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Coarctation of the Aorta in Adults: is It Cardiac or Cardiac Surgery Problem?

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Abstract. The paper presents the analysis of practical possibilities of the method of multislice spiral computed tomography (MSCT) angiography of the thoracic and abdominal segments of the aorta and intravenous bolus contrast enhancement (Omniscan, Omnipaque) in the diagnosis of coarctation of the aorta in adults. 5 cases of primary diagnosis of coarctation of the aorta confirmed by MSCT-angiogram of the thoracic and abdominal segments of the aorta are presented. Clinical monitoring of patients with suspected coarctation of the aorta (CoAo) was accentuated. Symptoms of CoAo in adults included holosystolic murmur, hypertension in the upper extremities and hypotension in the lower ones, a weak or even absent pulse in the lower extremities, difference in the physical development of the trunk and lower extremities.

Keywords: coarctation of the aorta in adults; clinical signs; multislice spiral computed tomography angiography of the aorta

Problem statement and analysis of the recent research

Coarctation of the aorta (CoAo) is one of the most common congenital heart defects (CHD) that accounts for approximately 6-15% of all live births with CHD being 2 to 2.5 times more common in men than in women [1, 2]. The incidence of neonatal CDH is about 800 infants per 100,000 births. Around 50 infants are born with coarctation of the aorta and 40 newborns have isolated coarctation of the aorta (with or without patent ductus arteriosus). Among infants with isolated CoAo 10% die within the first few months of life and 20% die within the first year of life due to cardiac failure. Chronic cardiac failure is the leading cause of death of 10% of babies at the age of 1-4 years. About 50% of deaths among children in the first 10 years of childhood and about 25% of deaths among children up to the age of 20 years are associated with isolated CoAo [3].

The average life expectancy in CoAo is about 35 years [4]. CoAo is localized more frequently in the aortic isthmus region distally to the left subclavian artery close to Botallo's duct or corresponding ligament [5].

There was established that in the aortic wall in the area of coarctation (stenosis) the sclerotic process develops and progresses rapidly resulting in intimal thickening and then, (due to turbulence of blood flow after passing through the constriction) aneurysmal expansion is formed (4.5-5.5 cm) with further aneurysm development (>5.5 cm).The collateral circulation intensifies sclerotic changes in the vessels of the upper part of the body. The systolic blood pressure gradient between the upper and lower parts of the body leads to the formation of two circulation regimes – hypertensive and hypotensive in the upper and lower parts of the body, respectively [6].

CoAo is difficult to diagnose in adults as after 18 years of age due to the collateral circulation hemodynamics is stabilized, complaints become rare and measurements of blood pressure in the lower extremities and pulse rate at femoral arteries have not become a mandatory algorithm for the examination of patients with arterial hypertension (AH) in outpatient settings yet.

The objective of the research was to determine features of CoAo course and the possibilities of its early detection in adults and develop clinical recommendations to be added to Medical Care Protocols.

The research aimed at the selection of the most characteristic clinical and instrumental signs of CoAO in adults to identify such patients in the outpatient settings and rationalize the tactics for further treating such patients.

Materials and methods

The study included 5 people who applied to Treatment & Diagnostic Center "Simedgroup" due to discomfort in the heart area (atypical, non-anginal chest pain), high blood pressure (BP), increased pulsation in the vessels (carotid pulse, brachial arteries), and dizziness. The algorithm for the examination included clinical monitoring (complaints, heart rate, BP and pulse rate at the upper and lower extremities), MSCT- angiography of the aorta.

Results and discussion

The diagnosis of CoAo in all patients was confirmed by MSCT and intravenous bolus contrast enhancement (Omniscan, Omnipaque) of the thoracic and abdominal segments of the aorta and its branches.

Cases of CoAo in adults listed below were diagnosed within the two-year period of 2013 and 2015.

Case history No 1

A 34-year-old patient L., resident of the village of Tysmenychany Nadvirna district, Ivano-Frankivsk region, applied to "Simedgroup" complaining of discomfort in the heart area, dull headache, periodic increase in BP (>140/90 mm Hg), episodic irregular heartbeat. The patient developed hypertension at the age of 15 years. Antihypertensive drugs were taken irregularly. During 2 pregnancies resulting in normal deliveries (8-year-old and 4-year-old girls) she received methyldopa in order to control hypertension. The patient was examined by an endocrinologists (no pathology was detected). Hypertensive uncomplicated cerebral crisis (BP 180-190/100-110 mm Hg) with signs of transient ischemic attack (TIA) was diagnosed on the background of antihypertensive treatment in January 2015.

Objective data. The patient' height – medium; the type of constitution – normosthenic; the skin covering – clean, the skin colour – normal; BP in the upper extremities - 170/100 mm Hg, BP in the lower extremities – 60/30 mm Hg; lung sounds – vesicular; heart sounds – rhythmic, heart rate – 85 beats per minute, diastolic shock and holosystolic murmur with epicenter in fossa jugularis; no peripheral edemata.

Laboratory findings. Complete blood count: hemoglobin – 110 g/l; erythrocytes: 3.4 x 10^{12}/l; colour index – 0.90; leucocytes – 6.3 x 10^{9}/l; ESR – 14 mm/hour.

Biochemical analysis of blood: metanephrine (24-hour urine) – 212.79 mcg/daily (N – 25-31 mcg/daily); aldosterone – 62.09 pg/ml; serum K^{+}– 4.3 mmol/l (N 3.5 – 5.1 mmol/l); serum N^{+} – 135 mmol/l (N – 136 – 145 mmol/l); INR – 1.0.

Electrocardiography: normal sinus rhythm and HR of 85 beats per minute were detected; the Sokolow-Lyon index was 37 mm, $R_{V5}>R_{V4}$, there were signs of early repolarization of the left ventricle (LV).

24-hour Holter ECG monitoring: the average HR of 85 beats per minute throughout the day and 63 beats per minute through night, isolated left ventricular extrasystoles, sinus tachycardia during the active period of the day, post-tachycardic left ventricular myocardial hypoxia, long QT syndrome (580 mc), 143 episodes of tachycardia for 24 hours were detected.

Echocardiography: the aorta – 2 cm (the level of aortic root), the left atrium – 3.2 cm; end-diastolic dimension (EDD) – 4.7 cm; end-systolic dimension (ESD) – 3.3 cm; end-diastolic volume (EDV) – 102 ml, end-systolic volume (ESV) – 44 ml, ejection fraction (EF) – 59%, SWTd – 1.2 cm, SWTds – 1.31 cm, PWTd – 1.0 cm, PWTs – 1.29 cm; the ratio of wall thickness to left ventricular radius – 0.42; fibrosis of the aortic walls, anterior mitral valve leaflet prolapse mitral regurgitation 1+; supplemental chord of the left ventricle, hypokinesis of the basal and medial anterior, anteroseptal segments, and the anterior and septal apical segments.

Ultrasonography of the abdominal organs, thyroid gland (TG) and adrenal glands revealed hypodynamic biliary dyskinesia; thyroid tissue was homogeneous, blood supply was found to be preserved; the size and structure of adrenal glands were normal.

Multislice spiral computed tomography (MSCT) angiography of the thoracic and abdominal segments of the aorta. MSCT-angiography of the thoracic segment of the aorta revealed that there was a severe narrowing of the aortic lumen (critical stenosis) of 0.15 cm 2.1 cm distally to the left subclavian artery (Fig. 1A).

Diagnosis: Congenital heart defect: coarctation of the descending thoracic aorta (Ø-0.15 cm) DeBakey type III A with co-existent stage II arterial hypertension in the upper part and hypotension in the lower part of the body.

In this case CoAo was oligosymptomatic. The patient had two pregnancies resulting in normal deliveries. She performed her professional duties and household tasks without any limitations.

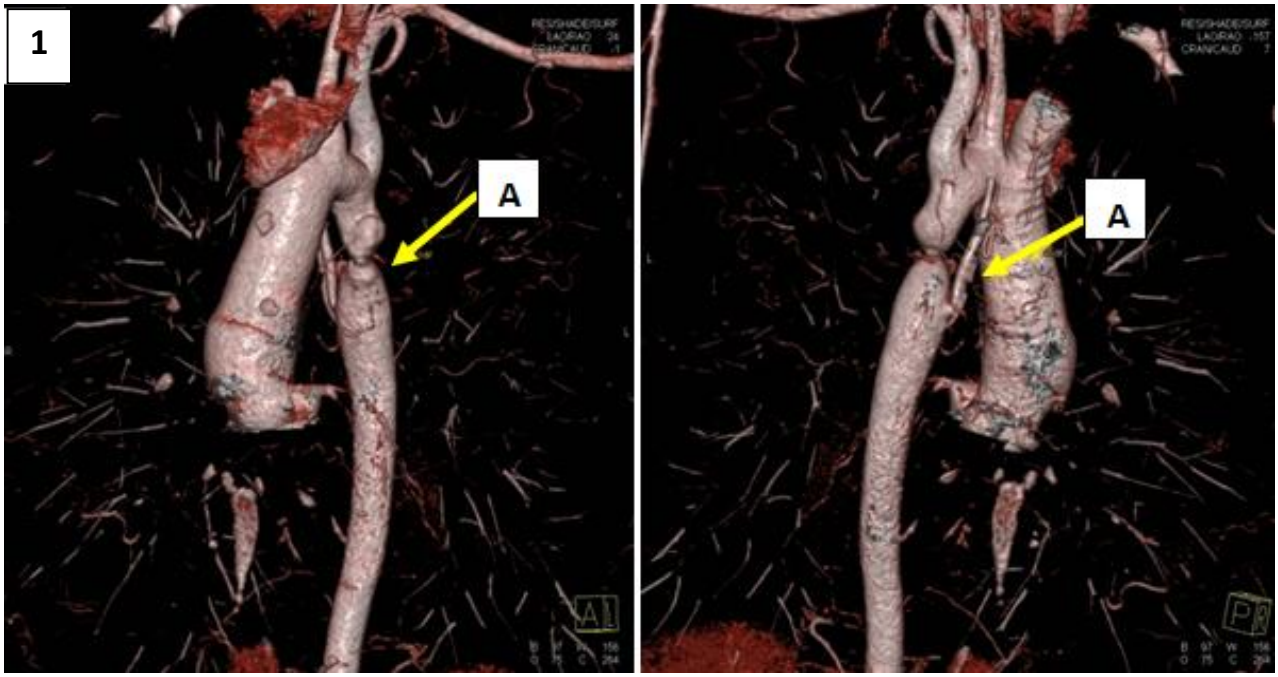

Fig. 1. MSCT of the thoracic segment of the aorta (with contrast enhancement) in a 34-year-old patient L

Characteristic features of late diagnosis of CoAo in a 34-year-old patient L. included subclinical patterns (absence of the pulse in the upper part of the body), the underestimation of strengthened second tone of the aorta and holosystolic murmur with epicenter in the jugular fossa, carelessness of the doctors regarding mandatory measurement of blood pressure in the lower extremities in case of elevated brachial BP and pulselessness at aa. femorales, popliteae et dorsalis pedis. The investigation of brachial-tibial pulse wave velocity or carotid–tibial pulse wave velocity (using rheography) and MSCT could be helpful in timely diagnosing CoAo in this case (Fig. 1).

Case history No 2

A 22-year-old patient M. complaining of dizziness, high BP >140/90 mm Hg, throbbing pain in the temples, discomfort near the breastbone on the left side, claudication when walking applied to "Simedgroup" to be examined and determine the cause of hypertension.

Clinical examination allowed us to establish concentric left ventricular hypertrophy (LVH), over-dipper hypertension and hyperkinetic hemodynamic state.

MSCT-angiography of the thoracic and abdominal segments of the aorta revealed that there was a sudden narrowing of the aortic lumen by 0.3 cm in the descending aorta immediately beyond the truncus brachiocephalicus and the left subclavian artery (Fig. 2A). The lumen of the right and left subclavian arteries, and the brachiocephalic trunk was extended.

There was observed a collateral network around the anterior chest wall and two collateral arterial branches having diameter of 0.75 cm (Fig. 2B).

Diagnosis: Congenital heart defect: coarctation of the descending thoracic aorta (Ø-0.3 cm) DeBakey type III A with co-existent hypertension in the upper part and hypotension in the lower part of the body.

Characteristic features of this case included the presence of high-grade aortic coarctation with the signs of hyperkinetic hemodynamic state and hypertensive cerebral crises which could be avoided with timely diagnosis using MSCT.

Case history No 3

A 25-year-old patient T. complaining of frequent headache, palpitation, discomfort near the breastbone on the left side applied to "Simedgroup" to be examined. These symptoms have been present since childhood. The patient has never been examined by a physician. His BP was 180/110 mm Hg; HR was 86 beats per minute; the pulse at a.radialis dextra was high, hard, and fast; on the left side deficient pulse was felt.

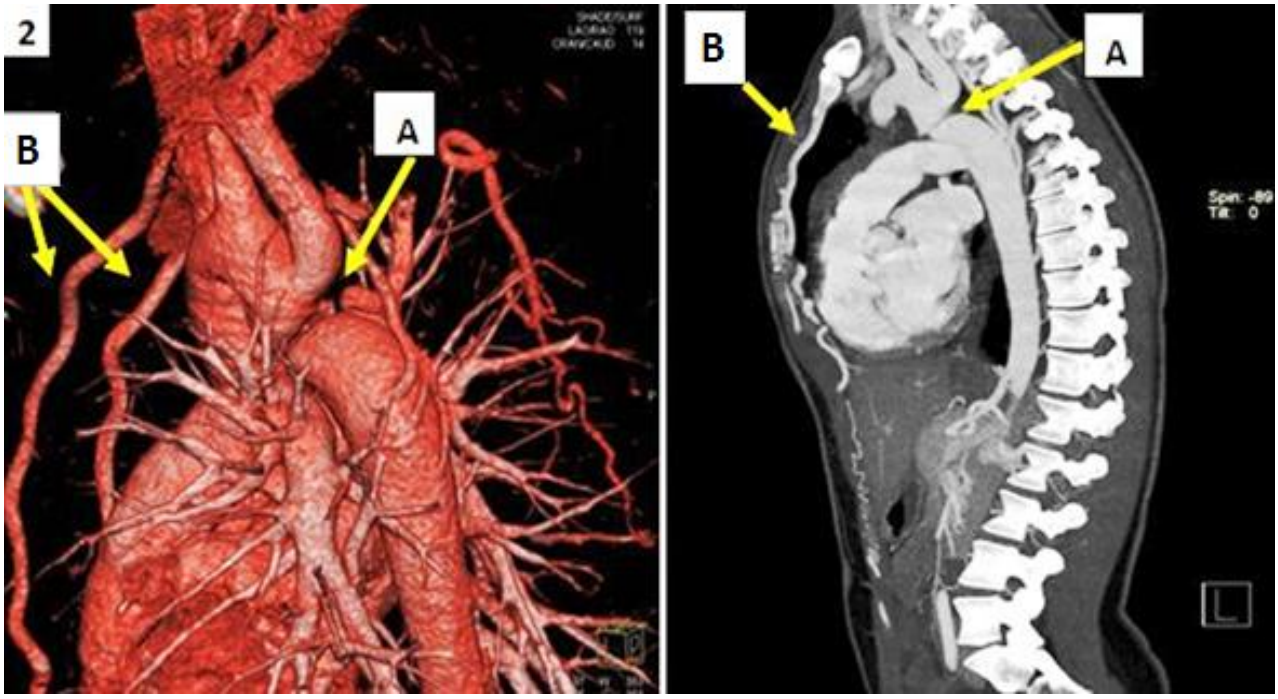

Fig. 2. MSCT of the thoracic and abdominal segments of the aorta (with contrast enhancement) in a 22-year-old patient M

ECG and EchoCG results: concentric left ventricular hypertrophy, signs of early repolarization of the ventricles (ECG), EF – 66%, EDD – 6 cm, EDV – 180 ml, left ventricular myocardial mass index (LVMMI) – 230 g/m^2 (EchoCG).

MSCT-angiography of the thoracic and abdominal segments of the aorta revealed that there was a narrowing of the aortic lumen in the narrowest portion by 0.45 cm 2-3 cm distally to the left subclavian artery (Fig. 3A). The width of the lumen increased gradually being 1.65 cm at the level of the diaphragm and 1.3 cm at the level of renal arteries. Uniform extensions of the lumen, brachiocephalic trunk, common carotid and left subclavian artery were also observed (Fig. 3B).

Diagnosis: Congenital heart defect: aneurysmal dilatation of the ascending aorta, coarctation of the descending thoracic aorta (Ø-0.45 cm) DeBakey type III B with stage II secondary hypertension.

Characteristic signs of CoAo in this case were cerebral and cardiac symptoms caused by aneurysmal dilatation of the ascending aorta and vessels of the brachiocephalic trunk.

Aneurysmal dilatation of the left subclavian artery resulted in pulse asymmetry: it was high on the right and low on the left.

Case history No 4

A 33-year-old patient K. applied to “Simedgroup” as long-term treatment of hypertension with different antihypertensive drugs did not result in expected lowering BP to a target BP < 140/90 mm Hg. She continued to suffer from persistent headache with a throbbing pain in the temples. The patient underwent surgical repair of coarctation of the aorta 5 years ago (2010) and her condition has been satisfactory for 1.5-2 years. The above mentioned complaints have been present since 2012.

MSCT-angiography of the thoracic and abdominal segments of the aorta revealed that the diameter of the aorta at the level of its arch in the widest area was 2.0 cm, and in the area of the transition of aortic arch to the ascending aorta its diameter was 1.2 cm and its lumen was 0.9 cm (4A).

Diagnosis: Congenital heart defect: recoarctation of the descending thoracic aorta (Ø-0.9 cm), condition after ineffective repair of coarctation of the aorta (2010), proximal left subclavian artery stenosis, stage II secondary AH.

Characteristic feature of this case is ineffective surgical repair of CoAo as indicated in the literature [5]. Authors indicated that in any case of CoAo including recoarctation of the aorta surgical intervention is needed.

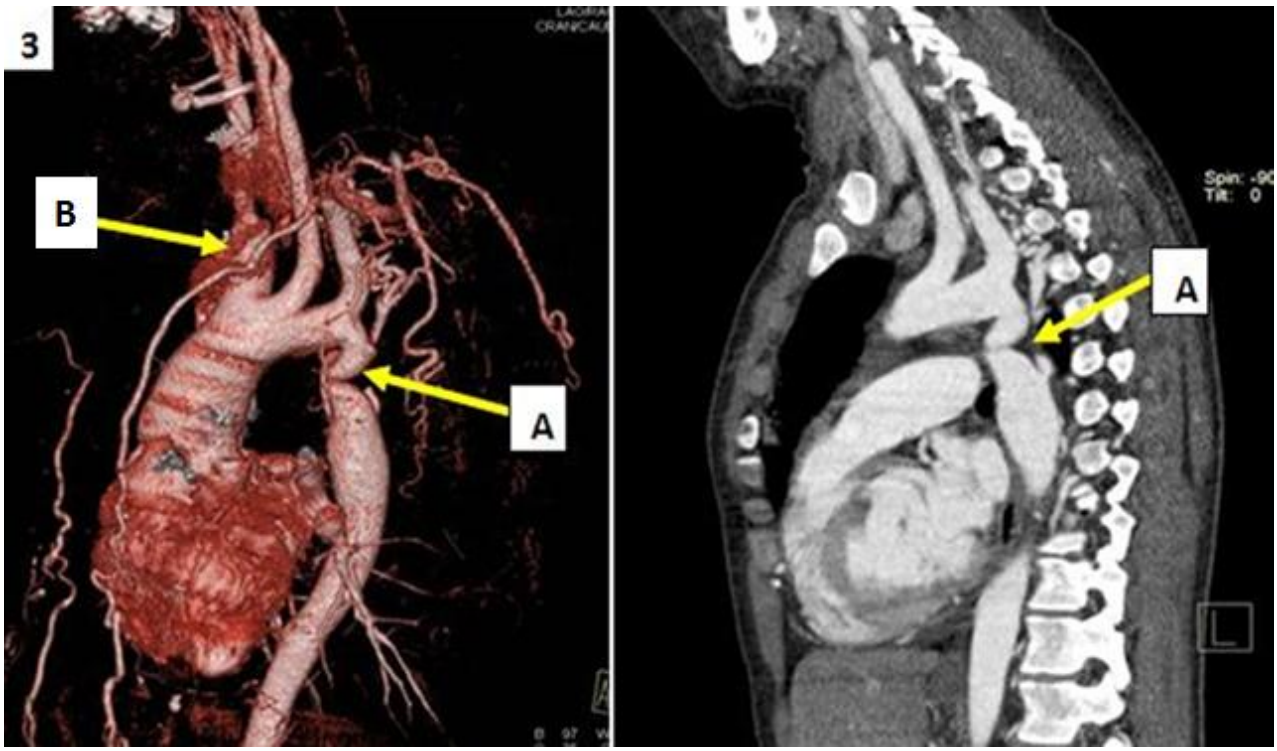

Fig. 3. MSCT of the thoracic and abdominal segments of the aorta (with contrast enhancement) in a 25-year-old patient T

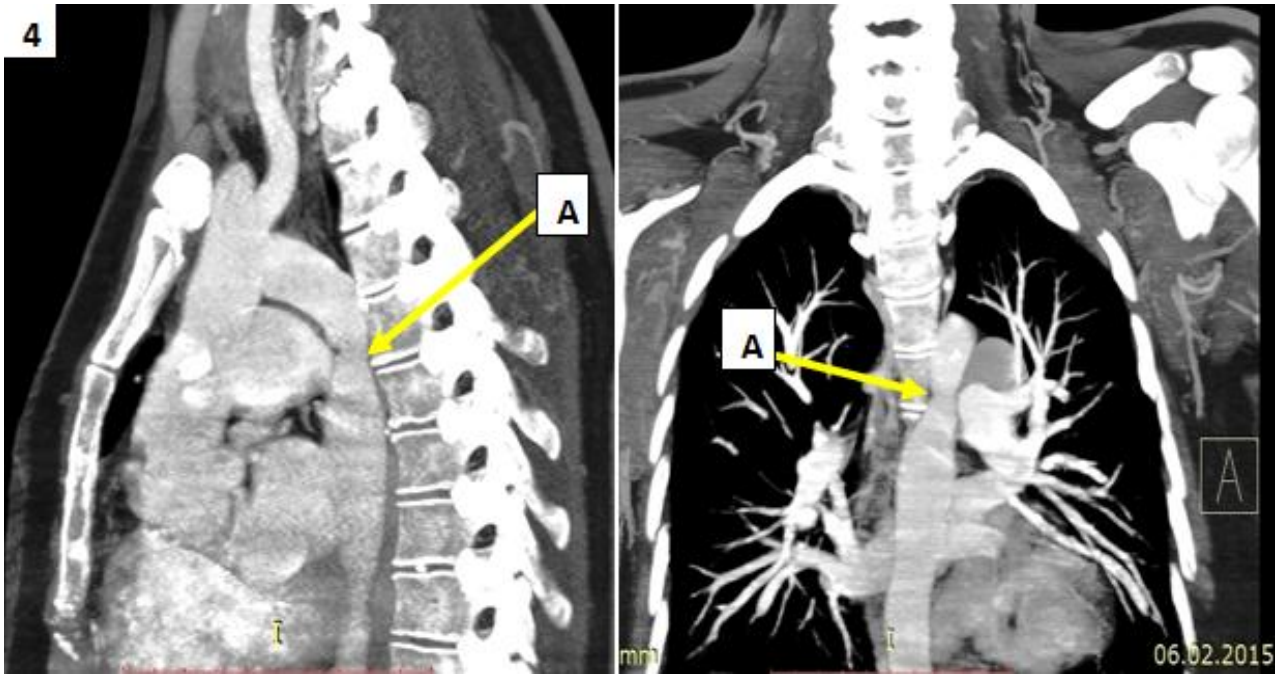

Fig. 4. MSCT of the thoracic segment of the aorta (with contrast enhancement) in a 33-year-old patient K

Case history No 5

A 34-year-old patient P. has been ill since childhood. The patient constantly complained on severe headache. She associated the disease with high BP diagnosed at school age. The patient was exempted from physical education due to hypertension. During the examination BP in the upper extremities was 160/98 mm Hg and in the lower ones it was 90/60 mm Hg, pulse at the left femoral artery was of poor volume.

MSCT–angiography of the thoracic segment of the aorta revealed that there was a narrowing of the aorta by 1.25 cm within 0.5 cm (5A).

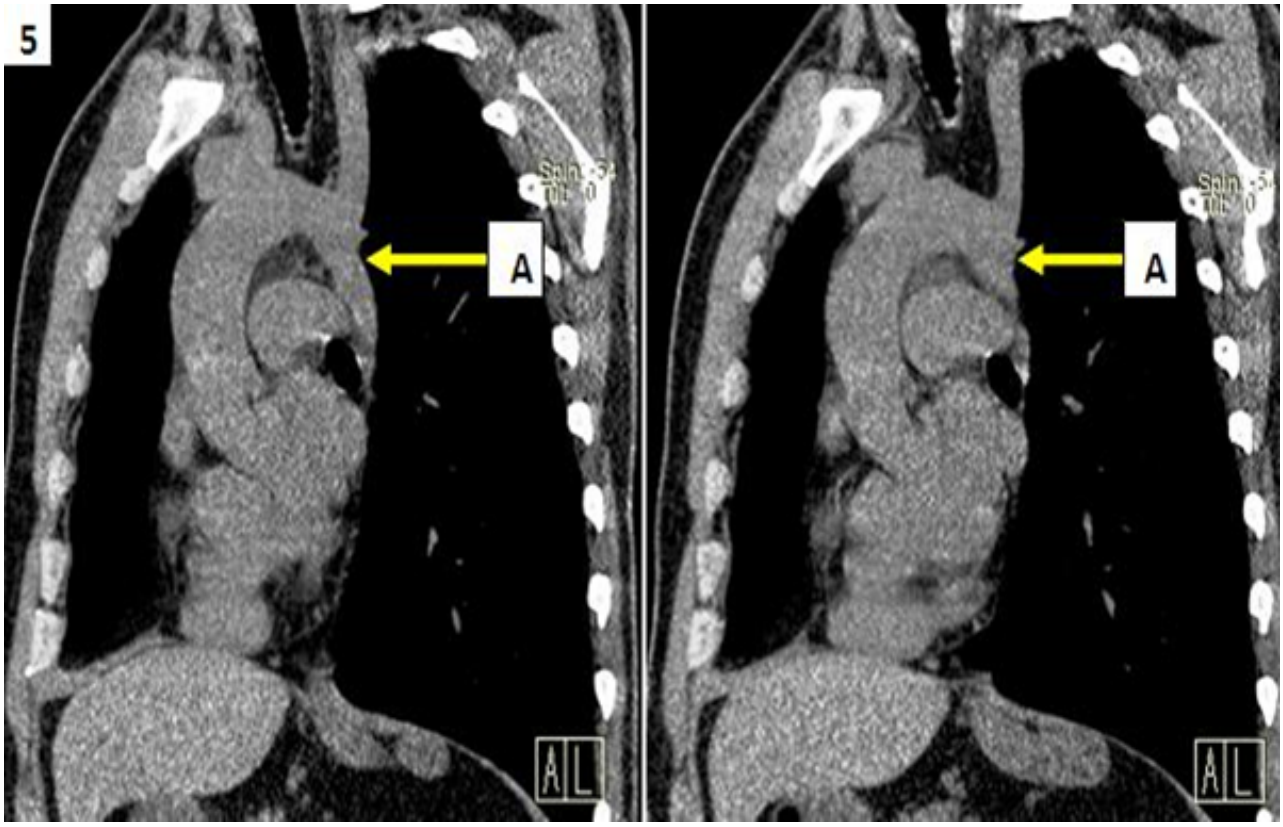

Fig. 5. MSCT of the thoracic segment of the aorta in the parasagittal plane (without contrast enhancement) in a 34-year-old patient P

Thus, CoAo in adults has a number of features which complicate its timely diagnosis.

In general, CoAo in adults has subclinical patterns; the upper and lower parts of the body often develop properly (no disproportion); normal physical activity and working efficiency are preserved. Inadequate attention of physicians during the examination to patients with hypertension also contributes to untimely diagnosis of CoAo. It is about mandatory BP measurement not only at brachial but femoral arteries as the strength of the pulse reduces significantly in CoAo. Therefore, in hypertension at a younger age multislice spiral computed tomographic angiography with contrast enhancement of the aorta must be used.

Conclusions

1. Coarctation of the aorta remains relevant for providing medical care to cardiac patients being ill since birth to adulthood and requires the attention of a neonatologist, pediatrician as well as cardiologist and cardiac surgeon.

2. The causes of late detection of coarctation of the aorta in adults include the absence of typical clinical signs of CoAo, overestimation of cases in which any increase in BP is associated with hypertensive disease, inadequate attention of physicians to measurements of BP in the lower extremities and evaluation of the arterial pulse volume in the upper and lower parts of the body (in CoAo pulse volume at the femoral artery decreases at early stages of the disease).

3. CoAo in adults is often accompanied by hypertensive complicated cerebral crises, excessive production of catecholamines, concentric left ventricular hypertrophy, long QT syndromes and early repolarization of the myocardium.

4. Multislice spiral computed tomography angiography of the aorta is the most reliable method for diagnosing coarctation of the aorta in adults. It should be used when treating all young patients with hypertension. Low pulse volume which can be palpated in the femoral arteries and low blood pressure secondary to hypertension in the femoral arteries is a simple, available to every doctor and effective auxiliary method of screening this pathology.

References

1. Panichkin YuV, Cherpak BV, Ditkivskyi IO, et al. Stenting of the aortic isthmus in coarctation in adolescents and adults. Shchorichnyk naukovykh prats Asotsiatsii sertsevo-sudynnykh khirurhiv Ukraiiny. Kyiv. 2010;465-466.

2. Kovalenko VN. Guide to cardiology. Kovalenko VN, editor. Kyiv. Morion. 2008;935-937.
3. John W. Kirklin, Brian G. Barratt-Boyes. Cardiac surgery. 2nd ed. Churchill Livingstone. New York. 1993;1273-1275.
4. Burakovskiy VI, Bokeriya LA. Cardiovascular surgery. Meditsina.Moscow. 1989;298-310.
5. Ozeryanskiy NA, Aksenova NA, Vashkeba VYu, et al. Case of surgical treatment of coarctation of the aorta in adult patient. Kardiokhirurhiia ta interventsiina kardiolohiia. 2014;2(7):28-32.
6. Dykukha SO. Combined autoplasty of the aorta in surgical repair of coarctation. Ukraiinskyi medychnui chasopys. 2001;3(23):127-128.