

MINISTRY OF HEALTH OF UKRAINE
ODESA NATIONAL MEDICAL UNIVERSITY

Faculty Medicine

Department Surgery, Radiological Diagnostics, Radiation Medicine,
Therapy and Oncology

APPROVED BY
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METHODOLOGICAL RECOMMENDATION
FOR PRACTICAL CLASSES OF THE ACADEMIC DISCIPLINE

Faculty, course Medical 6th year

Academic discipline Surgery
(name of the discipline)

PRACTICAL CLASSES

Practical class № 23

Topic: “Aortic arch syndrome: clinic, differential diagnosis, treatment approaches”

Approved:

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PRACTICAL CLASSES

Practical class № 23

Topic: “Aortic arch syndrome: clinic, differential diagnosis, treatment approaches”

1.Relevance of the topic

Among diseases of main arteries, aortic arch syndrome occupies a special place due to the complexity of diagnosis, the severity of clinical manifestations and complications, and the difference in approaches to conservative and surgical treatment. In the majority of patients, and mainly those of young working age, the disease quickly leads to serious complications in the absence of treatment, significantly reducing work capacity.

Despite the relatively low frequency of manifestations in the population, doctors of any specialty need to be able to detect aortic arch syndrome, properly organize examination and treatment of the patient. In the future, during outpatient observation of patients, careful monitoring of the degree of inflammatory and ischemic manifestations of the disease is necessary.

2. Objectives:

2.1. Learning objectives:

A student of higher education must learn how to:

- | | |
|--|-----------|
| 1. Identify anamnestic and clinical objective signs of the syndrome of aortic arch (non-specific aorta arteritis). | II level |
| 2. Basic principles of diagnosis of non-specific aortoarteritis | II level |
| 3. Assign an examination plan with using laboratory, X-ray, ultrasound methods examination. | III level |
| 4. Provide emergency conservative care to patients on non-specific aortoarteritis | III level |
| 5. Determine indications for surgical intervention and theoretically know the methodology their implementation. | II level |

2.2. Educational objectives:

1. Formation of a professionally meaningful personality of the doctor
2. To emphasize the participation of the national surgical school in the development of modern methods of treatment of the aortic arch syndrome

3. Interdisciplinary integration.

№	Disciplines	To know	To be able to
1	2	3	4
I. Previous disciplines			
1.	Anatomy	The structure of the aorta and large branches, blood supply to the neck, head, and limbs.	To be able to differentiate various vessels during operations and their branches
2.	Physiology and pathophysiology	Features of blood circulation of the brain, neck, upper limbs	Be able to interpret hemodynamic disorders in patients with aortic arch syndrome.
3.	Histology	The structure of the arterial wall.	To be able to interpret histological data by micro preparations.
4.	Pharmacology	The mechanism of action of drugs used in the treatment of aortic arch syndrome	
II. Intersubject integration			
1.	Nonspecific aortoarteritis	Signs of the disease, X-ray and ultrasound signs of characteristic changes in the main vessels	Interpret the data of hagiography, ultrasound and endoscopic examination.
2.	Atherosclerosis of vessels	Know etiology and clinical manifestations	Purposefully collect anamnesis, be able to find signs of chronic ischemia.
3.	Endarteritis (Thromboangiitis)	Know the clinical picture, differential diagnosis	To be able to examine the patient and put the primary diagnosis.
4.	Thrombosis of main vessels.	Know the features of the clinical picture and diagnosis of acute ischemia of limbs and individual bodies	Be able to carry out a differential diagnosis, interpret X-ray and duplex data research, to put readings before treatment.

4. Content of the class

Aortic arch syndrome is the most common manifestation of nonspecific aortoarteritis. Nonspecific aortoarteritis (NAA) - systemic disease of autoimmune genesis, which leads to the walls of the aorta, main arteries and ischemia of the corresponding organ.

Until the mid-60s, there were various names for nonspecific aortoarteritis -

pulseless disease, aortic arch syndrome, young women's arteritis, occlusive thrombarteriopathy, brachiocephalic arteritis, syndrome of the middle part of the aorta, Martorell syndrome or obliteration syndrome of supra-aortic trunks, aortitis syndrome, etc. Term Takayasu's disease was introduced into clinical practice by Caccamise, Whitman in 1952.

Among the brachiocephalic arteries, the subclavian arteries are most often affected, especially the left. The clinic of the lesion includes symptoms caused by ischemia of the upper limb and symptoms of ischemia of the brain. The main complaints of patients suffering from ischemia of the upper limbs are weakness in the hands, numbness and rapid fatigue of the hands during physical exertion. To one degree or another, symptoms of ischemia of the upper limbs are observed in more than 90% of patients with lesions of the subclavian arteries.

Clinical manifestations of transient ischemic attacks are characterized by attacks of loss of consciousness, impaired vision up to blindness, transient paresis and paralysis, dizziness with the development of unsteadiness and loss of balance. In most cases, these symptoms are recorded in patients with an open circle of Willis.

At the same time, with this localization of NAA there is the greatest difference between the large volume of damage to the branches of the aortic arch and the poverty of clinical symptoms is possible. In more than 50% of patients with damage to the brachiocephalic arteries, the course of the disease is neurological asymptomatic. Only 11% of patients with a lesion of the branches of the aortic arch suffered an acute brain injury before entering the clinic blood circulation.

Diagnosis of the lesion of the brachiocephalic arteries, as a rule, is not complicated. Most often registered:

- absence of pulsation on one or both radials' arteries,
- systolic noise in the supraclavicular region with damage to the subclavian arteries,
- systolic noise over the general sleepy one's arteries,
- blood pressure gradient between the upper limbs.

Duplex scanning and X-ray contrast play a decisive role in the diagnosis of the syndrome research.

The purpose of duplex scanning is primarily to assess:

- morphological changes in the affected arteries;
- the state of the internal carotid arteries in patients with stenoses or occlusions of the common carotid arteries.

Ultrasound features characteristic of nonspecific aortoarteritis:

- 1) thickening of the walls of the common carotid arteries to a maximum of 4.0-4.5 mm,
- 2) the thickness of the walls directly correlates with the severity of inflammation process,
- 3) artery walls - with clear, even contours in the chronic stage of the process,
- 4) vague contours,
- 5) unclear differentiation from the paravasal tissue of the outer layers of the affected arteries indicate ongoing inflammation,
- 6) uneven inclusions confirm the calcification of the arteries, which in turn indicates the antiquity of the present process,

- 7) due to prolonged segmental stenoses, even in patients with hemodynamically insignificant stenoses, all speed indicators increased. In this regard, assessment of the degree of stenosis based on velocity values in non-specific aortoarteritis difficult.

It is extremely important to assess the state of the internal carotid arteries in patients with stenoses or occlusions of the common carotid arteries, because their patency largely determines the feasibility of surgical intervention. Until now, X-ray contrast examination remains the gold standard for non-specific diagnosis aortoarteritis.

According to the nature of the lesion in the subclavian arteries, occlusions prevail (more often in the second and third segments). Nonspecific aortoarteritis is characterized by prolonged lesions of the common carotid arteries. Diffusion constrictions smoothly transition into unchanged areas. Often, the common carotid arteries retain a filiform lumen and the process extends up to the bifurcation, sometimes with an extension to it. However, as a rule, the internal carotid arteries remain patent. A detailed picture of the lesion of the brachiocephalic arteries can be obtained at MRI.

Epidemiology. Previously, it was believed that non-specific aortoarteritis the disease is quite rare and is found only in the countries of the East (Japan, India, China, Korea, etc.), in connection with which the disease is also called an oriental disease. However, the latest data from the literature certainly indicate that it is more extensive spread.

The actual frequency of nonspecific aortoarteritis in different regions varies quite significantly range

It is assumed that the difference in clinical manifestations of non-specific aortoarteritis is due to the peculiarities of climatic conditions affecting the course of the disease. However, in the absence of convincing facts about the cause of the disease and the possibility of a polyetiological nature, it can be explained by various etiological factors.

The etiology of nonspecific aortoarteritis is still unknown. At the initial stages of studying this pathology, a number of versions were expressed regarding its genesis (tuberculosis, syphilis, rheumatism, etc.). They found no confirmation.

There is an innate theory of nonspecific aortoarteritis. A congenital defect of fusion of the embryonic dorsal aorta at the 3rd week of intrauterine development may play a role in the genesis of the disease.

The autoimmune theory of the occurrence and progression of non-specific aortoarteritis, first expressed in 1962 by Judge, has gained more and more supporters.

Connection of nonspecific aortoarteritis with other diseases and hereditary factors. The issue of the influence of hereditary factors on the occurrence and progression of nonspecific aortoarteritis has not yet received its final solution.

Aortoarteritis combined with other diseases in the genesis of which immune disorders are involved are discussed in the literature. Thus, nonspecific aortoarteritis is associated with Crohn's disease, various forms of rheumatoid arthritis, glomerulonephritis, cardiomyopathy, and systemic lupus erythematosus. However, almost all publications refer to individual cases and it is not possible to reveal the true frequency of the combination of these diseases.

Histocompatibility antigens, responsible for the immune response, are associated with the frequency of development of nonspecific aortoarteritis. Despite the presumed genetic predisposition, additional factors are necessary for its occurrence factors.

Autoimmune mechanisms. More than 40 years have passed since the first assumption about the role of immune disorders in the genesis of nonspecific aortoarteritis. What proof of the role of autoimmune aggression in the development of aortic arch syndrome has been obtained by science today?

Back in the mid-1930s, it was established that the first symptoms of autoimmune diseases often appear during pregnancy, which is known to be accompanied by a change in immunological reactivity.

The point of view about the role of immuno-pathological processes in the development of NAA is confirmed by the existing, albeit rare, combination of this pathology with other autoimmune diseases: Crohn's disease, nonspecific ulcerative colitis, systemic lupus erythematosus, rheumatoid arthritis, nodular periarteritis, glomerulonephritis. There are observations of nonspecific aortoarteritis in immunodeficient patients' conditions.

It was established that in 25-41 % of patients, antibodies to cardiolipin are detected, and an increase in the level of antibodies is associated with an occlusive lesion of blood vessels, the development of arterial hypertension, and an aortic valve lesion. In recent years, it has been proven that in 38-95% of cases, antibodies to endothelial cells are present in the sera of patients with non-specific aortoarteritis. At the same time, the frequency and level of their detection is significantly higher than with others diseases involving an autoimmune component (systemic lupus erythematosus, scleroderma, obliterative thromboangiitis).

As a possible mechanism for the occurrence of autoimmune aggression, the authors propose a change in the permeability of the vascular wall under the influence of some factors. Violations of cellular immunity are of particular importance in the pathogenesis of nonspecific aortoarteritis.

Thus, there is no doubt that nonspecific aortoarteritis is accompanied by pronounced immune disorders. However, the effector link of the immune defect remains unclear. Moreover, the fact whether the immune defect is primary or arises in response to acute or subacute inflammation is controversial.

Pathological anatomy. It is established that with non-specific aortoarteritis, the pathological process mainly affects the aorta, as well as large trunks departing from it (arteries of elastic and muscle-elastic types). At the same time, a characteristic feature of the disease is the predominant defeat of the proximal segments departing from the aortic branches, while the distal sections of the affected arteries, as a rule, remain intact. Brachiocephalic arteries and the thoracoabdominal segment of the aorta with renal and visceral arteries are most often involved in the process of nonspecific aortoarteritis. Characteristic involvement in the process of several arterial segments

The pathological process in nonspecific aortoarteritis is quite variable depending on the stage of the inflammatory process. As a result of pronounced inflammatory or autoimmune processes, all elastic structures in the media and adventitia are destroyed, with the subsequent formation of granulomatous, and ultimately fibrous and scar tissues. With the transition of the inflammatory process to the intima of the artery, the latter sharply thickens, scleroses and, thus, causes the

narrowing of the lumen of the affected arteries.

As the disease progresses, granulomatous changes turn into fibrous ones, there is fragmentation and destruction of elastic fibers in the adventitia and media. A similar change occurs with muscle elements. In the end, a chronic stage of the disease occurs, the result of which is severe sclerosis and fibrosis of all layers of the wall of the affected vessel.

In practice, it is quite difficult to clearly distinguish the stages, because with nonspecific aortoarteritis, as a rule, in different segments of the aorta and artery, you can see the development of different phases of the pathological process in separate segments of the artery at the same time.

The pathological physiology of the aortic arch syndrome (nonspecific aortoarteritis) corresponds to diseases characterized by impaired blood circulation in various arterial pools.

Variants of hemodynamic disorders are caused by ischemia of various organs. The principal mechanism of compensation of blood circulation of different basins is arterial hypertension of a systemic nature caused by renovascular, coarctation, cerebroischemic or mixed genesis.

Classification and clinical picture

The most common clinical classification is a classification in which all types of damage are conditionally divided into 4 types:

- I - isolated involvement of arc branches in the process aorta;
- II - a lesion of only the thoracoabdominal segment of the aorta with visceral branches and renal arteries, without involvement of the branches of the aortic arch;
- III or mixed type - a combination of these two options
- IV type of lesion, in which any departments of the aorta can be affected, but with the obligatory involvement of the pulmonary arteries.

The clinical picture of nonspecific aortoarteritis is very diverse and multifaceted, which is due primarily to the mosaic nature of the lesion of certain arterial segments. At the same time, scissors between the clinical symptoms of the disease and morphological changes of the aorta and main arteries are characteristic. The weight of vascular damage is always greater than the clinical manifestations of the disease.

In the vast majority of cases, a diagnosis of non-specific aortoarteritis can be made already at the initial examination of the patient only on the basis of a standard angiological survey, which includes auscultation of vessels and blood pressure measurement in all extremities.

The most common syndromes are lesions of the branchial arch of the aorta, coarctation, renovascular hypertension, and general inflammatory manifestations.

The syndrome of general inflammatory reactions (general, cardiac and pulmonary symptoms) is recorded in the acute period of the disease. During this period, sweating, unmotivated temperature rises, and inflammatory changes of blood, acceleration of ESR, positive reaction to C-reactive protein are noted. Patients often complain of pain along the course of the main arteries. Shortness of breath and palpitations are possible. Cardiac symptoms can also be caused by coronary disease, and pulmonary symptoms - by the presence of damage to the branches of the pulmonary artery. In general, inflammatory reactions are observed

in 2/3 of patients. Often this syndrome precedes the hemodynamically significant stage violations.

The syndrome of renovascular hypertension is observed when the renal arteries are damaged. An isolated lesion of the renal arteries in nonspecific aortoarteritis is rarely observed. More often, the lesion of the renal arteries is combined with the lesion of the branches of the aortic arch or the thoracoabdominal part of the aorta. Renal artery stenosis, as a rule, is located in the mouth or the first segment. Renal artery occlusions are registered much less often.

The main complaints are headache, shortness of breath, pain in the heart, caused by systemic hypertension. Damage to the renal arteries leads to higher arterial hypertension than in coarctation syndrome. For this syndrome, a typical increase in systolic pressure up to 180-250 mm Hg, diastolic pressure - up to 100-160 mm Hg.

With non-specific aortoarteritis, the true blood pressure level can be determined only when measuring it on four limbs. Sometimes, when both subclavian arteries and the thoracoabdominal segment of the aorta are affected, the pressure on the upper extremities may not be determined or may remain normal.

The true level of blood pressure in such patients can be determined only when probing the aorta.

Auscultation is of great importance in diagnosis, in which systolic noise in the epigastric region is diagnosed in almost 80% of cases.

In the diagnosis of this syndrome, ultrasound duplex scanning and angiography are the most informative. Renal artery damage is characterized by a limited area of stenosis, therefore local increased velocity and asymmetry of blood circulation in both renal arteries become important during duplex scanning. MRA reveals a lesion of the aorta and renal arteries, but the picture is usually clearer with an angiographic examination, which allows to specify the nature and length of the lesion. With aortitis of this localization, it is not uncommon that even with occlusion of the renal artery, its distal section remains patent.

The syndrome of chronic abdominal ischemia is, according to the literature, from 5 to 66%. It is quite rarely observed in patients without involvement of the thoracoabdominal aorta or renal arteries in the process.

The rarity of its manifestation is due to good collateral ways of blood circulation compensation.

Most often, blood circulation compensation is carried out at the expense of the lower mesenteric artery through the arc of Riolan. Functional disorders of intestine prevail. Characteristic manifestations are stool instability, unexplained change in diarrhea to constipation. Sometimes patients lose a lot of weight.

Diagnosis is primarily related to auscultation: a rough systolic noise is usually heard under the xiphoid process.

Duplex scanning and two-projection angiography are priority studies.

Mesenteric artery is sharply thickened. Ultrasound can determine the length of the affected segment and assess the degree of artery stenosis, as well as the condition of the distal arterial bed. This is of fundamental importance for choosing the method of surgical intervention.

With X-ray contrast examination, it is possible to visualize stenosis of the abdominal trunk and upper mesenteric artery in the lateral projection. Narrowing, as a rule, is localized in the mouth. When the thoracoabdominal part of the aorta is

damaged, the abdominal trunk and the upper mesenteric artery are the first to be involved in the process, when the process is localized in the infrarenal part of the aorta, the lower and upper mesenteric arteries are mainly affected arteries

It has been established that in none of the other known diseases are the collaterals between visceral arteries so strongly developed as in nonspecific aortoarteritis. The developed arc of Riolan, filled retrogradely during angiographic examination in young patients, is a pathognomonic symptom of nonspecific aortoarteritis. The same pattern of damage can be detected with electron beam or spiral tomography.

Aortic bifurcation lesion syndrome combines infrarenal and brachial artery lesions. The frequency of detection of this syndrome in nonspecific aortoarteritis is 18-20%. At the same time, in 25% of patients, the arteries of the lower extremities below the Poupart's ligament are involved in the process, more often - the femoral arteries.

The main symptoms are signs of ischemia of the lower limbs and pelvic organs: pain in the lower limbs, intermittent lameness, weakness in the lower limbs and their cooling. Often, especially when the aorta is damaged, high intermittent lameness with pain in the hip muscles is observed.

Almost 50% of patients with this syndrome have impaired sexual function, which is more often manifested by erectile dysfunction. It is quite likely that the occlusion of the internal iliac and lumbar arteries plays an important role in the genesis of these symptoms.

The main clinical symptoms are weakening or absence of pulsation, blood pressure on the lower extremities. A systolic noise is heard over the abdominal aorta and femoral arteries.

Diagnosis is carried out taking into account blood pressure measurement, duplex scanning and angiographic examination. The latter has priority in this syndrome. The angiographic picture is characterized by stenosis or occlusion of the infrarenal section of the aorta. At the same time, the femoral and iliac arteries often remain intact or little changed.

Coronary syndrome, or the so-called coronary diseases, occurs in no more than 8-10 % of patients. Most patients with coronary artery disease, according to the world literature, are female (86%) of Japanese nationality.

The main manifestations of coronary artery disease are angina, pain in the heart area, palpitations with a tendency to tachycardia, shortness of breath, and rapidly passing changes on the ECG. The development of a fatal myocardial infarction, which occurs without any warning signs, is not uncommon. Coronary arteritis can lead to myocardial infarction due to stenosis or occlusion of the artery, as well as myocardial ischemia during exercise due to the rigidity of the coronary artery and the impossibility of its adequate dilatation.

In the diagnosis of the syndrome, the following are important:

- ECG with functional tests, myocardial scintigraphy, Holter monitoring;
- coronary angiography.

The diagnosis of coronary artery disease can be suspected on the basis of ECG data.

The main ECG signs of current coronary artery disease are depression of the *ST wave* and inversion of the *T wave*. More than 29% of patients with hemodynamically

significant lesions of the coronary arteries did not show signs of myocardial ischemia on the ECG.

Holter monitoring helps in diagnosis ECG monitoring and myocardial scintigraphy. Coronaritis is associated with aortic regurgitation in almost 30% of cases, and mitral insufficiency in 5%.

Coronary angiography is the gold standard for clarifying the nature, localization and length of the lesion.

Aortic insufficiency syndrome is diagnosed in different regions with quite different frequency. It is rare in Ukraine: in no more than 2-3 % of patients, and in Japan - up to 50%. The development of the syndrome is caused by infiltration and expansion of the ascending part of the aorta. In the future, dilatation of the aortic ring occurs with the occurrence of aortic insufficiency. The clinical picture is characterized by complaints of pain in the area of the heart and behind the sternum, shortness of breath. Diagnosis is based on blood pressure determination and auscultation. The main symptom is a decrease in diastolic pressure. Auscultatively, a diastolic noise is heard over the aorta and in the third-fourth intercostal space to the left of the sternum.

The degree of insufficiency of the valve, data on the size of the ascending aorta are clearly determined with the help of echocardiography.

Angiography with ventriculography is appropriate only in certain cases.

Pulmonary artery disease syndrome was first discovered only in 1973.

Cough, haemoptysis, and pain behind the sternum usually serve as clinical manifestations. At the chronic stage of the process, clinical symptoms may be quite insignificant. Often the only symptom can be a cough mixed with sputum. Cases of asymptomatic lesion of the pulmonary artery in non-specific aortoarteritis are also described. The most frequent lesion of the pulmonary artery is diagnosed in combination with the localization of the process in the branches of the aortic arch or thoracoabdominal aorta.

X-ray examination and lung scanning are of great importance in the diagnosis of the syndrome. The final study is angiogram.

At X-ray research reveals increased transparency of the affected segments when scanning the lungs in the lesion area, vascularization may deteriorate. With the help of angiogram, it is possible to detect stenoses or occlusions of the branches of the pulmonary artery, as well as its deformation.

Aneurysmal syndrome, or aneurysmal expansions, are observed in 7-22% of patients, which in 14% of cases may be the only manifestation of the pathological process.

As a rule, aneurysmal expansions are located in the thoracic (about 60%) and abdominal aortas (40%). Nonspecific aortoarteritis is also characterized by the presence of multiple aneurysms. As a rule, aneurysmal expansions are diffuse in nature, bag-shaped aneurysms are much less common.

The clinical picture consists of symptoms of aneurysm compression of adjacent organs and is determined by the location and size of the aneurysmal protrusion, but these symptoms may not be expressed.

When the process is localized in the abdominal aorta, as a rule, there is a concomitant lesion of the renal arteries with the development of arterial

hypertension syndrome. Isolated aneurysm development without a stenotic process in other basins is diagnosed extremely rarely, with the exception of the South African region.

Diagnosis is based on auscultation and palpation. As a rule, a systolic noise is heard above the aneurysmal expansion, and when the process is localized in the abdominal aorta, its expansion can be detected by palpation.

Ultrasound, computed tomography, MRI and angiographic examination play an important role in the diagnosis of the lesion. Ultrasound scan, transoesophageal echography is the simplest type of diagnosis. CT and MRI allow not only to estimate the size and length of aneurysmal expansion of the aorta, but also to estimate the degree of calcification vessels.

Angiography should be performed when the process is localized in the thoracic aorta. To solve the question about the state of visceral arteries at deviations from aneurysmal expansion, it is advisable to perform the study in projections.

Differential diagnosis

Differential diagnosis of NAA should first of all be carried out with atherosclerotic lesions of the aorta, since only these diseases cause multiple lesions of different segments of the aorta and main arteries.

Nonspecific aortoarteritis mainly affects women aged 20 to 40. Atherosclerotic lesions are usually observed in patients older than 40 years, but it is possible in people younger than 40 years. Differential diagnosis is based on the features of the clinical course, the time of onset of the disease, the typical localization of the process, and the results of instrumental research methods.

A lesion of the brachiocephalic arteries caused by nonspecific aortoarteritis usually occurs in young women under 30 years of age, the process is located in the second and third segments of the subclavian arteries, in the common carotid arteries. In men over 40 years old, with atherosclerosis, the process is most often localized in the area of bifurcation of carotid arteries, common carotid arteries are rarely stenosed. Subclavian arteries, as a rule, have a lesion in the first segment, and very rarely the process is localized in the distal segments.

In non-specific aortoarteritis, the thoracoabdominal part of the aorta is often involved in the process; in atherosclerosis, this segment of the aorta is extremely rarely damaged.

In aortoarteritis, isolated lesions of the iliac and femoral arteries are rarely diagnosed. The infrarenal part of the aorta, iliac and femoral arteries are favourite localizations of the atherosclerotic process.

In non-specific aortoarteritis in young patients, arterial hypertension is often recorded, in atherosclerosis in young patients it is rare.

In patients with nonspecific aortoarteritis, as a rule, pronounced calcification of the affected segments of the aorta, in young patients with atherosclerosis, calcification is rarely observed.

During ultrasound echolocation in nonspecific aortoarteritis, the wall of the affected segments of the aorta and arteries is thickened, peri-aortic inflammation and fibrosis are often present, in atherosclerosis, the intima-media complex is thickened, but the adventitia and peri-aortic space are usually intact.

On angiograms in patients with nonspecific aortoarteritis, prolonged segmental narrowing is observed, which smoothly transition into unchanged areas,

in atherosclerosis, usual local narrowing with uneven and sometimes eaten edges.

Another disease with which nonspecific aortoarteritis sometimes has to be differentiated is obliterating Thromboangiitis. The disease affects small and medium-sized arteries. In contrast to non-specific aortoarteritis, mostly men are affected. As a rule, with obliterating Thromboangiitis affects the arteries of the lower extremities, sometimes the distal segments of the arteries of the upper extremities are involved in the process, much less often - the infrarenal segment of the aorta. Visceral branches and branches of the aortic arch are involved in the process casuistically rarely.

Sometimes the differential diagnosis is difficult between non-specific aortoarteritis thoracoabdominal aorta and hypoplasia of the aorta. The clinical picture of both diseases may be similar, as in non-specific aortoarteritis in thoracoabdominal localization, inflammation activity may be low. Often, only with the help of an X-ray contrast study, it is possible to reveal the true genesis of an occlusive lesion. It should be remembered that visceral and renal arteries are almost never involved in the process of hypoplasia of the aorta. The second important differential diagnostic feature is the thickness of the aortic wall. Computed tomography and ultrasound transoesophageal echolocation play an extremely important role in the difficulties of differential diagnosis of stenosing lesions caused by nonspecific aortoarteritis and hypoplasia of the aorta. The thickness of the aortic wall in nonspecific aortoarteritis is always greater than in patients with hypoplasia of the descending aorta, in whom the wall thickness does not exceed the norm. In addition, patients with hypoplasia of the aorta practically never have calcification of the aorta.

Renal and internal carotid arteries can be affected due to fibromyalgia dysplasia, in which, however, the aorta itself is never affected. Fibromuscular in the carotid basin dysplasia is localized in the internal carotid artery without involving the common carotid arteries in the process, which is practically not observed in nonspecific aortoarteritis.

A detailed examination of patients in the vast majority of cases makes it possible to make the correct diagnosis.

Drug treatment

Treatment of nonspecific aortoarteritis is a complex task. In case of inadequate treatment, the process turns into protracted subacute stage and contributes to progression disease, what fixed in 88% of patients. In more than 61% of cases, lesions develop in previously intact places. Even when clinical remission is achieved, the appearance of new lesions is quite likely. Approximately 20% of patients suffering from aortic arch syndrome do not have relapses of inflammation and do not need any anti-inflammatory immunosuppressive therapy. As shown by clinical observations, the appointment of non-steroidal drugs, antibiotics and anticoagulants of direct action is ineffective in the treatment of the activity of the process of non-specific aortoarteritis. The most typical and generally accepted scheme of treatment of the acute and subacute stage is therapy with small doses (according to 30-60 mg per day) of corticosteroids in combination with antiplatelet therapy. The frequency of effectiveness of such therapy ranges from 25 to 72%

glucocorticoids has gained particular popularity. Although the combination of corticosteroids with cytostatic is an important factor in improving the long-term results and survival of patients with systemic vasculitis, it has side effects and increases the risk of developing hospital-induced diseases.

According to the literature, at this time preference is given to methotrexate, as the least toxic drug that causes fewer complications.

Plasmapheresis and chemosorption were used, but their effectiveness in terms of achieving stable and long-term remission is controversial.

Another quite priority area is specific plasmapheresis. The method is based on the removal of plasma (1200-1600 ml) from the bloodstream and/or its passing through a sorbent of imotin with a total trypsin-binding capacity of 20 mg.

Surgical treatment

The question of choosing a conservative or surgical method of treatment, as well as the methods of conservative treatment themselves, is still the subject of disputes by domestic and foreign authors. At the level of modern knowledge, these methods should not be alternatives to each other.

The following are extremely important position:

- patients with non-specific aortoarteritis should be operated on after the acute and subacute stages have subsided inflammation;
- the method of choice in the pre- and postoperative periods of anti-inflammatory therapy is pulse therapy with cyclophosphane and methylprednisolone;
- it is preferable to perform the operation after 3-4 months. after the completion of the effective pulse therapy.

The first successful operations on the branches of the aortic arch were performed in the early 50s. However, most vascular centres even today have at their disposal the experience of only single interventions on the branches of the aortic arch in NAA.

Operative treatment of the brachiocephalic branches of the aortic arch is indicated for:

- transient disorders of cerebral blood circulation in the presence of hemodynamically significant one's defeats;
- hemodynamically significant stenoses or occlusions of the common carotid arteries;
- Vertebral - subclavian steal syndrome with clinical manifestations;
- sub compensated and decompensated upper ischemia limbs

The question of intervention in the asymptomatic course of the disease has not yet been finally resolved. In contrast to atherosclerosis, in nonspecific aortoarteritis there is no mechanism of embolization from a disintegrating plaque.

Surgical intervention is indicated for patients with multiple lesions of the aortic arch, with occlusions and hemodynamically significant stenoses of the common carotid arteries, even with an asymptomatic course of the disease.

Today, most clinicians prefer less traumatic extra thoracic interventions. To perform extra thoracic interventions, require a suitable, i.e. intact, section of the donor artery, which can be the common carotid artery in case of carotid-subclavian

shunting or the subclavian artery in case of subclavian - carotid prosthetics. Unfortunately, this is not always possible with the multiple nature of the defeat, which often simply does not allow performing a simpler intervention.

Aortobicarotid operations are accompanied by the largest number of complications, primarily the occurrence of a haemorrhagic stroke caused by a mismatch of blood inflow and outflow and an increase in blood pressure observed in the first hours and days after surgery.

The use of balloon angioplasty in patients with NAA is questionable.

Performing simultaneous operations on extracranial and renal arteries is not justified, because the severity of the patients' condition, traumatic single-moment reconstructions sharply increase the risk of developing intra- and postoperative complications. Expedient phased reconstructions.

The sequence of correction of one or another arterial pool is decided taking into account the hypotensive test, which allows to assess the tolerance of the brain to artificially created moderate hypotension.

Dispensary observation

It should be remembered that patients with non-specific aortoarteritis require dispensary observation at least once a year, which includes:

- activity definition process;
- duplex scanning not only of reconstructed vessels, but also of other arterial basins for timely detection of new one's defeats;
- anti-inflammatory cursi pulse therapy or supportive therapy with methotrexate when determining high activity process

5. Plan and organizational structure occupation

№	Main stages of the classes, their function and content	Learning objectives in mastery levels	Control and training methods	Methodological support materials	Time min
1	2	3	4	5	6
Preparatory stage					
1.	Organization of the lesson				5 min
2.	Setting educational goals and motivation of the topic				10 min
3.	CONTROL of the initial level of knowledge, skills, abilities.				
	1. Etiopathogenesis atherosclerosis of vessels.	II	Level II methods		

	2. Anatomy and physiology of aorta and main vessels 3. Clinical picture of a patient with chronic limb ischemia 4. Algorithm of examining patients for pathology vessels 5. Differential diagnosis of atherosclerosis and endarteritis. 6. Interpretation of examination data - blood tests - general and coagulogram, ultrasound, X-ray	II II II II III	1. Individual oral survey. 2. Written theoretical survey. 3. Solving typical problems 1. Solving atypical situational problems	Problems of the II level Tables, Slides. Video recordings Equipment, Radiographs and Medical history	60
The main stage					
4.	Formation of professional skills and abilities				
	1. Master the methods of objective examination of patients with chronic ischemia upper limbs	III	Method of formation of practical training skills	Educational equipment - orientation maps	30 min.
1	2	3	4	5	6
	2. Curate a patient with atherosclerosis of vessels. 3. Participate in duplex vascular scanning and angiography 4. Participation in surgical intervention on carotid arteries vessels	III III III	Method of formation of skills: a) Professional training in solving non-typical problems b) laboratory-experimental decisions tasks	Unusual problems in form: patient, case histories, test situational tasks, business games, dressing	

5. Bandage the patient in the postoperative room		III			
The final stage					
5.	Control and correction of the level of professional skills and abilities	III	Control method: Individual control of practical skills	Equipment	60
6.	Summarizing the results of the class				3 min.
7.	Homework, study literature on the topic			Oriented map of independent work with literature	2 min.

6. Materials on methodical provision of classes.

6.1. Control materials for the preparatory stage occupation.

Questions

1. General classification of vascular diseases by etiological feature.
2. The main clinical signs of acute and chronic ischemia limbs.
3. Acute brain disorder of blood circulation.
4. Differential diagnosis of atherosclerosis and endarteritis.
5. Methodology of vascular examination (duplex scanning, angiography).
6. Tactics of the surgeon and features of the examination of a patient with vascular pathology.
7. Indications for conservative treatment of chronic limb ischemia.
8. Indications for surgical interventions for signs of cerebral ischemia or limb ischemia.
9. Methods of operative interventions on vessels.

Situational tasks

1. Patient A., 60 years old, was hospitalized in the surgical department with complaints of weakness, pain in the upper limb, impaired active movement, and sensitivity in the fingers of the same limb. What can be the cause of complaints?
Answer standard: thrombosis of the main artery of the limb.
2. Patient Yu., 54 years old, complained of repeated short-term dizziness, weakness, hearing impairment. On auscultation, there is a systolic noise in the projection of the right carotid artery. Which is the most likely diagnosis?
Answer standard: Stenosis of the right carotid artery with transit ischemic attacks.
3. Patient M., 30 years old, complained of increased sensitivity of the fingers of the upper and lower extremities to cold, a change in their colour, and periodic pain at rest. He has been ill for about 3 years after hypothermia in the water.

Intermittent lameness 300 m. What is the diagnosis?

Answer standard: Endarteritis, CHI 2.

6.2. Materials for methodical support of the main stage occupation

1. What is endarterectomy?

- A. Stitching arteries
- B. Resection of part of the artery.
- C. Removal of the middle and upper layer of the artery.
- D. Bypass artery.
- E. Removal of atherosclerotic intima and media

Answer standard: D.

2. Patient A., 35 years old, was brought to the surgical hospital in a serious condition with signs of ischemia of the upper limbs, trophic disorders on the fingers, weakening of pulsation in the carotid arteries, absence of pulsation in the arteries of the upper limbs. Active movements and sensitivity in the fingers are present. He has been sick for about a year. The most important test to determine the diagnosis?

- A. Laboratory.
- B. Overview X-ray of chest organs.
- C. Auscultation, ECG
- D. Ultrasound examination.
- E. Duplex scanning of vessels.

Answer standard: D.

3. Which etiological theory of non-specific aortoarteritis is the most proven?

- A. Infectious
- B. Allergic.
- C. Immunological
- D. Rheumatology.

Answer standard: C.

Differential diagnosis of NAA with atherosclerotic lesions of vessels

Sign	NAA	Atherosclerosis
Age of the patient	from 20 to 40 years	over 40 years old, but maybe in people younger than 40 years old

Damage to the brachiocephalic arteries	usually occurs in young women under 30 years old, the process is located in the second and third segments of the subclavian arteries, in the common carotid arteries	In men over 40 years old, the process is most often localized in the area of bifurcation of carotid arteries, common carotid arteries are rarely stenosed. Subclavian arteries, as a rule, have a lesion in the first segment, and very rarely the process localized in the distal segments
Thoracoabdominal section of the aorta	often involved in the process	this segment aorta is extremely damaged rarely
Isolated defeat of the iliac and femoral joints arteries	rarely diagnosed	favourite localizations
Arterial hypertension	in young patients often fixed	in young patients it is rare.
Pronounced calcinosis of the affected segments of the aorta	often	in young patients with atherosclerosis calcinosis is rarely observed
With ultrasound echolocation:	wall affected segments of the aorta and	thickened complex intima-media, however

	the artery is thickened, often peri-aortic inflammation and fibrosis	adventitia and peri-aortic space, as a rule, intact
On angiograms:	are observed prolonged segmental narrowing, which smoothly transition into unchanged areas	ordinary local narrowing with unequal, a sometimes with worn edges

Diagnostic criteria for NAA (aortic arch syndrome):

(Arned et al.: The American College of Rheumatology 1990 Criteria for the classification of Takayasu Arteritis, *Arthritis Rheum.* 33:1129-1134, 1990.)

1. Onset of the disease before the age of 40 years.
2. Intermittent lameness (muscle complaints when moving, at least in one limb, especially in hands).
3. Weakened radial and/or ulnar pulsation arteries.
4. The difference in systolic pressure on both arms is more than 10 mm Hg.
5. Vascular noises over the subclavian artery during auscultation (from one or on both sides) or above the abdomen aorta.
6. Angiographic detection of typical vascular changes of the aorta, its branches or larger arteries in the proximal parts of the upper or lower extremities (most often focal-segmental, stenotic or occlusive changes), which cannot be explained by atherosclerosis or fibromuscular dysplasia or the like reasons.

Probable NAA is indicated by the presence of at least three of these six criteria.

M. Takayasu (M. Takayasu) is a Japanese ophthalmologist who in 1908 described changes in the fundus and types of arteriovenous anastomoses and microaneurysms in a 21-year-old patient ; in subsequent reports, it is noted that patients with similar changes in the fundus often do not have a pulse.

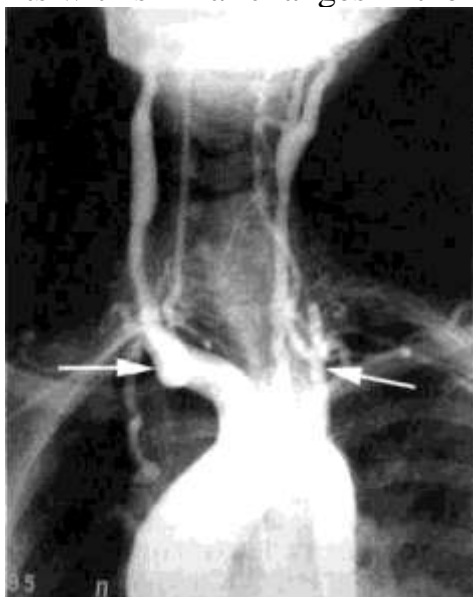


Fig . 1. Angiogram of patient M., 24 years old. Numerical lesions of the branches of the aortic arch. Arrows indicate occlusion of the left and right subclavian arteries.



Fig. 2. Macro preparation parts of the ascending aorta arch ; brachiocephalic occlusion trunk, left carotid and right subclavian arteries, pronounced thickening and swelling of the ascending aorta.



Fig. 3. Macropreparation thoracic aorta (transverse section). The lumen of the aorta passes the match head.

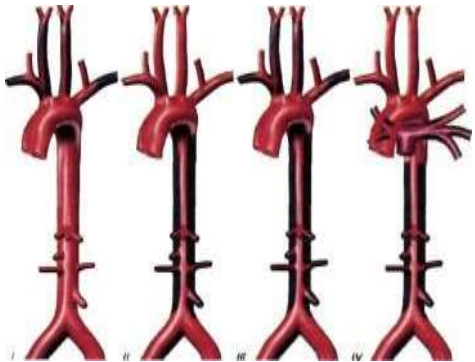


Fig. 4. Types of lesions of the aorta according to classification of E.Lupi-Herrera and others (1977)



Fig 5. Magnetic resonance angiography with contrast. Arrows indicate stenoses of both renal arteries.



Fig . 6 . Compensation of blood circulation in the case of occlusion of the abdominal trunk and superior mesenteric artery is carried out due to the developed arc of Riolan (indicated by an arrow), functioning in the cranial direction.

6.3. Control materials for the final stage occupation.

Situational tasks

1. Identify the diseases (column 1) corresponding to the location of the lesion (column 2).

- | | |
|------------------------|---|
| 1. Syndrome Lerisha . | a) maxillary artery; |
| 2. Prune Belly. | b) arch of aorta and its branches; |
| 3. Syndrome Takayasu . | c) arteries of the base of the skull; d) intestinal vessels; d) bifurcation of the aorta. |

Answer standard: 1-d, 2-d, 3-b.

2. Aortic arch syndrome is not characterized by: a) disappearance of the pulse on the upper extremities; b) disappearance of the pulse on the lower extremities; c) writing spasm; d) dizziness and headaches; e) deterioration of vision up to complete blindness

Answer standard : b, c.

3. A 24-year-old patient complains of pain in her hands, a feeling of numbness, coldness in her forearms and hands at rest. In the last week, the patient's condition has worsened. During the examination: the skin of the upper extremities is of normal colour, but when the fingers are squeezed and spread for 1 minute, they are pale, cold, and pain appears in the fingers. Active movements in the joints in full, there is no sensitivity disorder. Pulsation of the brachial artery is reduced in the distal third of the shoulder, it is vaguely determined in the radial and ulnar arteries. What disease does the patient have? What treatment is indicated for the patient? What treatment should be recommended to the patient to prevent repeated similar ones diseases?

Answer standard : Nonspecific aortoarteritis (aortic arch syndrome) with damage to the arteries of the upper extremities. The patient is shown conservative treatment: therapy with small doses (30-60 mg per day) of corticosteroids in combination with antiplatelet therapy.

• **What is the main symptom of aortic arch syndrome?**

A. Dizziness and weakness.

B. Abdominal pain.

C. Acute chest pain.

D. Violation of urination.

• **What symptom can indicate a violation of blood supply to the upper extremities in aortic arch syndrome?**

A. Violation of pulsation on the wrists.

B. Chest pain.

C. Edema of the lower extremities.

D. Pain in the neck.

• **What disease should be ruled out if aortic arch syndrome is suspected?**

A. Myocardial infarction.

B. Stenosis of the arteries of the lower extremities.

C. Acute pancreatitis.

D. Aneurysm of the abdominal aorta.

• **Which of the diagnostic methods is the most informative for suspected aortic arch syndrome?**

A. Magnetic resonance angiography (MRA).

B. Electrocardiogram (ECG).

C. Chest X-ray.

D. Ultrasound of abdominal organs.

• **What is the main surgical method used to restore blood flow in aortic arch syndrome?**

A. Stenting or shunting of affected arteries.

B. Aortocoronary bypass.

C. Removal of blood clots.

D. Arterial resection.

• **Which of the listed complications is most likely in aortic arch syndrome?**

A. Acute stroke.

B. Hepatitis.

C. Jade.

D. Gastric ulcer disease.

• **What is the most important preventive measure to prevent complications in aortic arch syndrome?**

A. Maintenance of normal blood pressure.

B. Regular intake of antibiotics.

C. Limitation of physical activity.

D. Drinking a lot of water.

• **Which of the following is an effective method of preventing the progression of aortic arch syndrome?**

A. Cholesterol control.

B. Regular physical activity.

C. Taking hormonal drugs.

D. Limiting carbohydrates in the diet.

• **What process is the main cause of aortic arch syndrome?**

A. Atherosclerosis.

B. Viral infection.

C. Chest trauma.

D. Lack of vitamins.

• **Which of the listed measures is mandatory after surgery for aortic arch syndrome?**

A. Appointment of anticoagulants.

B. Taking antibiotics.

C. Intensive physical rehabilitation.

D. Administration of hormonal drugs.

Literature:

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