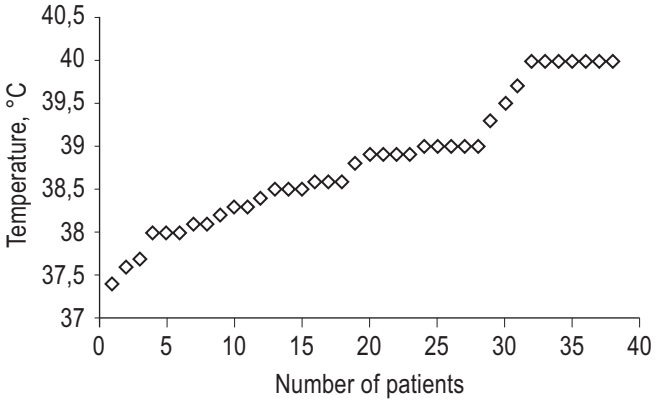

Fig. 6. Body temperature of patients with cardiac myxoma (fever of unknown genesis) [41]

Some studies show that the ratio of women to men is 2.05:1 and 0.75:1 for left and right atrial myxomas, respectively, and the specific weight of pathology detection in paediatric practice is very high at present [27, 31].

Myxomas are subdivided into three types: 1) sporadic, "non-familial" — the most common; 2) familial, transmitted by "inheritance"; 3) myxoma as part of a myxoma complex (Swiss-syndrome, Carnay-complex) — most often multiple myxomas [13, 15].

The etiology of myxoma occurrence remains unclear nowadays. Most authors identify three theories of cardiac myxomas, which in turn are also subject to doubt and criticism: 1) the theory of cardiac myxomas as a result of mural thrombus development; 2) the theory of viral genesis (the influence of Coxsackie B4 virus and various virus-like particles has been identified and is known in the literature; however, viral culture of these particles could not be obtained); 3) the theory of tumour genesis [10].

MORPHOLOGY (ON THE EXAMPLE OF SOFT, POLYP-LIKE CARDIAC MYXOMAS)

Benign cardiac tumours may originate from peri-, myo-, and endocardial rudiments, among which there are both organ-nonspecific, such as angiomas, fibromas, lipomas, etc., and organ-specific, such as myxoma, rhabdomyoma, and papillary fibroelastoma [2]. Myxomas are mesenchymal tumours of the heart, which most often grow from the oval fossa of the interatrial septum of the left atrium, have a pedicle and capsule, as well as an ovoid or lobular structure, according to L.A. Bokeria et al. (2003). Myxoma has a different structure and localisation (Fig. 7), but always originates from the endocardium with remnants of embryonic mucoid tissue or from the endothelium with subsequent myxomatous degeneration.

According to the macroscopic picture, cardiac myxomas are usually divided into two types: 1) soft, polypoid villous (Fig. 8); 2) dense oval-round (Fig. 9).

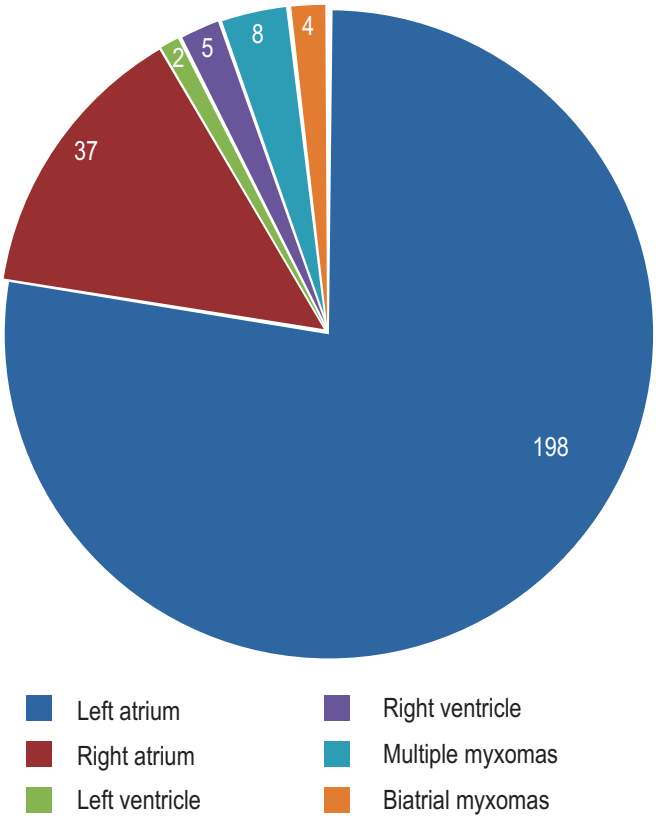

Fig. 7. Localisation of cardiac myxomas according to the data of the A.N. Bakulev National Centre of Cardiovascular Surgery Russian Academy of Medical Sciences [10]

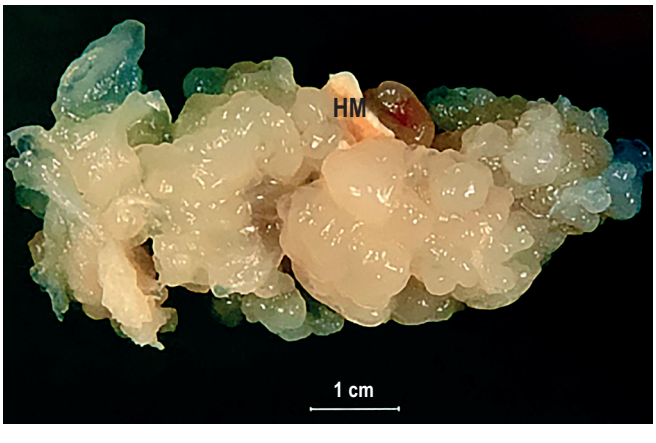

Fig. 8. Giant villous myxoma. Macroscopy. HM — heart myxoma [2]

In turn, soft, polypoid myxomas are divided into lobular, with gelatinous irregular surface and insignificant number of villi, and villous, built mainly of villi of gelatinous consistency. There are also mixed variants of macroscopic structure.

All myxoid soft tissue tumours within the benign structure are really quite common. All tumours of this type share the same myxoid appearance, while histogenetically and structurally these tumours have little in common. The myxoid properties of tumours are determined by the mucoid core matrix

enriched with polysaccharides during tumour transformation, which gives them a jelly-like macroscopic appearance [14, 16].

The morphological characterisation of polypoid villous myxomas is as follows:

1. According to K. Reynen et al. (1993) and M.L. Grebenc et al. (2000), the size of the tumours ranges from 0.6–1.5 cm to 15 cm, most commonly 5–6 cm.

2. A tumour pedicle is always presented, attached to the endocardium, the pedicle tissue contains vessels that supply the deep portions of the tumour tissue; the superficial portions are supplied directly from the lumen [2].

3. The tumour is jelly-like, villous, and of a bunched structure.

4. The shape is elongated, irregularly round, oval, round-oval (Fig. 10, a), sharply elongated (Fig. 10, b).

5. Smooth or slightly bumpy surface lined by a layer of mixed endothelial cells (Fig. 11, a), which may form invaginates (Fig. 11, b) and a network-like syncytial structure with cells lying deeper (Fig. 11, c).

6. Microscopy reveals small area haemorrhages, often with clusters of siderophages, attributed to repeated haemorrhages in the tumour tissue (Fig. 12).

7. Against the background of frequent traumatisation with subsequent haemorrhages, sclerosis of the tumour tissue is formed, which gives the tumour a whitish appearance and tissue density, and sometimes necrosis foci are observed (Fig. 13).

8. The tumour villi are lined with a single layer of endothelial cells, the tissue is friable, weakly basophilic with eosinophilic areas, mixoma cells in these structures are often arranged chaotically, but sometimes there is also an arrangement along the axis of the villi; blood supply of the villi is carried out in single cases or is absent at all.

9. With a large number and dense packing of villi — the solid structure of myxoma is observed.

10. Sometimes focal fibrosis is noted in tumours of this type due, most likely, to the reasons described in point 7.

DIAGNOSTIC

Cardiac myxomas are histologically benign, they can form hemodynamically significant pathology and be prognostically fatal because of their position. They can mimic not only any cardiac disease but also infectious, immunological and malignant processes. Thus, differential diagnosis should be made with cardiac valve pathology, acute and progressive chronic heart failure, cardiomegaly, infective endocarditis, cardiac rhythm and conduction disorders, syncope and signs of systemic and pulmonary embolism [16, 33, 34].

Symptoms depend on the size, mobility and location of the tumour. Echocardiography (the most accessible, relatively cheap method, which is the “gold standard” for the study of morphological structures of the heart), including transesophageal echocardiography, is the most important diagnostic tool. Computed tomography (CT) [39] and magnetic resonance imaging (MRI) can also be used as

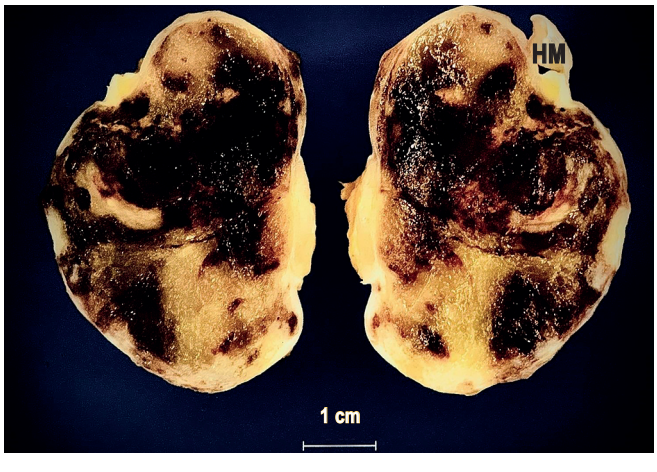

Fig. 9. **Biatrial dense myxoma. Macrophotography. HM — heart myxoma [2]**

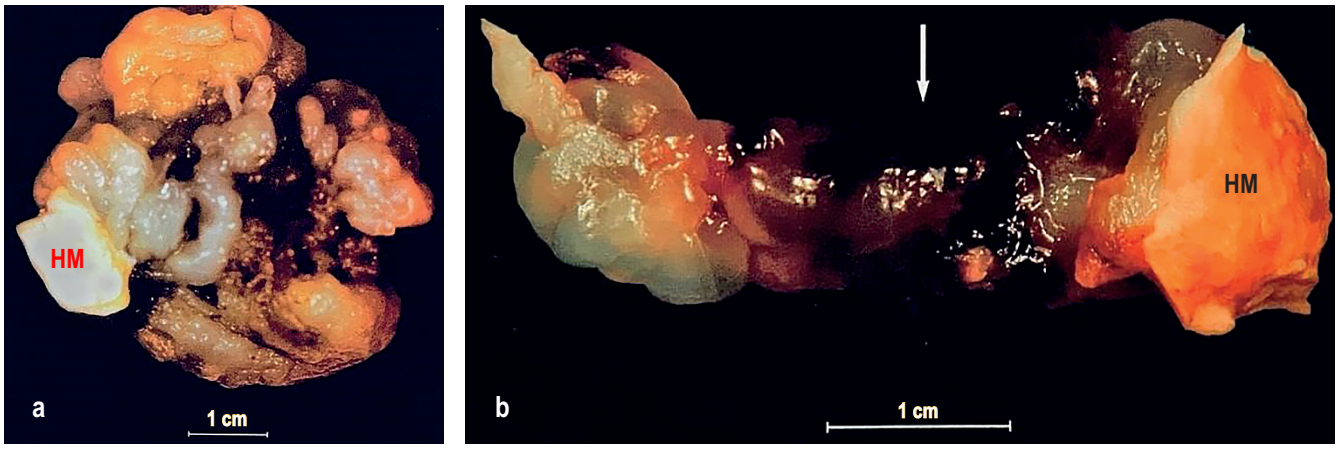

Fig. 10. **Medium-sized round soft myxoma (a); medium-sized soft gelatinous finger-shaped myxoma with haemorrhage in the middle third (b) (indicated by arrow). Makrophotographs. HM — heart myxoma [2]**

informative additional diagnostic methods. The use of coronary angiography in patients over 40 years of age is usually required to detect coronary vascular lesions — coronary artery disease [33].

At the initial stage of diagnostic search, of course, the doctor should pay attention to the symptom complex, which is presented in Table 3.

E.A. Shry et al. (2001) state that during auscultation the physician will hear auscultatory signs characteristic of mitral and tricuspid insufficiency [36, 37]. Electrocardiogram (ECG) also does not provide a clear answer to the question: is there a myxoma or not? When studying ECG films, a number of nonspecific changes are determined, various arrhythmias and cardiac conduction disturbances may be observed. The leading method in diagnostics of heart tumours is echocardiography (Table 4) [11].

When performing transthoracic echocardiography in patients with suspected volumetric cavity formation — myxoma, it is necessary to differentiate it from a large atrial thrombus. Compared to myxoma, the structure of thrombus is quite homogeneous, and the source is most often the atrial auricle, if we talk about the left atrium [11].

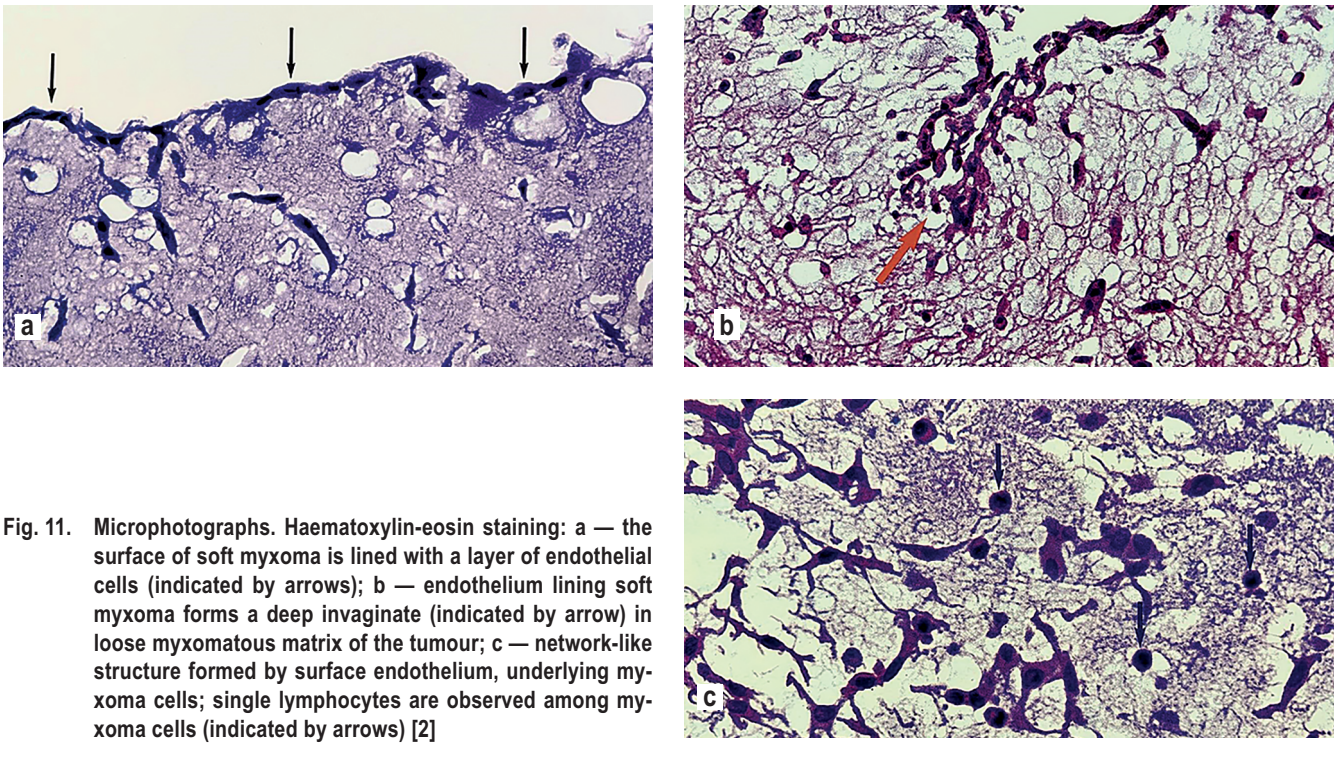

Fig. 11. Microphotographs. Haematoxylin-eosin staining: a — the surface of soft myxoma is lined with a layer of endothelial cells (indicated by arrows); b — endothelium lining soft myxoma forms a deep invaginate (indicated by arrow) in loose myxomatous matrix of the tumour; c — network-like structure formed by surface endothelium, underlying myxoma cells; single lymphocytes are observed among myxoma cells (indicated by arrows) [2]

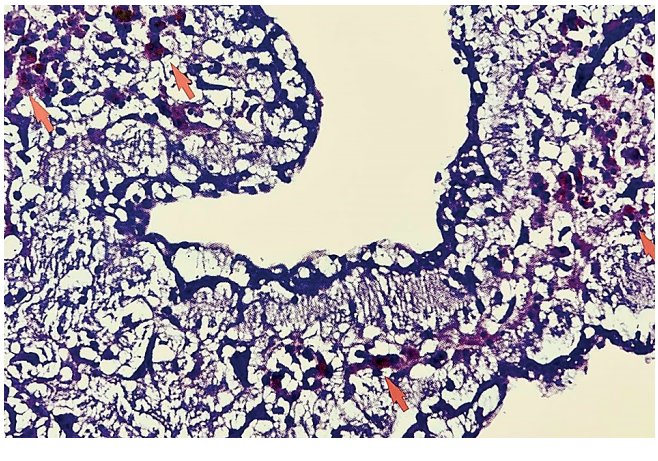

Fig. 12. A villous myxoma with small haemorrhages. Aggregation of siderophages in the villi (indicated by arrows). Microphotograph. Haematoxylin-eosin staining [2]

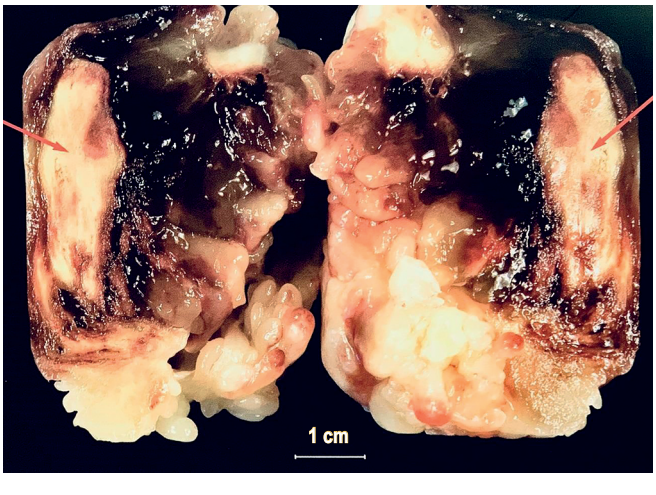

Fig. 13. Tissue necrosis of soft villous myxoma (indicated by arrows) on the cut surface. Macrophotography [2]

3D-echocardiography is of great importance in the diagnosis of cardiac masses. The increasing availability of 3D echocardiography (3DE) over the last decade has increased knowledge of the structure and function of cardiac haemodynamics [21]. The advantages of 3D-echocardiography include the elimination of geometric assumptions, quantification of volumes of complex geometries, and viewing structures from any point, all of which were not possible with traditional 2D-echocardiography (Fig. 14) [18, 19, 40].

A remarkable example of the use of 3D-echocardiography and its high significance in the diagnosis of cardiac tumour diseases, including atypical localisation, was demonstrated by M. Cottini et al. (2016) in a clinical observation of a rare case of left atrial myxoma localisation on its posterior wall (Fig. 15).

SURGICAL TREATMENT METHODS

Currently, the largest experience has been accumulated by cardiac surgeons of all countries in the surgical treatment of cardiac myxomas. The surgical tactics in establishing the diagnosis of cardiac myxoma is to perform surgical treatment in a short period of time as a matter of urgency [24]. Myxoma removal is the only radical and, from the point of view of the treatment, correct method.

The tumour is approached through an incision in the right atrium (there is also access to the left atrium through its roof) (Fig. 15, *a*). The interatrial septum (IS) is opened in the region of the fossa ovalis. A flap of septal tissue free of tumour tissue is created. The tumour is detached until the tumour pedicle is clearly defined (Fig. 15, *b*). The point of attachment is often located on the IS, allowing complete removal of the tumour along with a significant amount of normal IS tissue. If the tumour attaches to the left atrial wall or mitral valve, the amount of normal tissue resection must be determined on a case-by-case basis.

Small retractors are used to dilate the opening in the IS (Fig. 15, *c*) -— this makes it possible to extract the tumour from the left atrium without damaging it. Some patience is required to extract the entire tumour without damage. The tumour can be divided into parts (e.g. in case of giant size), but this is undesirable; it is preferable to take out the whole tumour. After completion of the main stage of the operation, the IS defect is sutured (Fig. 15, *d*), including the use of

Table 3

Symptom-complex diagnostic search in suspected cardiac myxoma [4]

Symptoms		
Nonspecific:	By localisation:	Signs of valve pathology:
• weight loss; • fever; • weakness; • fatigue; • syncopal attacks	• symptoms of systemic embolism; • symptoms of pulmonary embolism; • sudden cardiac death	• mitral stenosis: signs of left ventricular insufficiency — dyspnoea, cough, haemoptysis, pulmonary congestion, symptoms of peripheral inflammation, chest pain; symptoms of IC obstruction, left ventricular outflow tract (signs of pulmonary hypertension, hydrothorax — in case of large myxomas); • tricuspidal stenosis: signs of right ventricular insufficiency — ascites, hepatomegaly

Table 4

Echocardiographic picture of myxoma in a 'classic' case [4, 11]

B-mode	M-mode	Doppler	Further actions	The main
Myxomas are visualised often as mobile structures located near the fossa ovalis of the interatrial septum, they may pendulum between the atria and ventricle	Multiple parallel echoes, thickening of EF slope; normal DE-amplitude	No colour flow signal in the tumour area	Transesophageal and/or 3D echocardiography	Detection of myxoma, clarification of its localisation, determination of size, degree of obstruction, valve insufficiency, the size of affected cavities
To clarify the localisation and determine the size of the tumour	In myxomas, cavities may be enlarged	Mitral valve obstruction, tricuspid valve obstruction, and left ventricular outflow tract or right ventricular outflow tract obstruction are possible	Emergency surgical intervention on an urgent basis	M-mode gives results as in valve stenosis
The size and function of the involved cardiac chambers should be assessed	–	Assessment of the severity of valve insufficiency	–	If a myxoma is detected, surgery is urgent
NB! After radical correction of the pathology, regular echocardiographic monitoring is required due to the high rate of tumour recurrence				

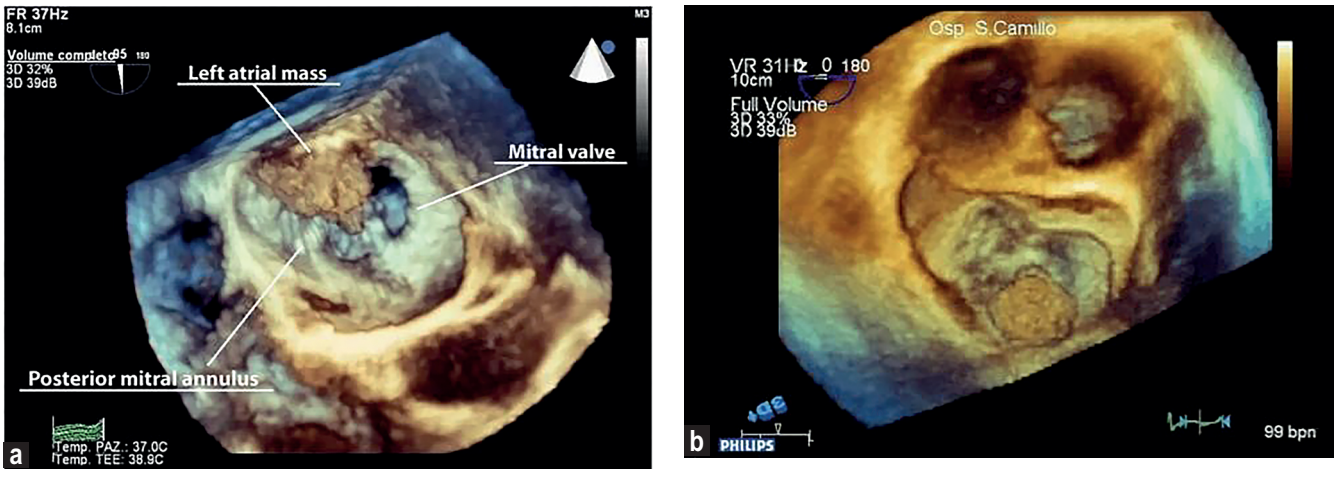

Fig. 14. Real-time three-dimensional (3D) echocardiography using a surgical view of the left atrium. The myxoma was demonstrated to be located near the posterior mitral annulus with a diameter of 2.5 × 2 cm [20]

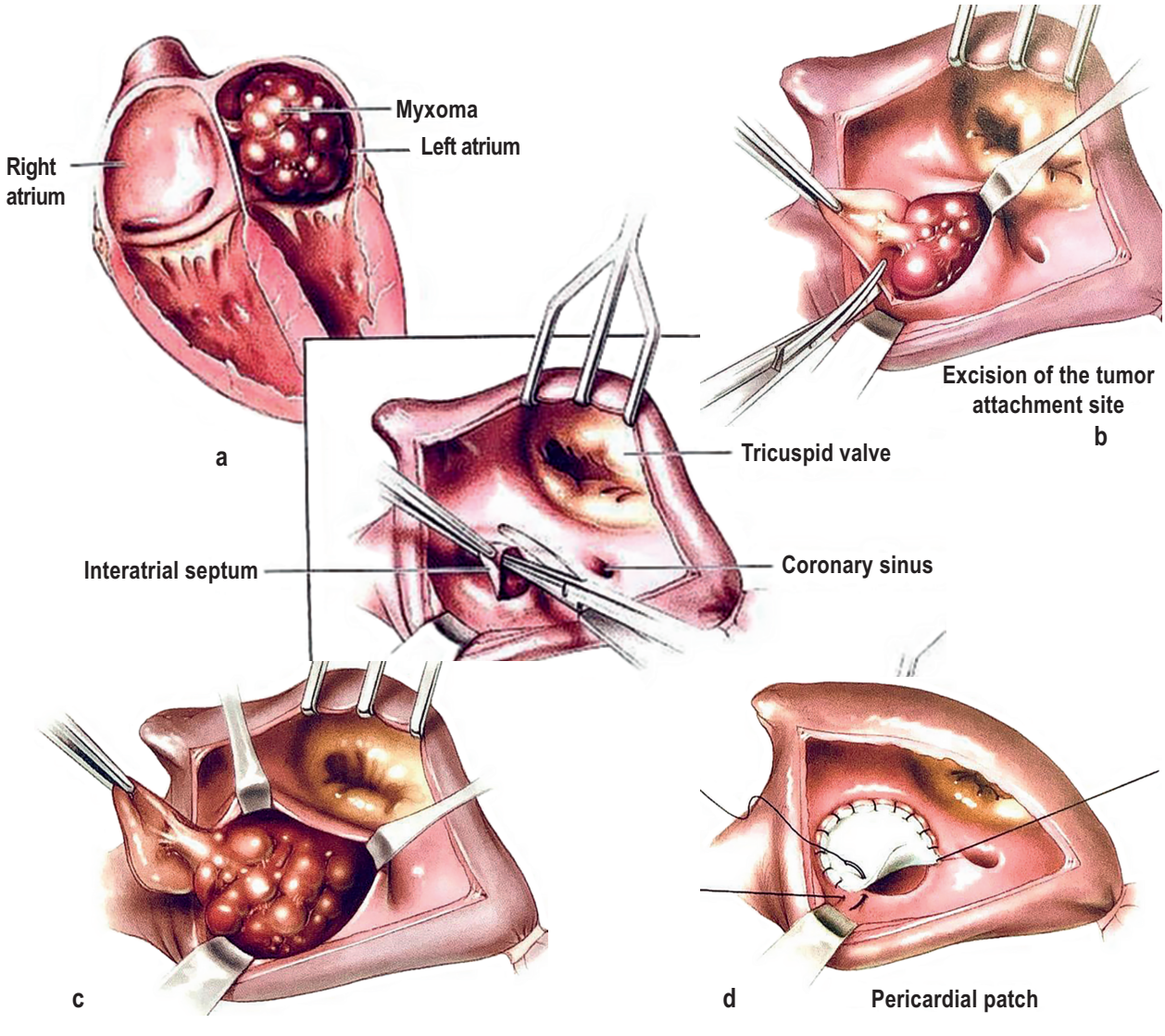

Fig. 15. Stages of surgical treatment of left atrial myxoma (a–d). An explanation is in the text [6]

Table 5

Indications and contraindications for surgery developed by the A.N. Bakulev National Centre of Cardiovascular Surgery

Indications	Contraindications
1. The initial condition of the patient. 2. The risk of thromboembolic complications with tumour fragments. 3. The probability of obstruction of heart valves. 4. Risk of surgical intervention (EuroScore II). 5. Metastasis to other organs. 6. Degree of involvement of cardiac structures. 7. The age of the patients. 8. The presence of concomitant diseases	1. Severe CNS damage due to frequent recurrent material emboli, most commonly seen in patients with large "jelly-like" left atrial myxomas. 2. Extensive left ventricular myocardial infarct zone with cardiac rhythm disturbances. 3. Postinfarction left ventricular myocardial aneurysm with severe coronary lesions. 4. Severe incurable concomitant congenital heart disease in children
Cardiomegaly, high hypertension of the small circulatory circle, previous cardiac surgery are high-risk factors for surgery. They are not contraindications.	

Note: CNS — central nervous system

pericardial patch by suturing it to the rim of the defect with "prolene 4/0" thread [6].

THE CLINICAL CASE OF THE LEFT ATRIAL VILLOUS MYXOMA

Patient L., 80 years old, was admitted with complaints of dyspnoea, pressure pain with minor physical activity.

She was routinely hospitalised for additional examination, correction of therapy and determination of further treatment tactics 08.11.2019 at the cardiology department of the State Budgetary Institution "Leningrad Regional Clinical Hospital" (LRCB).

Anamnesis morbi: there is a long history of hypertension, maximum figures of blood pressure (BP) — 220/100 mm Hg, adapted to 120-130/70 mm Hg, constant therapy was not received. For a year, dyspnoea and high BP values have been bothering the patient. Progressive course of the disease. She was observed by a cardiologist at the place of residence. On 09.10.2019 she was admitted to the hospital at the place of residence, where according to EchoCG in the position of the anterior leaflet of the mitral valve, a volumetric formation up to 20 mm in diameter was detected, with periodic obturation of the outlet from the LP and the formation of severe mitral stenosis.

Anamnesis vitae. 1. Operations, including outpatient: cholecystectomy, appendectomy, uterine extirpation in 1990. 2. Gynaecological anamnesis: pregnancies 7; deliveries 2, abortions 5. 3. Allergological anamnesis is not aggravated. 4. Epidemiological anamnesis is not aggravated; 5. No blood transfusions in the last 6 months.

Physical examination: the condition is stable, of average severity. Consciousness is clear, contact. The skin is pale pink, without pathological rashes. Peripheral lymph nodes are not enlarged. Pulse — 90 beats per minute, rhythmic. BP — 140/80 mm Hg on both sides. Heart tones are muffled, rhythmic. First tone amplification at the apex of the heart and diastolic murmur in the projection of the mitral valve are heard. In the lungs breathing is rigid, D = S, without wheezing. The tongue is moist, clean. The abdomen is not swollen, it participates symmetrically in respiration. On palpation soft, painless. Peritoneal symptoms are negative. The liver is palpatorily unchanged. The spleen is not palpated. Kidneys are not palpated. Urination is said to be sufficient, without peculiarities. Stool is normal. Diuresis is normal.

Results of examinations confirming the diagnosis:

Transthoracic EchoCG dated 06.11.2019: hyperechogenic mass of large size in the left atrium, more likely myxoma, with mitral valve obturation and formation of critical mitral stenosis, with a significant risk of embolism into the large circulation circle. Sharp dilatation of the left atrium, significant dilatation of the right atrium, moderate dilatation of the right ventricle. First degree pulmonary hypertension. Moderate concentric hypertrophy of the left ventricle.

ECG from 06.11.2019: Sinus rhythm with the number of heart contractions — 90 bpm. Violations of intra-atrial conduction. Probable enlargement of the right ventricle. Nonspecific repolarisation changes.

On the basis of the presented data the clinical diagnosis was made: D15.1 Benign neoplasm: Heart. Left atrium formation with formation of critical mitral stenosis — indications for urgent surgical treatment were determined.

Preoperative transesophageal echoCG was performed (Fig. 16) — the diagnosis was confirmed.

Surgical intervention was performed (operating team D.G. Gramatikov, V.K. Noginov, L.Yu. Artyukh): median sternotomy was performed under endotracheal anaesthesia. Pericardotomy. At revision — cardiomegaly. An artificial circulatory apparatus (ACA) was connected according to the scheme (aorta-vena cava-left ventricle). An extracorporeal blood circulation. Thermal blood non-selective cardioplegia antegradely into the aortic root. Access to the left atrium through the right atrium and interatrial septum (Fig. 17).

At revision, a 4.0 × 5.0 cm voluminous gelatinous mass on a pedicle was found in the left atrial cavity, which was completely

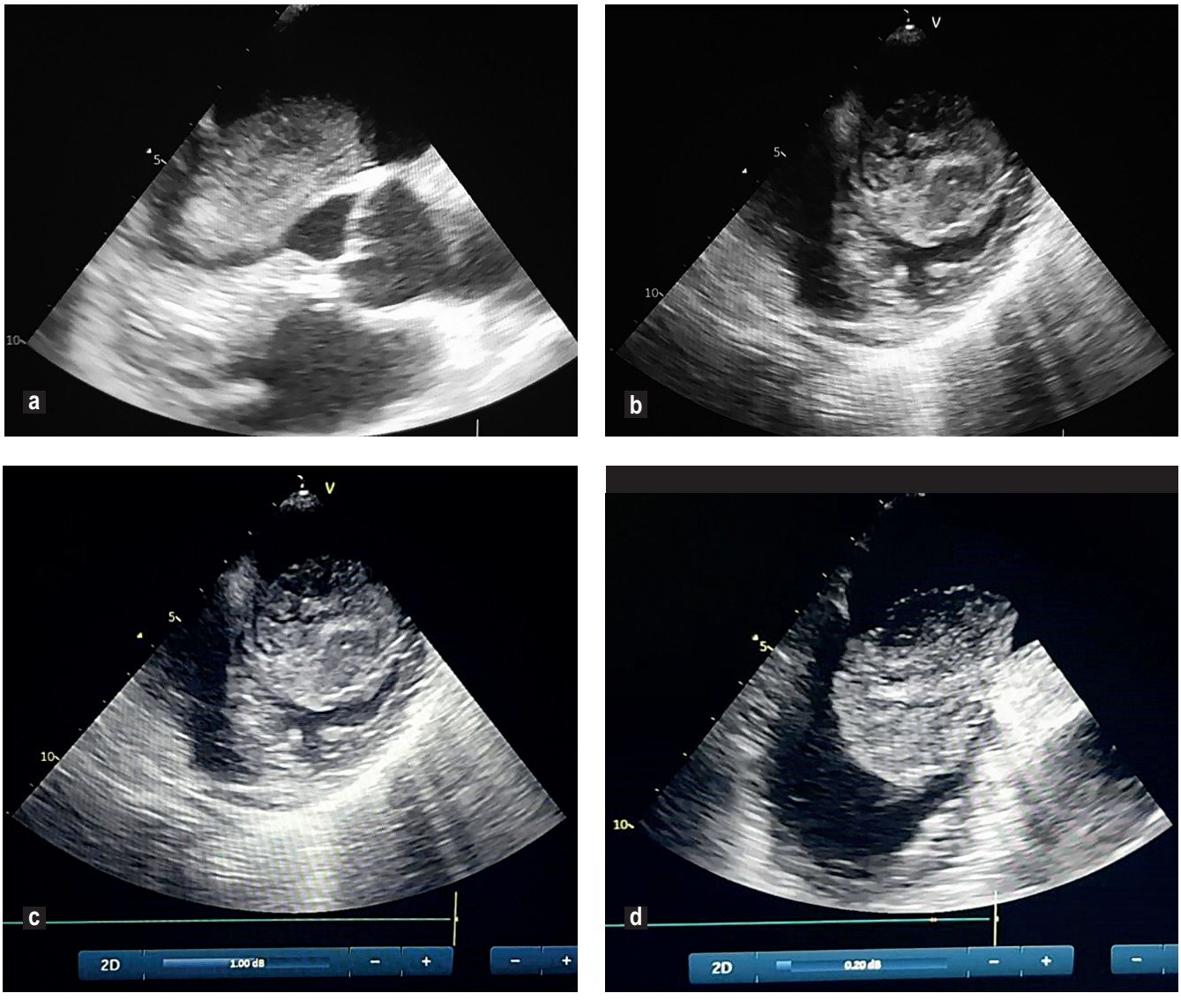

Fig. 16. Transesophageal EchoCG. In the left atrium, a roundish mass with a diameter of 4 × 5 cm, of inhomogeneous structure, with irregular fragments along the edges, is visualised. The mass creating mitral valve stenosis is attached to the MV ring on the side of the anterior leaflet (a rare variant of localisation). Mitral insufficiency of the first degree. (Observation (preoperative) of the Leningrad Regional Clinical Hospital, 2020)

evacuated from the left atrial cavity (Fig. 18). Suture of the interatrial septum, double-row suture of the right atrium. The aortic clamp was removed. Prevention of air embolism. Restoration of cardiac activity. Transesophageal EchoCG — the neoplasm fragments were not visualised (Fig. 19).

Hemodynamics was stabilised. The artificial circulatory apparatus was stopped and disconnected. Metal osteosynthesis of the sternum. Layer suture of the postoperative wound. Transfer to the department of anaesthesiology, resuscitation and intensive care. The early postoperative period is smooth. Postoperative sutures heal by primary tension, the sternum is stable. On 15.11.2019, the patient was transferred to the cardiology department of the interdistrict hospital for further treatment and rehabilitation. At the time of discharge, the condition is stable. Haemodynamic parameters are within normal values.

Histological examination data: a mass represented by loose edematous fibrous tissue with hemorrhagic impregnation and a cluster of spindle-shaped cells with indistinct contours — villous myxoma.

CONCLUSION

Although histologically benign, myxomas are potentially dangerous because of the high risk of systemic and cerebral embolism, so surgical intervention preceded by a thorough diagnostic search should be planned for these patients immediately and also performed immediately.

ADDITIONAL INFORMATION

Author contribution. L.Yu. Artyukh — development of the design of the review, writing the text of the article, general editing, literary search; N.R. Karelina, D.G. Gramatikov, I.N. Sokolova, M.Yu. Erina — review design editing, general editing, literary search; V.K. Noginov, V.V. Sklyarova, A.A. Prokhorycheva, M.R. Gafiatulin — literary search. The final version has been read and approved by all authors.

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Consent for publication. Written consent was obtained from the patient for publication of relevant medical information within the manuscript.

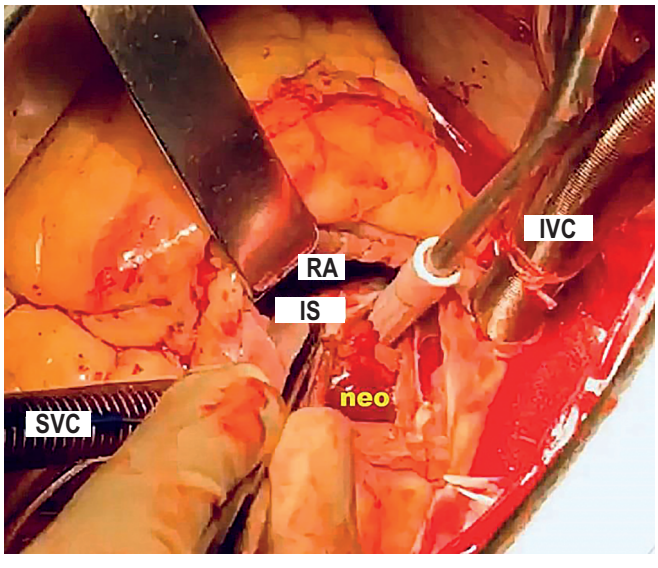


Fig. 17. **Intraoperative photograph. The artificial circulation apparatus was connected according to the scheme (aorta-vena cava-left ventricle). Extracorporeal circulation and thermal blood non-selective cardioplegia. Access to the left atrium through the right atrium and interatrial septum. A mass of jelly-like consistency on a pedicle is visualised under the interatrial septum. SVC — cannula placed in the superior vena cava; IVC — cannula placed in the inferior vena cava; IS — interatrial septum; RA — right atrium; neo — formation in the left atrial cavity**

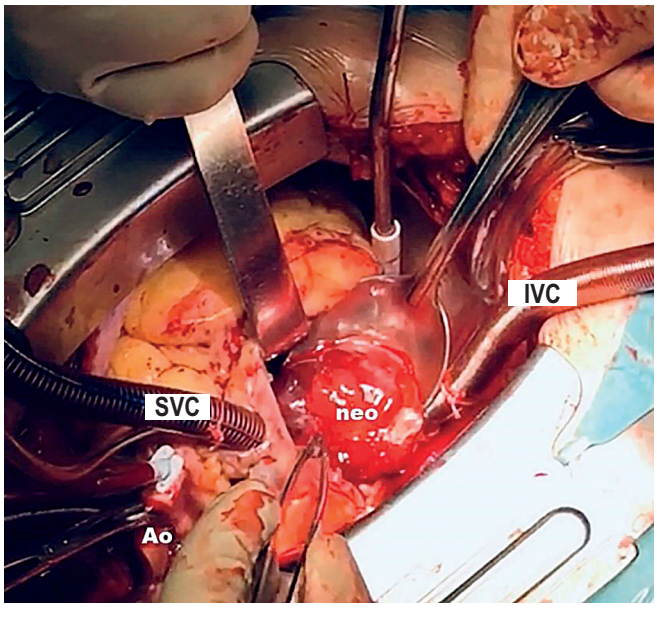
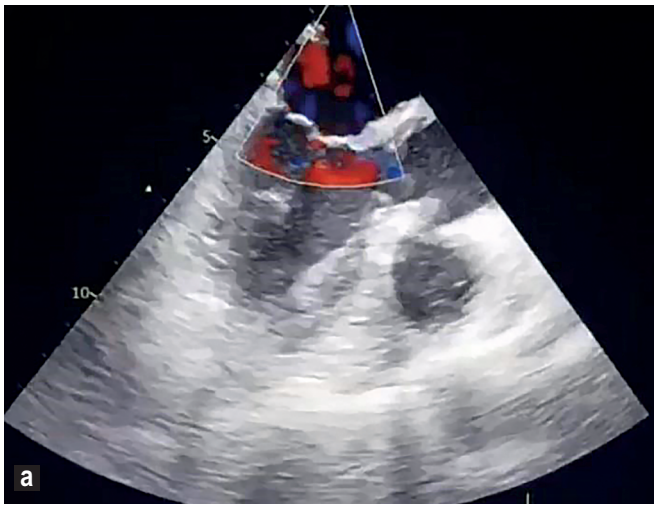


Fig. 18. **Stenotic mass of the left atrium. Intraoperative photograph. SVC — cannula placed in the superior vena cava; IVC — cannula placed in the inferior vena cava; neo — formation in the left atrial cavity; Ao — ascending aorta, constricted**



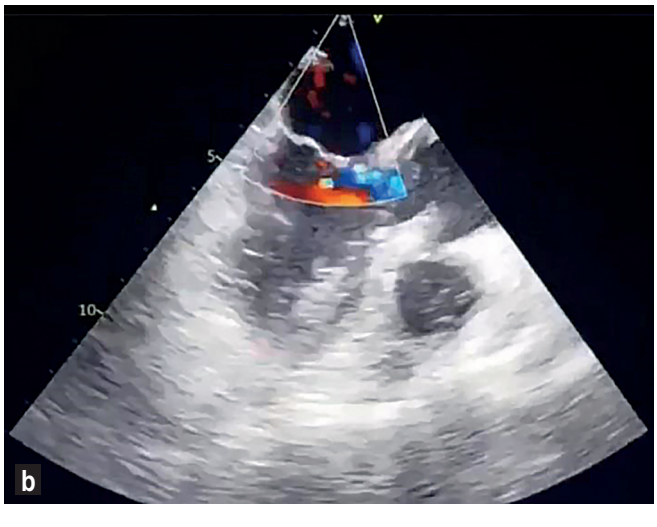


Fig. 19. **Transesophageal echocardiography. Postoperative data**

ДОПОЛНИТЕЛЬНАЯ ИНФОРМАЦИЯ

Вклад авторов. Л.Ю. Артюх — разработка дизайна обзора, написание текста статьи, общее редактирование, литературный поиск; Н.Р. Карелина, Д.Г. Грамматиков, И.Н. Соколова, М.Ю. Ерина — редактирование дизайна обзора, общее редактирование, литературный поиск; В.К. Ногинов, В.В. Склярова, А.А. Прохорычева, М.Р. Гафиатулин — литературный поиск. Окончательная версия прочитана и одобрена всеми авторами.

Конфликт интересов. Авторы декларируют отсутствие явных и потенциальных конфликтов интересов, связанных с публикацией настоящей статьи.

Источник финансирования. Авторы заявляют об отсутствии внешнего финансирования при проведении исследования.

Информированное согласие на публикацию. Авторы получили письменное согласие пациентов на публикацию медицинских данных.

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