ISSN 1607-419X
ISSN 2411-8524 (Online)
УДК 616.12-008.331.1

Rare cases of secondary hypertension: coarctation of aorta and idiopathic mid-aortic syndrome in middle aged patients

N. G. Avdonina[1], Yu. S. Yudina[1], I. V. Basek[1], O. G. Garbusova[2], V.Yu. Rakhmanov[3], K. V. Polyakov[4], A. I. Pushkarev[4], I. V. Emelyanov[1], N. E. Zvartau[1], A. O. Konradi[1]

[1] Federal Almazov Medical Research Centre, St Petersburg, Russia
[2] Far-Eastern State Medical University, Khabarovsk, Russia
[3] Clinical Diagnostic Center, Khabarovsk, Russia
[4] Federal Cardiovascular Center, Khabarovsk, Russia

Corresponding author:
Natalya G. Avdonina,
Federal Almazov Medical Research Centre,
2 Akkuratov street, St Petersburg, 197341 Russia.
Phone/fax: + 7(812)702–37–56.
E-mail: avdonina@almazovcentre.ru.

Received 15 November 2014; accepted 30 November 2014.

Abstract

In recent years, different advanced technologies for diagnosis and treatment of secondary endocrine and renovascular forms of arterial hypertension have been developed. However, rare causes of hypertension are still difficult for identification, especially when manifested with atypical clinical features. Two cases of atypical clinical manifestation of aorta coarctation at different level are discussed in this article. The article pays special attention to the causes and approaches to hypertension that usually manifests in childhood. At the same time in adult clinical practice young patients presenting with elevated blood pressure require careful examination and exclusion of secondary forms of hypertension even when the elevation of office blood pressure is mild-to-moderate.

Key words: secondary hypertension, coarctation of aorta, idiopathic mid-aortic syndrome

For citation: Avdonina NG, Yudina YuS, Basek IV et al. Rare cases of secondary hypertension: coarctation of aorta and idiopathic mid-aortic syndrome in middle aged patients. Arterial'naya Gipertenziya = Arterial Hypertension. 2014;20(6):600–607.

Редкие варианты симптоматической артериальной гипертензии: клинические случаи коарктации аорты и среднеаортального синдрома у больных среднего возраста

Н. Г. Авдонина[1], Ю. С. Юдина[1], И. В. Басек[1], О. Г. Гарбузова[2], В. Ю. Рахманов[3], К. В. Поляков[4], А. И. Пушкарев[4], И. В. Емельянов[1], Н. Э. Звартау[1], А. О. Конради[1]

[1] Федеральное государственное бюджетное учреждение «Федеральный медицинский исследовательский центр имени В. А. Алмазова» Министерства здравоохранения Российской Федерации, Санкт-Петербург, Россия
[2] Государственное бюджетное образовательное учреждение высшего профессионального образования «Дальневосточный государственный медицинский университет» Министерства здравоохранения Российской Федерации, Хабаровск, Россия
[3] Краевое государственное бюджетное учреждение здравоохранения «Клинико-диагностический центр» Министерства здравоохранения Хабаровского края, Хабаровск, Россия
[4] Федеральное государственное бюджетное учреждение «Федеральный центр сердечно-сосудистой хирургии» Министерства здравоохранения Российской Федерации, Хабаровск, Россия

Контактная информация:
Авдонина Наталья Георгиевна,
ФГБУ «ФМИЦ им. В.А. Алмазова» Минздрава России, ул. Аккуратова, д. 2, Санкт-Петербург, Россия, 197341.
Тел./факс: +7(812)702–37–56.
E-mail: avdonina@almazovcentre.ru

Статья поступила в редакцию 15.11.14 и принята к печати 30.11.14.

Резюме

В последние годы достигнуты большие успехи в выявлении и лечении многих симптоматических гипертензий эндокринного, реноваскулярного характера. Однако особую сложность представляет диагностика редко встречающихся заболеваний, особенно при стертой или нетипичной клинической картине. В статье представлены два клинических случая редких форм вторичной артериальной гипертензии — коарктации аорты и среднеаортального синдрома, которые характеризовались нетипичной клинической картиной и были выявлены у пациентов среднего возраста.

Ключевые слова: симптоматическая артериальная гипертензия, коарктация аорты, среднеаортальный синдром

Для цитирования: Авдонина Н. Г., Юдина Ю. С., Басек И. В. и др. Редкие варианты симптоматической артериальной гипертензии: клинические случаи коарктации аорты и среднеаортального синдрома у больных среднего возраста. Артериальная гипертензия. 2014;20(6):600-607.

Introduction

Currently, timely detection and correction of potentially reversible causes of elevated blood pressure (BP), so called symptomatic arterial hypertension (HTN), is considered a highly relevant topic. Great advances have been achieved in the diagnosis and treatment of primary hyperaldosteronism and other endocrine HTN, renovascular HTN, and algorithms and indications for screening were developed. Sometimes, however, in routine clinical practice, we are confronted with quite unexpected cases. This article presents two clinical cases of the accidental rather late (in middle-aged subjects) finding of congenital malformations, which usually manifest before the second decade of life. Moreover, there were no common clinical manifestations that would be indicative of the disease in order to start treatment. In the first case, it resulted in serious complications, while in the second one, an intervention was performed timely leading to the prognosis improvement only due to the persistence of the treating physician.

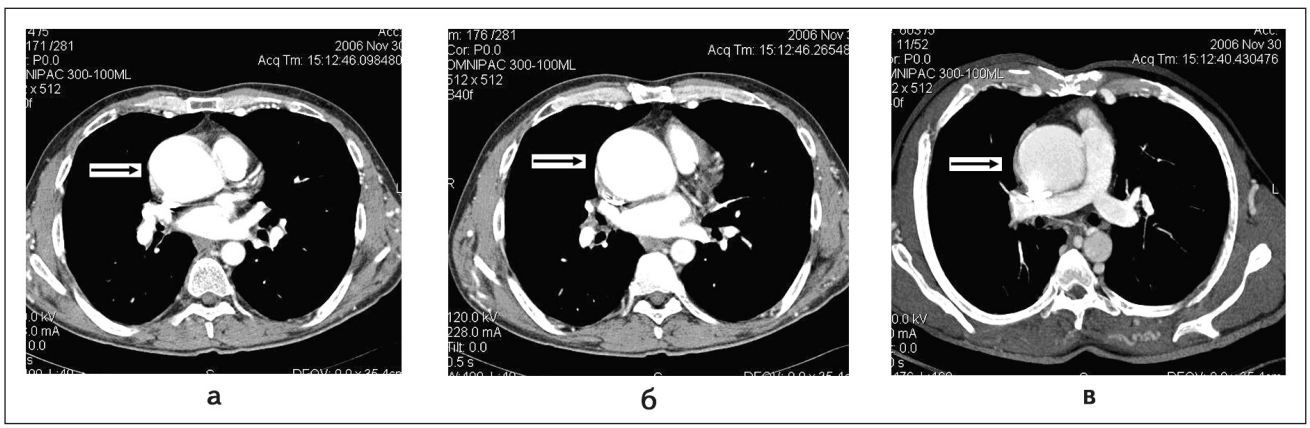
Clinical case № 1: aortic coarctation

Patient K. 54 years old was admitted to the V.A. AlmazovFederal North-West Medical Research Center for surgical treatment of the rheumatism-associated aortic valve stenosis in combination with angina pectoris III functional class (FC).

Since youth BP has been elevated. The patient was given an early discharge from military service on medical grounds due to HTN and suspected aortic valve disease (the murmur in the projection of the aortic valve was auscultated). In this regard, he was under the supervision of cardiorheumatologist, and the chronic rheumatic heart disease was diagnosed. Prescribed antihypertensive therapy was effective and included angiotensin converting enzyme inhibitors, beta-blockers, thiazide diuretics. For 2–3 years preceding admission to the hospital, the patient had experienced typical angina pectoris during exercise. The episodes had gradually progressed accompanying by shortness of breath. At the cardiologist's visit aortic valve stenosis was verified, and the patient was referred to the hospital for surgery.

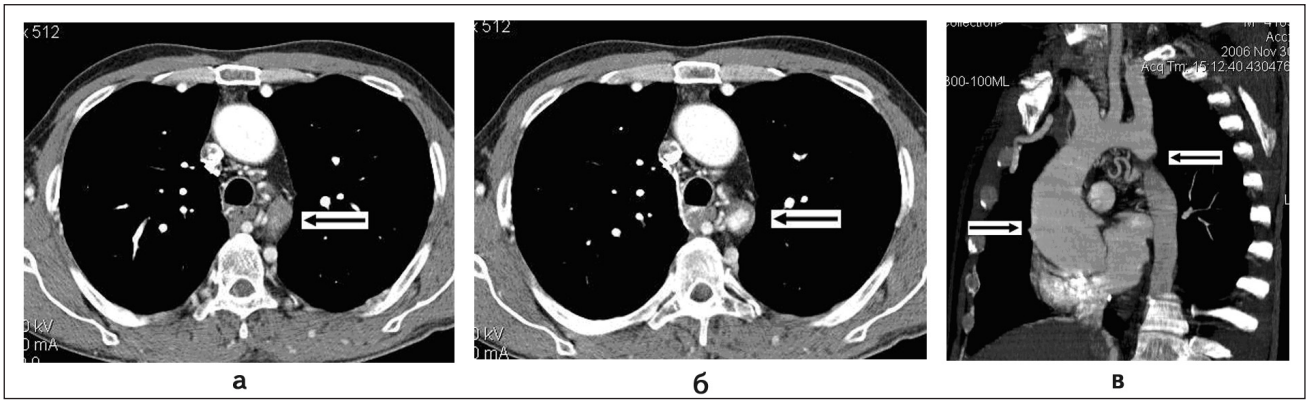
At admission the patient complained of general weakness, shortness of breath, episodes of compressive chest pain at moderate exertion (climbing 2 flights of stairs) that stopped spontaneously. The general state was satisfactory. The skin was clean and of physiological color and moisture. Office BP was elevated up to 160–170/90–100 mm Hg as measured on the brachial artery, and up to 190/120 mm Hg on the lower limbs. Pulse rate was rhythmic, symmetrical, and counted 66 beats per minute. Percussion demonstrated a moderate expansion of the left heart boundaries, and auscultation showed a rough systolic murmur, with a maximum above aortic valve, and transmitted to all auscultation points and the carotid arteries. In the lungs, vesicular breathing and no wheezing were auscultated. At palpation, abdomen was soft, painless, there was no enlargement of either liver, or spleen. No peripheral edema was found.

Figure 1. The patient Z., 54 years old. Aneurysm of the ascending aorta. Computed tomography angiography



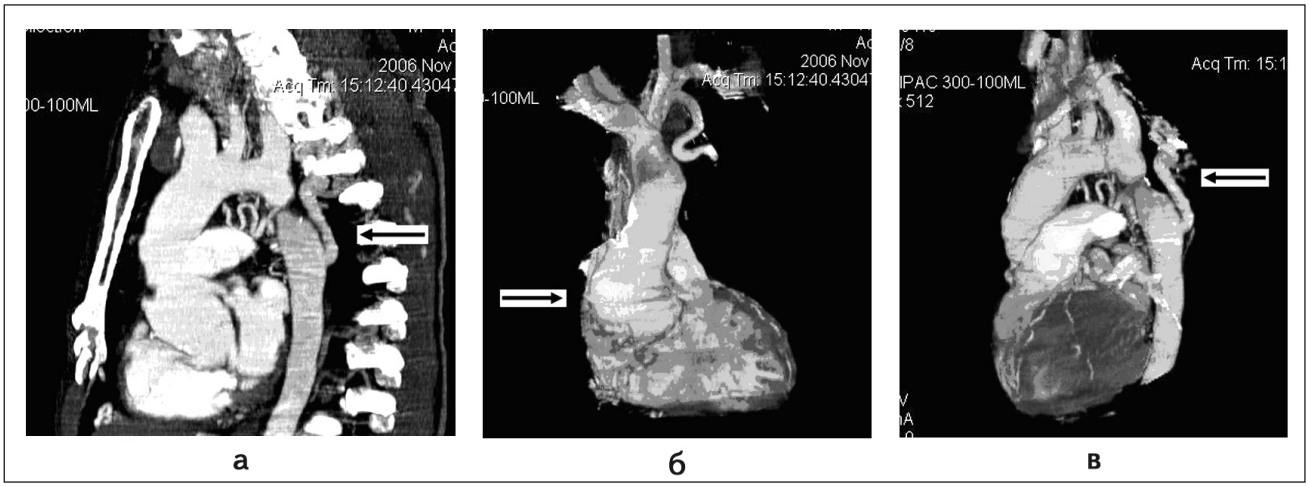
Note: At axial image aorta diameter is 6 cm at the level of coronary arteries (a, b) and pulmonary trunk (c) (→).

Figure 2. The patient Z., 54 years old. Aortic coarctation. Computed tomography angiography



Note: Axial images (a, b) show aortic coarctation with the narrowed diameter (till 1.5 cm) at the distance of 1.5 cm from left subclavian artery (contrast agent is not visualized over a distance of 0,37 cm) (→).

Figure 3. The patient Z., 54 years old. Aneurysm of the ascending aorta. Aortic coarctation. Computed tomography angiography



Note: There are two additional vessels (diameter 0,89 cm) coming from aorta 3,4 cm proximal to aortic stenosis, going up and joining subclavian arteries (→) (a). Aneurysm of the ascending aorta (→) (b). Aortic coarctation (c) with occlusion and collateral circulation (→) in the descending aorta.

Laboratory blood parameters were within reference range, no signs of the activity of rheumatic disease were found. Mild proteinuria was found. Electrocardiogram on admission showed a sinus rhythm 75 beats per minute, 1 single premature atrial beat, signs of left ventricular and atrial hypertrophy manifested by secondary repolarization changes, possible right ventricular fibrosis in anteroseptal area.

There were no focal changes on chest X-ray taken in two projections; vascular pattern was not changed. The examination showed right-sided pleuro-diaphragmatic adhesions; horizontal location of the heart, enlargement of the left heart chambers and the ascending aorta; narrow pulmonary artery; and a small erosion at the lower edge of the posterior parts of the IV–V ribs.

Echocardiography showed a congenital heart disease — bicuspid aortic valve, aortic valve opening of 14 mm, the pressure gradient of 19.1 mm Hg, a Aneurysma of the ascending, thoracic aorts and the arch of aorta (aortic diameter at the sinus level 34–44–60 mm) with the signs of the ascending aorta dissection, and a severe concentric left ventricular hypertrophy.

Due to the suspected aortic aneurysm dissection multislice computed tomography (MSCT) angiography ("Somatotom Sensation 4", Siemens) was performed at emergency including a topogram, native study (without a contrast enhancement)

and MSCT angiography in the arterial phase with intravenous bolus of contrast agent (Ultravist 370–100 ml, 3 ml/sec, the delay of 25 seconds). Post-processing analysis was performed, including three-dimensional reconstruction, MPR, MIP, VRT and SSD for the assessment of the location, shape, size of the aneurysm, the presence of thrombotic masses in the aneurysm cavity, dissection signs.

MSCT angiography demonstrated an aneurysm of the ascending aorta (II type by De Bakey) 6 cm in diameter and7 cm in length (Fig. 1). No signs of dissection or parietal thrombotic masses were identified. However, apart from the aortic aneurysm, a local narrowing of the aorta up to 1.3 cm (coarctation) was found at 1.5 cm from the left subclavian artery. The narrowed segment of the descending aorta was not visualized for 0.37 cm due to occlusion (Fig. 2). At a distance of 3.4 cm distal to the stenosis, two vessels of 0.89 cm in diameter were verified at two sides from the aorta extending upwards along the spine and connecting with the subclavian artery (Fig. 3). There was a developed network of collateral circulation t bringing blood to the descending aorta.

Thus, MSCT angiography identified previously undiagnosed congenital aortic abnormality — coarctation with signs of occlusion and the development of collateral blood circulation of the descending aorta, which likely was the cause of the ascending aortal aneurysm formation.

Coronary angiography, manometry, aortography were performed. Coronary angiography showed no atherosclerotic lesions or local stenosis of the coronary arteries, and a satisfactory coronary blood flow. Aortography demonstrated aneurysmal dilation of the ascending aorta with the maximum aortic diameter at the bulb level up to 5.7 cm. The aorta diameter at the level of brachiocephalic trunk was 3.0 cm, the diameter of the brachiocephalic trunk was 1.7 cm. There was a complete interruption of contrasting at the level of the aortic isthmus, the descending part was supplied through collateral vessels. There were no stenosis of subclavian or carotid arteries on either side. The blood pressure in the ascending aorta was 105/52 mm Hg, in the common femoral artery — 74/53 mm Hg. Prosthesis of the ascending aorta was performed. The surgery included valve-containing conduit (MedEng 25) implantation and reimplantation of the coronary arteries (Bentall operation), aorto-aortic bypass grafting (ascending aorta — thoracic aorta) by vascular prosthesis (diameter 12 mm) under extracorporeal circulation and cardioplegia.

The postoperative period was uncomplicated, BP stabilized at the level of 120–130/80–90 mm Hg, angina pectoris did not reoccur despite more active life style. Later on, combination antihypertensive therapy and anticoagulants were prescribed, and general state remains stable till now.

Clinical case № 2: midaortic syndrome

Forty-two year old patient K. referred to a doctor to get a medical permission for practicing amateur hockey. He had no limits in physical activities, and rarely visited medical doctors, had not attended any regular medical examinations, and considered himself healthy. Three years ago, at medical examination to obtain driving license an increased BP up to 150/100 mm Hg was found. For a short period of time, the patient received ACE inhibitors, but stopped due to the good general state. In addition, the patient complained of episodes of intense postprandial abdominal pain, requiring emergency care, episodes were regarded as “biliary colic”, and no profound examination was performed.

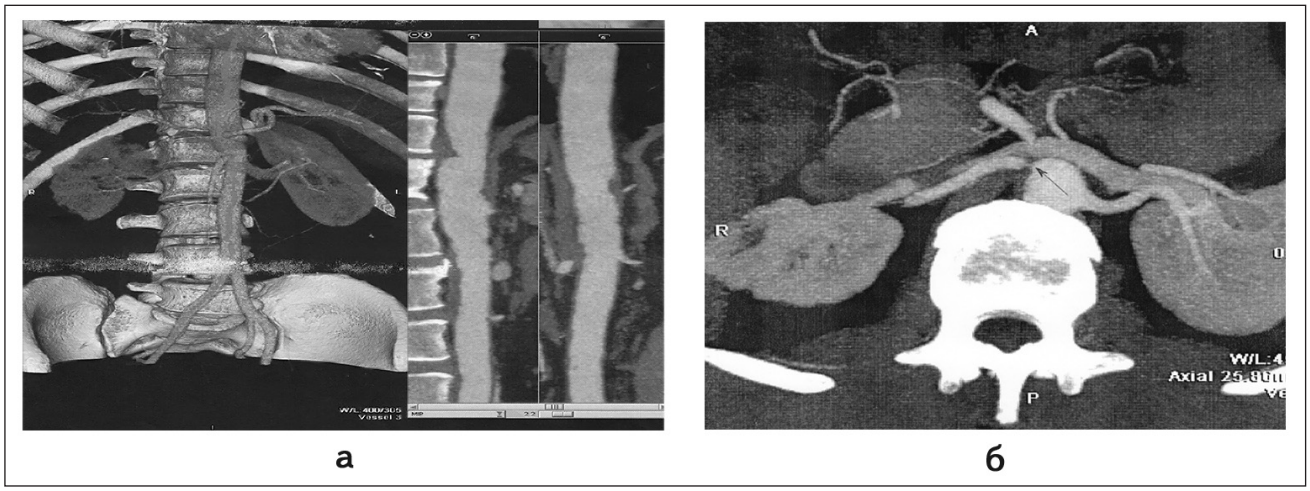
At admission the patient had no complaints. The general state was good. Pulse rate was rhythmic and symmetrical, and counted 66 beats per minute. BP — 170/110 mm Hg on both hands. The pulsation of peripheral arteries was clear, symmetrical without evident murmurs. Percussion and auscultation of chest did not show any abnormalities.

Electrocardiogram showed sinus rhythm with heart rate of 68 beats per minute, and the signs of left ventricular hypertrophy.

Echocardiography showed normal diastolic left atrial size — (3.2 cm), left ventricular end-diastolic dimension (5.5 cm), right ventricular end-diastolic dimension (2.5 cm), posterior wall thickness of left ventricle in diastole (1.1 cm), interventricular septum thickness in diastole (1.17 cm), ejection fraction (by Teichholz) of 62 %, and intact valves.

There was an increase in creatinine level up to 114 mmol/l (normal upper value is 110 mmol/l), microalbuminuria up to 100 mg/day. However, the estimated glomerular filtration rate was normal

Figure 4. The patient K., 42 years old. Computed tomography angiography



Note: (a) An extensive (22 cm) irregular thickening of the right aortic wall with the involvement of the right renal artery. The right kidney is small in size (a, b), and the contours are irregular, parenchyma is thin. There is a stenosis of the right renal artery up to 90 % (→) (b).

(85 ml/min). Ultrasound exam showed normal kidneys (size, parenchyma thickness were within normal limits, contours were equal, pyelocaliceal system was not enlarged, and respiratory excursions on both sides were normal).

In the absence of kidney disease and normal results of ultrasound examination, microalbuminuria and a mild increase in serum creatinine could be explained by the previously uncontrolled HTN, in particular, because target BP was achieved by felodipine 10 mg.

However, regarding BP level, 2 degree HTN, young age of HTN onset, microalbuminuria and increased creatinine level, contrast-enhanced MSCT of the abdomen and pelvis was performed (including topography, native study without contrast enhancement and MSCT angiography in the arterial phase with intravenous bolus of contrast agent Ultravist 300–100 ml and excretory phase).

MSCT showed an irregular contraction of the abdominal aorta extending for up to 22 cm (at the level of 11th thoracic vertebrae to the aortic bifurcation) due to the undulating irregular thickening of the aortic wall involving the right renal artery (Fig. 4 a, b) and the superior mesenteric artery leading to the significant stenoses.

The examination also showed a decrease in right kidney size — 4.3 × 4.7 × 8.5 cm, its contours were undulating and parenchyma was irregularly thin (Fig. 4a, b).No abnormality of the left kidney was found.

The excretory function of the right kidney was delayed at the excretory phase. Thus, MSCT enabled diagnosis of of the midaortic syndrome with the significant stenoses of the superior mesenteric and right renal arteries, and secondary right kidney shrinkage.

In The percutaneous balloon angioplasty of the renal artery preceded by aortoangiography of abdominal aorta, renal arteries and superior mesenteric artery was performed. It showed a stenosis of the right renal artery (90%), and mesenteric artery (90%) (Fig. 5 a, b). Percutaneous balloon angioplasty with stenting of the right renal artery was performed (Fig. 6 a). Balloon angioplasty with stenting of the mesenteric artery was performed as a second step (Fig. 6 b).

The postoperative period was uncomplicated. BP decreased till the normal values without antihypertensive therapy in the first days after surgery. The patient was discharged under the cardiologist's supervision, and recommendations included regular BP monitoring BP and renal function control once in 6 months. At 1.5-year follow-up BP level was normal without antihypertensive therapy stable, and not decrease in kidney function was found.

Discussion

We presented two cases of symptomatic HTN due to the congenital narrowing at different levels of the aorta.

Figure 5. Angiography. Stenosis of right renal artery achieving 90% (a). Stenosis of the superior mesenteric artery up to 90 % (b)

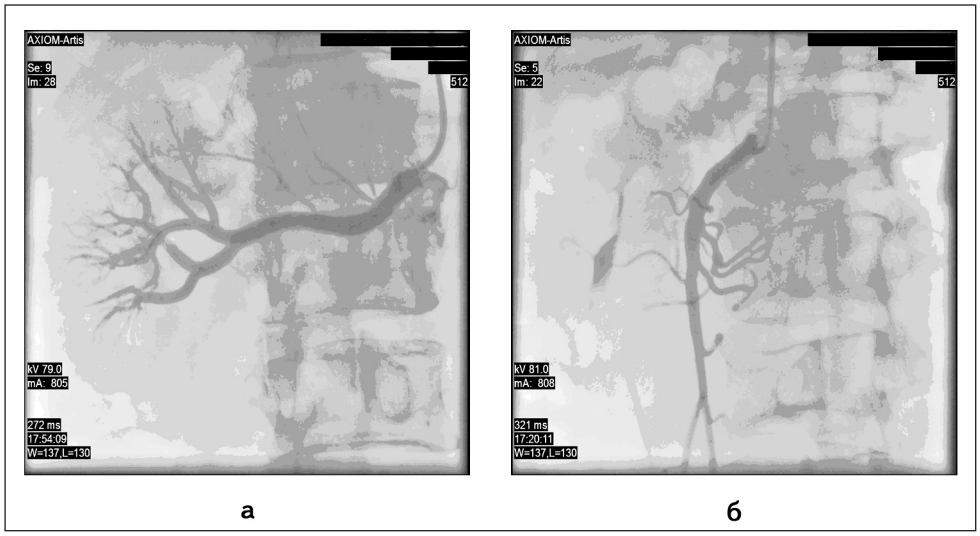
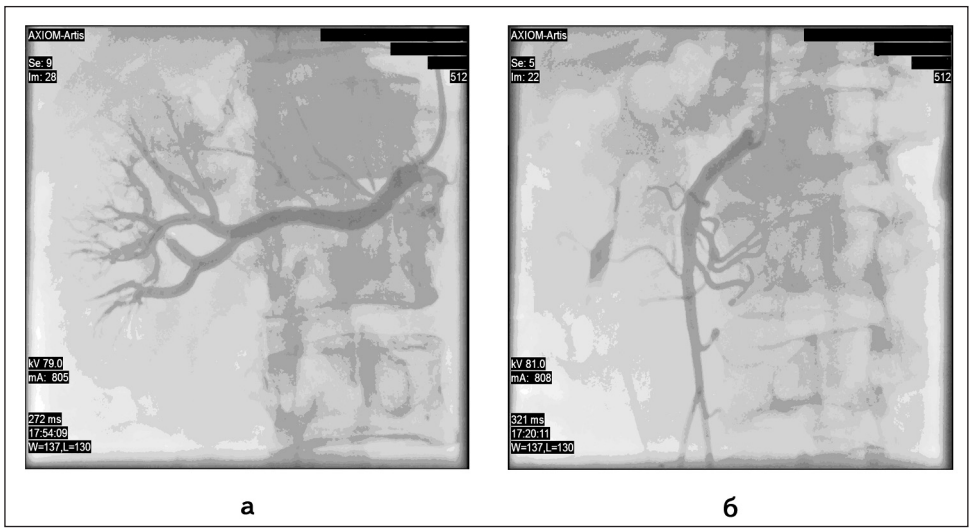


Figure 6. Angiography. Angioplasty with stenting of the right renal artery (a). Angioplasty with stenting of the superior mesenteric artery (b)



Aortic coarctation — a congenital segmental narrowing of the aorta, mostly found at the level of isthmus, was first described by JF Meckel in 1750. Its prevalence comprises 5.1–8.1% of all congenital heart diseases [2]. The most common location (88,3–98% of all cases of aortic coarctation) is ductus arteriosus, at the distance between the left subclavian artery and the arterial ligament, rarely (2%) — in the lower thoracic or abdominal aorta [3]. In our case a typical localization of aortic coarctation was observed. Moreover, in 60–70% cases, aortic coarctation co-exists with other congenital heart disease, like in our patient (a combination of aortic coarctation and bicuspid aortic valve). In addition, HTN was the only clinical manifestation in our patient.

Midaortic syndrome includes hypertension, absence of femoral artery pulsation and the murmur above abdominal aorta caused by the narrowing of the abdominal or distal parts of the descending aorta due to various etiologies [5, 6]. It is a rare pathology, occurring in 0.5–4.6% cases of all stenotic aortic diseases (including typical coarctation). The causes include Takayasu aortoarteritis, neurofibromatosis, fibromuscular dysplasia, retroperitoneal syndrome, mucopolysaccharidosis, Williams' syndrome, but an idiopathic abdominal aortic coarctation (atypical aortic coarctation) is the most common and manifests in adolescence. In 65% of patients, renal arteries are involved, and in 35% of cases visceral branches are also affected [6]. In our case, the location of midaortic

syndrome was typical, and despite the uncommon late onset of the disease it is likely to be idiopathic that was confirmed by the absence of any potential underlying disease.

The diagnosis was not established timely due to the atypical disease course, adequate collateral circulation in the first case and a moderate aortic narrowing and late onset in the second case. This led to the development of mild hypertension. In such cases, even slight changes in laboratory, instrumental and clinical data are very important. In the second case, microalbuminuria was, in fact, the only sign, and the further MSCT study allowed to establish correct diagnosis. Despite the benign course of the disease, at admission both patients demonstrated signs of complications such as heart failure, encephalopathy, kidney damage, and ischemia of gastrointestinal system. Surgery is the only approach to treat both congenital diseases, the operations vary from endovascular interventions to reconstructive surgery and aortic prosthesis. Successful surgical treatment results in improvement, target BP achievement, prevents target organ damage, and even leads to the regression of pathologic changes as observed in our cases.

In conclusion, even minor changes at examination should be considered in all hypertensive patients, and in particular in young and middle-aged subjects, even if HTN is mild and there is an evident response to treatment.

Conflict of interest
The authors declare no conflict of interest.

References

1. Fyler DC. Nadas' Pediatric Cardiology. Philadelphia PA, Hanley&Belfus. 1992:535–536.

2. Ben-Shoshan M, Rossi NP, Korns ME. Coarctation of the abdominal aorta. Arch Pathol. 1973;95(4):221–5.

3. Campbell M, Baylis JH. The course and prognosis of coarctation of the aorta. Br Heart J. 1956;18(4):475–495.

4. Razumovski AYu, Kulikova NV, Alkhasov AB et al. The mid-aortic syndrome: possibilities of vascular prosthetics. Ann Surg. 2011;4:75–79. In Russian.

5. Delis KT, Gloviczki P. Middle aortic syndrome: from presentation to contemporary open surgical and endovascular treatment. Perspect Vasc Surg Endovasc Ther. 2005;17 (3):187–203.

Author information:

Natalia G. Avdonina, MD, Researcher, Research Department for Hypertension, Almazov Federal Medical Research Centre;

Yulia S. Yudina, MD, Researcher, Research Department for Hypertension, Almazov Federal Medical Research Centre, St Petersburg, Russia;

Ilona V. Basek, MD, PhD, Head, Department of Radiology, Almazov Federal Medical Research Centre, St Petersburg, Russia;

Olga G. Garbuzova, MD, Physician, Far-Eastern State Medical University, Khabarovsk, Russia;

Vitaliy Yu. Rakhmanov, MD, Radiologist, Clinical Diagnostic Center, Khabarovsk, Russia;

Konstantin V. Polyakov, MD, Federal Cardiovascular Center, Khabarovsk, Russia;

Alexey I. Pushkarev, MD, Federal Cardiovascular Center, Khabarovsk, Russia;

Igor V. Emelyanov, MD, PhD, Senior Researcher, Research Department for Hypertension, Almazov Federal Medical Research Centre;

Nadezhda E. Zvartau, MD, PhD, Senior Researcher, Research Department for Hypertension, Almazov Federal Medical Research Centre;

Aleksandra O. Konradi, MD, PhD, DSc, Professor, Head, Research Department for Hypertension, Deputy Director on Research, Almazov Federal Medical Research Centre.