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# ФИБРИЛЛЯЦИЯ ПРЕДСЕРДИЙ И СЕРДЕЧНАЯ НЕДОСТАТОЧНОСТЬ В ДЕБЮТЕ AL-АМИЛОИДОЗА

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# Atrial Fibrillation and Heart Failure as the Onset of AL-Amyloidosis

**Резюме**

В практике кардиолога нередко встречаются пациенты с гипертрофией миокарда, фибрилляцией предсердий и сердечной недостаточностью. Выяснение причин этих состояний крайне важно для назначения этиологической терапии, улучшающей прогноз. В статье представлен клинический случай несвоевременно диагностированного амилоидоза у мужчины 53 лет. Несмотря на комплексную терапию, течение заболевания осложнилось развитием двусторонней пневмонии, сепсиса, синдрома диссеминированного внутрисосудистого свертывания, что привело к летальному исходу. На аутопсии подтвержден диагноз AL-системного амилоидоза (тип каппа) с массивным поражением сердца, почек, легких, печени, селезёнки, надпочечников, щитовидной железы, поджелудочной железы, желудочно-кишечного тракта, подкожной жировой клетчатки и артериальных сосудов костного мозга. Для прижизненной диагностики AL-амилоидоза и своевременного назначения патогенетической терапии необходимо проведение скрининга при выявлении гипертрофии левого желудочка, фибрилляции предсердий и сердечной недостаточности неясной этиологии.

***Ключевые слова:*** *амилоидоз сердца, амилоидная кардиомиопатия, хроническая сердечная недостаточность с сохраненной фракцией выброса левого желудочка, фибрилляция предсердий, гипертрофия левого желудочка, хроническая болезнь почек, протеинурия, нефротический синдром, кардиоренальный синдром*

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**Конфликт интересов**

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

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**Abstract**

Left ventricular hypertrophy, atrial fibrillation and chronic heart failure are often in the practice of a cardiologist. The etiology of these conditions is very important because the correct early treatment. We are presenting a case of a late diagnosis of amyloidosis in a 53-year-old man. Despite the complex therapy, the course of the disease was complicated by the development of bilateral pneumonia, sepsis, disseminated intravascular coagulation and the patient died. Autopsy confirmed the diagnosis of systemic AL-amyloidosis (type Kappa) with massive damage to the heart, kidneys, lungs, liver, spleen, adrenal glands, thyroid gland, pancreas, gastrointestinal tract, subcutaneous fatty tissue and arterial vessels of the bone marrow. Thus, screening for amyloidosis is necessary in idiopathic LV thickening, atrial fibrillation, and heart failure for timely intravital diagnosis and therapy.

***Key words:*** *cardiac amyloidosis, amyloid cardiomyopathy, chronic heart failure with preserved ejection fraction, atrial fibrillation, left ventricular hypertrophy, chronic kidney disease, proteinuria, nephrotic syndrome, cardiorenal syndrome*

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**Conflict of interests**

The authors declare no conflict of interests

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BP — arterial pressure, LV — left ventricle, GFR — glomerular filtration rate, AF — atrial fibrillation, ECG — electrocardiogram, EchoCG — echocardiography

## Introduction

AL amyloidosis is the most aggressive form of systemic amyloidosis, which affects the heart in 60% of patients and kidneys in 74% of patients [1]. The median survival of patients with cardiac diseases, if untreated, is no more than one year, and after the onset of heart failure symptoms — nine months [2, 3]. The first signs of AL amyloidosis may be atrial fibrillation (AF) and heart failure, which are the reason for seeking the cardiologist's advice. It is important for practitioners to be vigilant and prescribe the necessary examinations for patients with myocardial hypertrophy, AF and heart failure of unknown etiology for the early diagnosis of amyloidosis and timely prescription of therapy to improve the prognosis of the disease. The presented clinical case demonstrates the debut of systemic amyloidosis with the development of AF and heart failure in a middle-aged patient.

## Clinical case description

Male, 53 years old, without history of cardiovascular, thyroid and other diagnosed disease, or toxic exposure, presented with dyspnea on exertion. A month later, the electrocardiogram (ECG) recorded AF, tachysystole, low QRS complexes voltage in the precordial leads, signs of right ventricular hypertrophy with qR-type QRS complex, the absence of an adequate R-wave elevation in V3–V6 (Fig. 1).

The outpatient echocardiography (EchoCG) showed a 12 mm thickening of the walls of the left ventricle (LV) myocardium, 43 mm dilatation of the left atrium, with preserved LV ejection fraction — 59% based on the Simpson method. Attempts to restore sinus rhythm with amiodarone and electrical cardioversion had short-term results. However, AF recurrence occurred in 12 hours. Radiofrequency ablation was planned but postponed after finding thrombosis in the left atrial appendage by means of transesophageal echocardiography. The recommended drugs (vitamin K antagonist (warfarin 3.75–5 mg per day with INR monitoring), then rivaroxaban 20 mg/day; metoprolol succinate 25 mg/day, then bisoprolol 5 mg/day; perindopril 2.5 mg/day; torasemide 10 mg/day; spironolactone 25 mg/day) were taken regularly.

Five months after the onset of the disease, multislice computed tomography of the chest with contrast enhancement showed hyperuricemia 768 μmol/l, increased serum creatinine 155 μmol/l, a decrease in the calculated glomerular filtration rate (eGFR, CKD-EPI) 43.4 ml/min/1.73 $m^2$ (initial values of creatinine and eGFR are unknown, examined by a nephrologist, contrast-induced kidney damage is assumed, nephroprotective therapy is recommended).

Eight months after the onset of dyspnea for a week, there were syncopes with tongue biting, loss of bowel control, an episode of gross hematuria developed, which led to hospitalization to the V.M. Buianov State Clinical Hospital on October 24, 2018.

On admission, the condition was severe; the patient presented with the pallor of the skin, edema of the lower extremities, weakened breathing in the lower parts of both lungs, no wheezing, respiratory rate 23 per minute, blood saturation ($SpO_2$) 98%, irregular heart rate 90 beats per minute, blood pressure (BP) 116/68 mm Hg. Laboratory tests showed a decrease in hemoglobin to 104 g/l, hypoproteinemia — 55 g/l, hypoalbuminemia — 26 g/l, proteinuria — 10 g/l in a single portion of urine, increase in creatinine to 686 μmol/l, urea to 40.5 mmol/l, decrease in eGFR to 7.2 ml/min/1.73 $m^2$, aspartate aminotransferase — 58 IU/l (5–34), alanine aminotransferase — 45 IU/l (0–32), total creatine phosphokinase — 90 IU/l (21–215), MV fraction of creatine phosphokinase — 12 IU/l (0–25), total lactate dehydrogenase — 471 IU/l (225–450), gamma glutamyl transpeptidase — 1295 IU/l (9–39), alkaline phosphatase — 1598 IU/l (64–306), alpha-amylase — 392 IU/l (0–220), total bilirubin — 42.4 μmol/l (1.7–20.5), direct bilirubin — 35 μmol/l (0.86–5.00), troponin I — 0.120 μg/l (0.0–0.1, in real-time — an increase to 1.020 μg/l), antithrombin III — 67.8% of N (80.0–120.0), D-dimer — 324 ng/ml (64–550), N-terminal precursor of brain natriuretic peptide > 35 000 ng/l (Table 1).

Abdominal ultrasound showed a moderate increase in the size of both kidneys (left: 132 x 69 x 60 mm, volume 272 $cm^3$; right: 137 x 61 x 59 mm, volume 261 $cm^3$), hepatomegaly (left lobe oblique caudal size — 126 mm, thickness — 99 mm, right lobe oblique vertical dimension — 210 mm, thickness — 143 mm), splenomegaly (150 × 73 mm), a small amount of fluid in the abdominal cavity. Computed tomography of the brain showed no abnormality. EchoCG showed a thickening of the LV myocardium walls up to 22 mm with normal LV end-diastolic size (38 mm) and LV end-diastolic volume (60 ml), restrictive type of diastolic dysfunction with preserved LV ejection fraction (55%), left atrial size — 43 mm, left atrial volume — 98 ml end-diastolic right ventricular size — 42 mm, up to 7 mm pericardial separation (Table 2, Fig. 2).

Multislice computed tomography revealed bilateral hydrothorax, hydropericardium, signs of pulmonary congestion, ascites, enlargement of the liver, spleen and aortocaval lymph nodes up to 12 mm (Fig. 3).

Signs of hypertrophic cardiomyopathy in combination with nephrotic syndrome (proteinuria up to 10 g/l, daily proteinuria not assessed, hypoproteinemia 55 g/l, hypoalbuminemia 26 g/l, hypooncotic edema, hyperlipidemia) and decreased renal function allowed to suspect systemic amyloidosis in the patient. The hereditary (mutant) variant of transthyretin (ATTR) amyloidosis was excluded by direct sequencing of the entire coding sequence and regions of exon-intron junctions of the transthyretin gene, in which pathogenic and probably pathogenic variants of the nucleotide sequence were not detected in this gene. Immunochemical assay of 24-hour urine revealed an increase in the excretion of light chains of immunoglobulins — kappa up to 63.9 mg/l (norm < 7.31 mg/l), lambda up to 10 mg/l (norm < 4.03 mg/l). Bone marrow biopsy showed low plasma cell count (8%), which allowed to exclude multiple myeloma (Table 3).

Isolated amyloid deposits were found during aspiration biopsy of subcutaneous fat with Congo red staining and examination in polarized light (Fig. 4). Similar deposits were found in the walls of arterioles, the muscle lamina of the mucous membrane in the lamina propria of the colon mucosa, and the walls of the arterial vessels of the bone marrow trephine biopsy. When examined in polarized light, the birefringence with an apple-green and yellowish glow was demonstrated. Pachler's test with potassium permanganate excluded the AA-type amyloid.

After six sessions of veno-venous hemodiafiltration (VVHDF), a decrease in the level of serum creatinine to 169 μmol/l and urea to 6.2 mmol/l was observed. The course of the disease was complicated by bilateral pneumonia, sepsis (with an increase in procalcitonin up to 200 ng/ml, C-reactive protein up to 101 mg/l), disseminated intravascular coagulation syndrome (with a decrease in antithrombin III to 12.3% of N, an increase in D-dimer up to 21 400 ng/ml, a decrease in blood hemoglobin levels to 60 g/l, platelets up to 54 × $10^9$/l). Transfusion of erythrocyte mass, fresh frozen plasma and antithrombin III was performed.

On the 26th day of hospitalization, with ultrasound Doppler in duplex mode of the veins of lower extremities, occlusive thrombosis of the sural veins of the left lower extremity was visualized.

Despite the antibiotic, infusion therapy, diuretic, antiarrhythmic, gastroprotective, and hepatoprotective therapy, the patient died on the 27th day of hospitalization.

Based on clinical, laboratory and other diagnostic test data, a clinical diagnosis was:

**Principal diagnosis:** Systemic AL amyloidosis with involvement of the heart (grade III amyloid cardiomyopathy), liver, spleen, gastrointestinal tract, kidney, subcutaneous fat.

**Complications:** Persistent AF of unknown duration. CHA2DS2-Vasc 2 points/ HAS-BLED 3 points. Chronic heart failure IIB stage, III FC. Nephrotic syndrome. Chronic kidney disease stage 5 (glomerular filtration rate according to the CKD-EPI formula 12 ml/min/1.73 $m^2$), A4. Nosocomial bilateral polysegmental pneumonia. Respiratory failure degree 2. Sepsis. Disseminated intravascular coagulation syndrome. Severe thrombocytopenia. Coagulopathy. Recurrent nosebleeds. Severe normochromic normocytic anemia. Occlusive thrombosis of the sural veins of the left lower extremity on November 20, 2018. Pulmonary embolism on November 21, 2019. Generalized convulsive seizure on October 24, 2018.

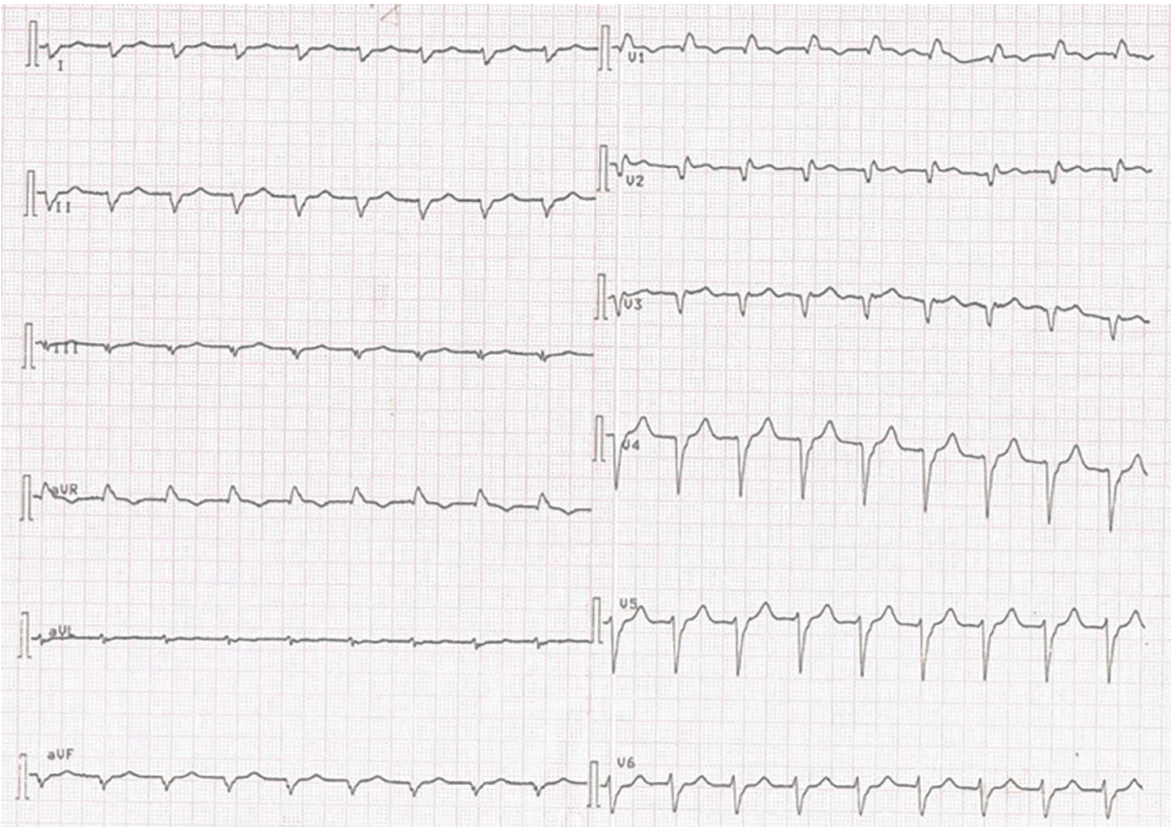


***Figure 1.*** *Electrocardiogram. Atrial fibrillation with ventricular rate 115 per minute, decreasing of r voltage in I, II, III, avL, avF, V3-V6, right ventricular hypertrophy qR type*

***Table 1.*** *Results of laboratory examinations in dynamics*

| Parameter | At the onset of the disease | In 4 months | In 8 months | Normal value |
|---|---|---|---|---|
| **Blood analisis** | | | | |
| Hemoglobin, g/l | 165 | 111 | 104 | 130-170 |
| Red blood cells, $10^{12}$/l | 5,29 | 3,8 | 3,46 | 4,28-5,78 |
| Hematocrit, % | 47,6 | 35,5 | 31 | 39,5-51,0 |
| The volume of erythrocytes, fl | 90 | 94 | 89,6 | 82-98 |
| Red blood cell hemoglobin content, pg | 31,2 | 29,4 | 30,1 | 27,9-33,2 |
| Platelets, $10^9$/l | 292 | 160 | 291 | 150-340 |
| White blood cells, $10^9$/l | 13,25 | 14,8 | 9,3 | 3,9-10,9 |
| **Blood chemistry** | | | | |
| Total protein, g/l | **67** | **61** | **55** | **65-85** |
| Albumin, g/l | | | 26 | 35-55 |
| Creatinine, μmol/l | 155 | 231 | **686** | 71-115 |
| Urea, mmol/l | 11,4 | 16,5 | 40,5 | 2,5-8,3 |
| Potassium, mmol/l | 4,8 | 4,8 | 4,9 | 3,5-5,5 |
| Sodium, mmol/l | 142 | | 138 | 135-150 |
| **Urine analysis** | | | | |
| Blood | Not | + | + | Not |
| Protein, g/l | Not | 1 | 10 | Not |
| Glucosae, μmol / l | Not | Not | Not | Not |
| Specific gravity, g/l | 1001 | 1004 | 1016 | 1005-1030 |
| Leukocytes, per 1 μl | 0-1 | 17 | 500 | 0 |
| pH | 6 | 6 | 6 | 5-7 |

***Table 2.*** *Dynamics of echocardiographic parameters*

| Parameter | At the onset of the disease | In 6 months | In 8 months | Normal value |
|---|---|---|---|---|
| Interventricular septum thickness, mm | 11 | 15 | 21 | Up to 10 |
| Left ventricular posterior wall thickness, mm | 12 | 14 | 22 | Up to 10 |
| Left ventricular ejection fraction, % | 59 | 52 | 55 | >55 |
| End-diastolic volume of the left ventricle, ml | 98 | - | 60 | |

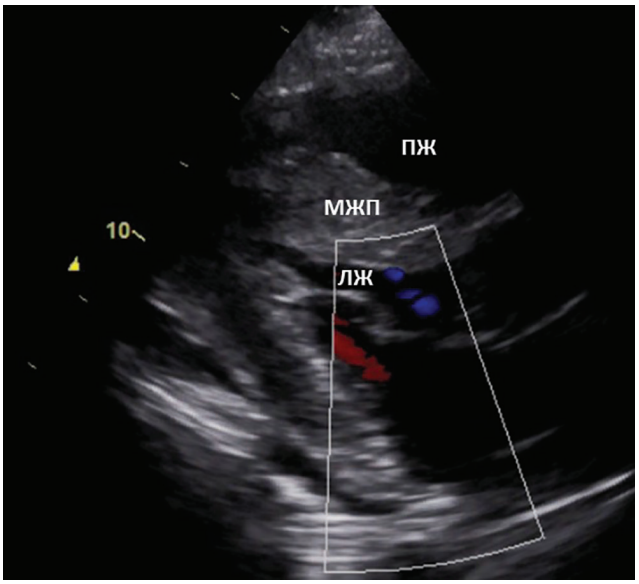


***Figure 2 A.*** *Echocardiogram (Photo of Dr. I.I. Ganieva). Parasternal long axis position (dyastola): left ventricular hypertrophy*
*ЛЖ — left ventricular, ПЖ — right ventricular, МЖП — interventricular septum*

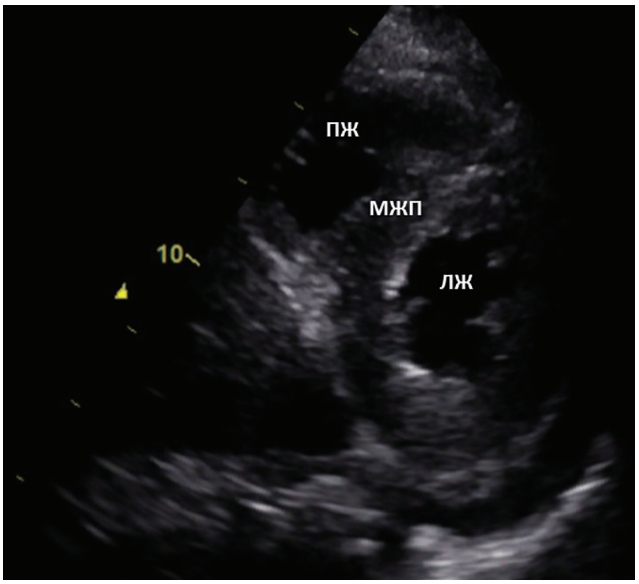


***Figure 2 B.*** *Echocardiogram (Photo of Dr. I.I. Ganieva). Parasternal position along the short axis of the left ventricle: severe left ventricular hypertrophy*
*ЛЖ — left ventricular, ПЖ — right ventricular, МЖП — interventricular septum*

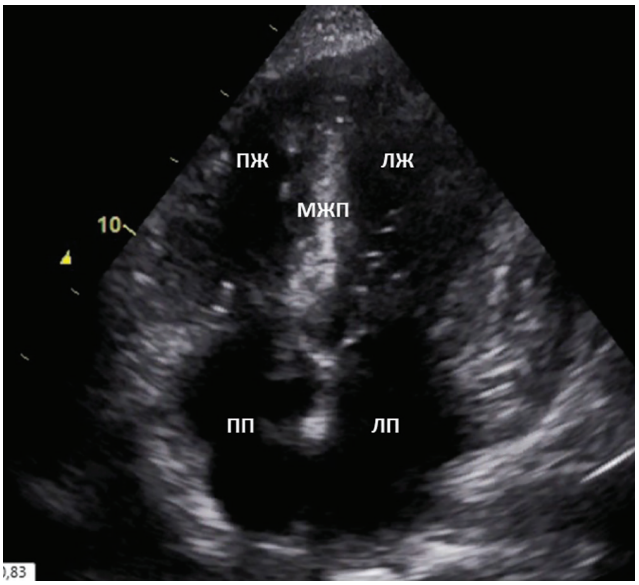


***Figure 2 C.*** *Echocardiogram (Photo of Dr. I.I. Ganieva). Apical four-chamber position: severe left ventricular hypertrophy*
*ЛЖ — left ventricular, ПЖ — right ventricular, МЖП — interventricular septum, ЛП — left atrium, ПП — right atrium*

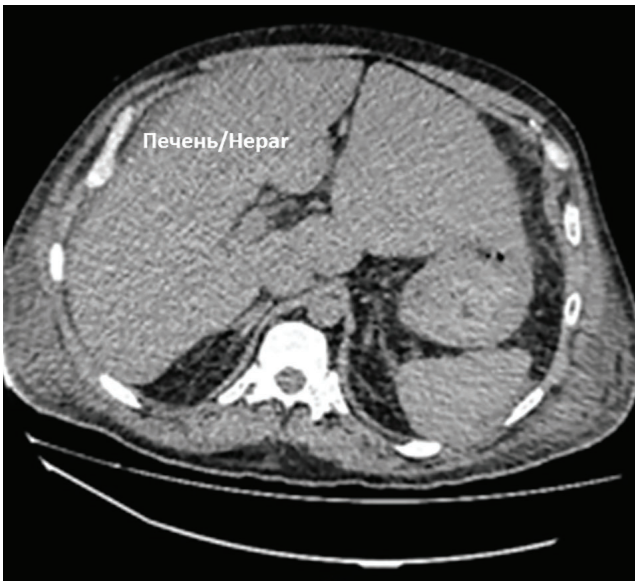


***Figure 3.*** *Computed tomography of the abdominal organs: hepatomegaly*

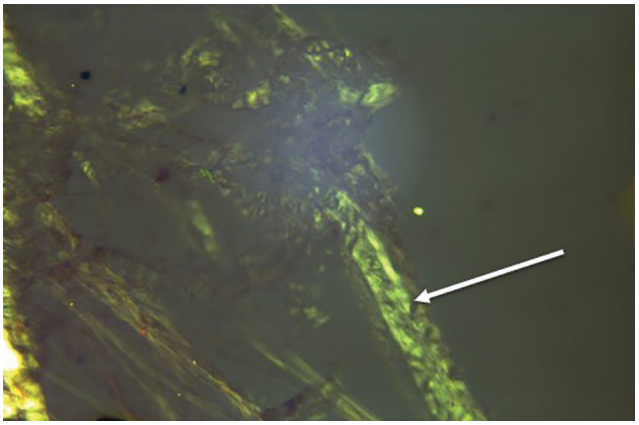

***Figure 4.*** *Biopsy of subcutaneous adipose tissue (Photo of Dr. EA Stepanova). Single irregularly spaced congophilic deposits, birefringence with apple-green glow (arrow). On a visual grage scale CR 1+, sometimes CR 2+*

**Concomitant disease:** Mixed encephalopathy.

**Invasive procedures:** Sessions of VVGDF of November 06, 2018; November 07, 2018; November 10, 2018; November 14, 2018; November 15, 2018; November 16, 2018.

During autopsy, the diagnosis of systemic amyloidosis (AL, IgG, κappa) was confirmed, a massive lesion of the heart (weight 885 g, normal — 310 g), kidneys (weight of the left kidney 291 g, right kidney 313 g, normal — 320 g), lungs (weight of the left lung 571 g, right lung 817 g, normal left lung — 325–480, right lung — 360–570 g), liver (weight 4120 g, normal — 1600 g), spleen (weight 320 g, normal — 150 g), adrenal glands, thyroid, pancreas, gastrointestinal tract, subcutaneous adipose tissue and arterial vessels of the bone marrow. Immunohistochemistry showed AL amyloidosis, type Kappa (Fig. 5–6).

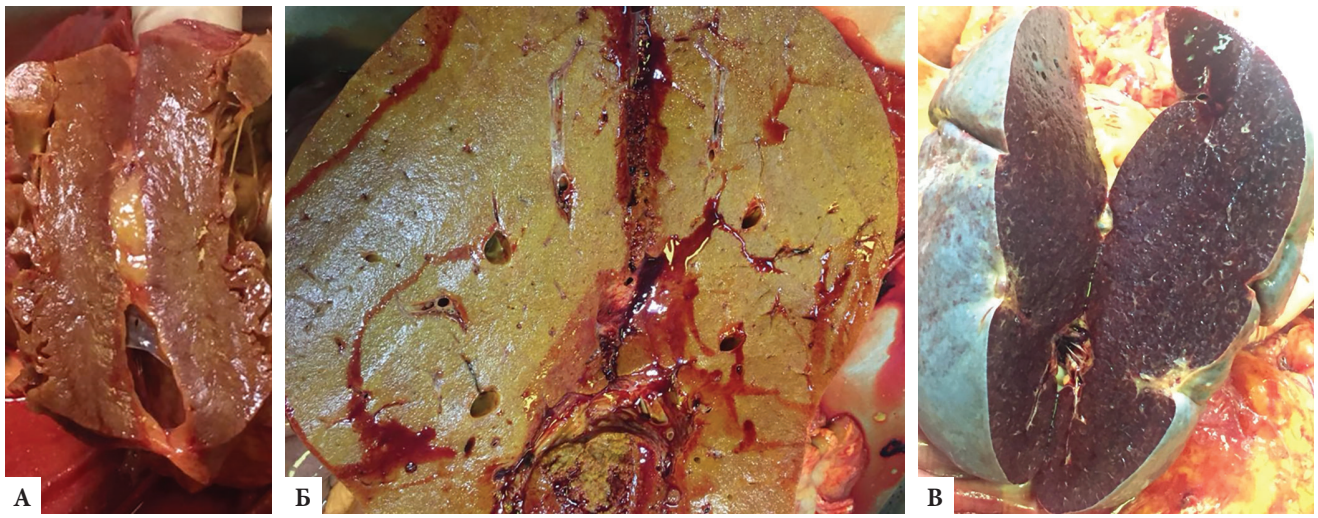


***Figure 5.*** *Macroscopic picture at autopsy: revealed massive damage to the heart (А), liver (Б), spleen (В) (Photo courtesy of Dr. EA Stepanova)*

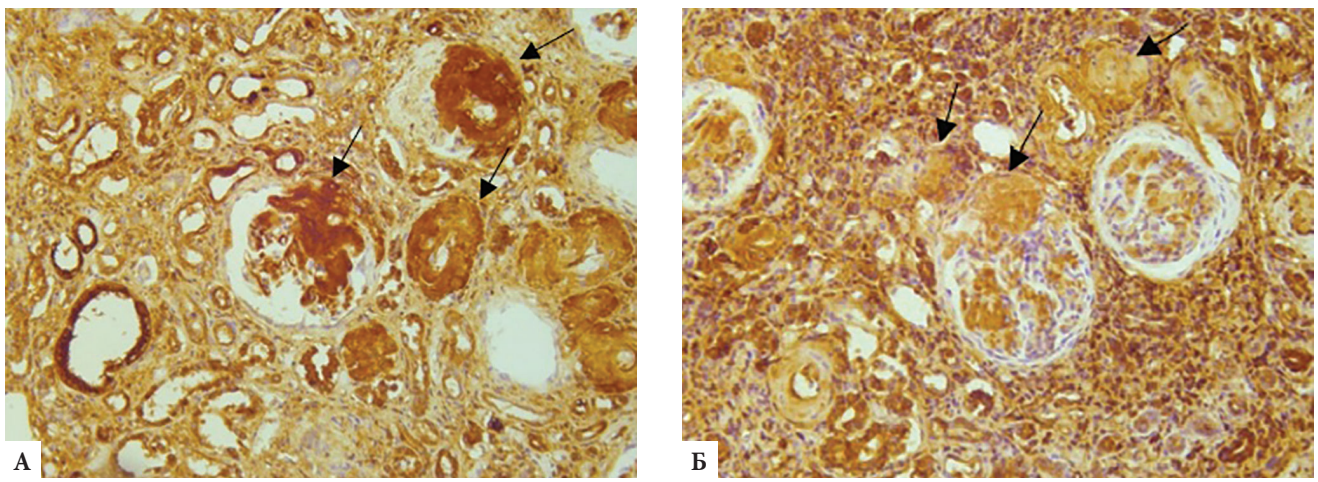


***Figure 6.*** *Immunohistochemical study (Photos courtesy of Dr.E. A. Stepanova)*
*А — Kidney, light chains K; Б — Kidney, light chains λ*

*Table 3. Myelogram*

| | Parameter | Normal value |
|---|---|---|
| Blasts, % | 2 | 0,1-2,8 |
| Neutrophilic promyelocytes, % | 2 | 1-4,1 |
| Neutrophilic myelocytes, % | 10 | 7-12,2 |
| Neutrophilic metmyelocytes, % | 4 | 8-15 |
| Neutrophilic stab, % | 5 | 12,8-23,7 |
| Neutrophil segmented, % | 24 | 13,1-24,1 |
| All neutrophilic elements, % | 45 | 52,7-68,9 |
| Eosinophils, % | 1 | 0,5-5,8 |
| Lymphocytes, % | 9 | 4,3-13,7 |
| Monocytes, % | 4 | 0,7-3,7 |
| **Plasma cells, %** | **8** | 0,1-1,8 |
| Erythroblasts polychromatophilic, % | 10 | 8,9-16,9 |
| Erythroblasts oxyphilic, % | 13 | 0,8-5,6 |
| Megaloblasts, % | 6 | 0 |
| All erythroid elements, % | 31 | 14,5-26,5 |
| Leukoerythroblastic index | 1,8 | 2,1-4,5 |
| Neutrophil Maturation Index | 0,6 | 0,5-0,9 |
| Maturation index of erythrokaryocytes | 0,9 | 0,7-0,9 |
| Megakaryocytes | + | + |

The patient's death was caused by systemic amyloidosis with damage to the heart, kidneys, liver, adrenal glands, thyroid and pancreas, gastrointestinal tract, complicated by bilateral focal pneumonia and acute heart failure.

## Discussion

Cardiac amyloidosis is observed in 33–60% of patients with AL amyloidosis [4, 5]. In AL amyloidosis, heart failure develops relatively early, in 22% of patients — as early as the onset of the disease [6, 7]. Cardiac damage in AL-amyloidosis is almost always associated with damage to other organs, most often the kidneys, as well as blood vessels, peripheral nervous system, the liver, gastrointestinal tract, and soft tissues [8]. Isolated cardiac involvement is observed in less than 5% of cases [4–6].

The presented patient with heart failure and AF had cardiorenal syndrome, monoclonal gammopathy, plasma cell dyscrasia [5, 9, 10]; the presence of amyloidosis was confirmed by an intravital morphological exam with Congo red staining and polarizing microscopy.

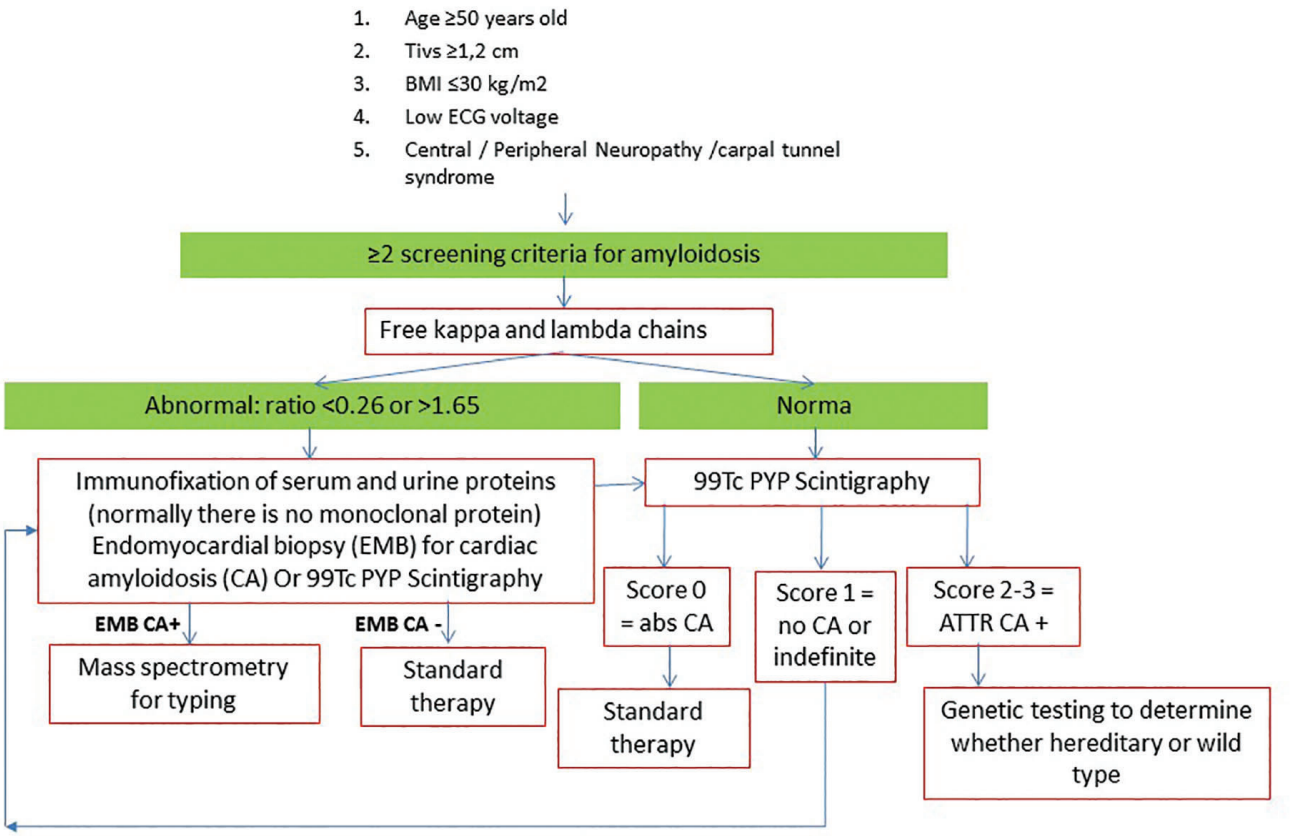


***Figure 7.*** *Algorithm for the diagnosis of cardiac amyloidosis in CHFpEF [5, 13]*
*ATTR CA — transtiretin cardiac amyloidosis, BMI — body mass index, CA — cardiac amyloidosis, CHFpEF — chronic heart failure with preserved ejection fraction; ECG — electrocardiogram; EMB AS — endomyocardial biopsy for cardiac amyloidosis; Tivs — interventricular septum thickness, 99mTc-PYP — 99mtechnetium pyrophosphate*

Due to the severity of the condition and late intravital diagnosis of amyloidosis, pathogenetic antiamyloid therapy was not prescribed to this patient, which led to a rapid progression of the disease and death. [5, 6, 11, 12].

For the timely diagnosis of cardiac amyloidosis in chronic heart failure with preserved ejection fraction (HFpEF), it is advisable to use the procedure shown in Figure 7.

Therefore, the variety and non-specificity of the clinical signs of amyloidosis often lead to a fatally late diagnosis of the disease. Suspicion of cardiac amyloidosis should arise with the idiopathic thickening of the LV myocardium walls to 12 mm or more, the presence of a restrictive type of diastolic dysfunction, idiopathic AF, chronic heart failure of unknown etiology, refractoriness to therapy, low voltage of the ECG teeth, arterial hypotension, syncope of unknown origin, pulmonary hypertension, nephrotic syndrome, and stage 4–5 chronic kidney disease. Electrophoresis of blood and urine proteins, identification of light chains of immunoglobulins, detection of amyloid deposits during intravital histological examination of various organs and tissues enable early and accurate diagnosis of AL amyloidosis, which is extremely important for improving the prognosis in such patients.

**Вклад авторов:**

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