A rare case of bilateral arteriovenous malformation of the lungs

Arteriovenous malformation of the lungs is a disease characterized by a pathological «direct» connection of the vessels of the pulmonary artery system with the pulmonary veins and the formation of a right-left intrapulmonary shunt. The incidence of arteriovenous malformation of the lungs (AVML) is 2—3 cases per 100,000 population, 10 % cases are diagnosed in children. The degree of manifestation of symptoms in arteriovenous malformation of the lungs directly depends on the vessels diameter at the level of which blood shunting occurs.

The disease is characterized by a typical triad of clinical manifestations: cyanosis (SaO₂ < 85 %), polycythemia (Hb > 185 g/l, Ht > 52 %, RBC > 5.1 · 10¹²/l), changes in the end phalanges of the fingers in the form of «drumsticks». The most severe complications of arteriovenous malformation of the lungs are hemothysis (pulmonary bleeding), hemothorax, abscess and cerebral infarction.

The basis for the diagnosis of the disease are radiological methods, in particular, helical computed angiography of the pulmonary vessels. Treatment of arteriovenous malformation of the lungs can be carried out by embolization of the arterial vessels of the site of the malformation by resection interventions on the lungs or a combination of the two methods described above.

Over the past 32 years, in the clinic of thoracic surgery and invasive diagnostic methods of the SI «National Institute of Phthisiology and Pulmonology named after F.G. Yanovsky NAMS of Ukraine» 20 were treated with a diagnosis of arteriovenous malformation of the lungs. In the literature, there are very few reports on the treatment of bilateral arteriovenous malformation of the lungs, so we would like to give a clinical example of just such a case.

The article presents a clinical example of bilateral arteriovenous malformation of the lungs.

Keywords
Arteriovenous malformation of the lungs, computer angiography, surgical treatment.
individuals with AVML will have HHT. On the contrary, approximately from 15 to 35 % of individuals with HHT will have AVML.

More often, 80 % of cases have primary (congenital) AVML. 47—80 % of AVML is a manifestation of Osler—Randu disease — hereditary hemorrhagic telangiectasia. Congenital AVML has a genetic link, so it can be passed from parents to children. Secondary AVMLs develop with liver cirrhosis (hepato-pulmonary syndrome), metastatic carcinomas, congenital heart defects, injuries, infections (actinomycosis), etc. [1, 2].

The degree of symptoms manifestation in AVML directly depends on the vessels diameter at the level of which blood shunting occurs. Thus, if AVML is localized between the segmental vessels of the lungs, then the clinical manifestations of the disease will be apparent, if the pathological connection is between subsegmental vessels or vessels of an even smaller caliber, then in such cases the pathology may be asymptomatic for a long time [1, 4].

According to clinical manifestations, patients with AVML can be conditionally divided into 2 age groups. In some patients, a bright clinical picture of the disease appears immediately after birth or at a young age [7, 9, 10]. In others, it occurs during puberty and full clinical symptoms develop as a rule at the age of 20—30 and sometimes later [5].

The most serious complications of AVML are hemoptysis (pulmonary bleeding), hemothorax, abscess, and cerebral infarction. The first two complications arise as a result of rupture or telangiectasia or directly the rapture of the vessel that shunts blood from the artery to the vein. Cerebral abscess and infarction occur as a result of thrombosis (with possible infection) of expanded shunt vessels with thinned walls, detachment of the thrombus under the influence of high pressure and its introduction into the pool of cerebral arteries [2].

Diagnosis of AVML includes the study of clinical manifestations of the disease, data of general clinical examination methods, and radiological research methods occupy a special place in this pathology. The most typical complaints in AVML are shortness of breath, rapid fatigue. In more than half of cases AVML is diagnosed only with a complicated course.

X-rays and x-rays of the chest make it possible to detect a fairly clearly contoured homogeneous shadow in the lungs of different shapes and sizes, which sometimes pulsates [3, 10]. Echocardiography is highly informative, especially with its transesophageal modification [7, 9, 10]. Sometimes dilation of the left heart can be determined [3]. Angiopulmonaryography is an absolutely proven method of X-ray diagnosis of AVML, especially with multiple lesions, and its modification in combination with magnetic resonance imaging has a resolution to vessels of the 5th order [6, 8]. Some researchers use Tc-99m MAA scanning to diagnose AVML [5].

Spiral computer angiography of pulmonary vessels is particularly accurate, which allows obtaining a color 3D image of the mosaic type of blood flow in pathologically changed vessels [8, 10].

Treatment of AVML can be carried out by embolization of the arterial vessels of the malformation area, by resection interventions on the lungs or a combination of two methods described above. The goal of the embolization method of therapy is to occlude all blood vessels supplying AVML. For embolization, either Gianturco springs (their diameter exceeds the diameter of the vessel by 1 mm) in combination with histoacryl as an embolizing substance or inflatable balloons (their diameter 2 times exceeds the diameter of the vessel) are most often used. Currently, no clear indications for vascular embolization in AVML have been developed in the world. Each clinic develops its own indications and contraindications for such an intervention. Some authors express the opinion that arteries with a diameter more than 3 mm should be embolized, although usually the diameter of the supplying arteries is between 4 and 10 mm. During long-term observation of patients treated with this method, a stable therapeutic effect is observed in 84 % of cases [4]. So, K. Matsuura et al. [6] reports the effective use of this method of treatment in 8 (57.1 %) patients out of 14. S. Hirota et al gives similar data [3]: excellent results were obtained in all 7 cases in the immediate postoperative period. However, after 1 year, 2 patients with giant AVMLs died due to bleeding from the contralateral lung and progressive pulmonary fibrosis. It is noted in the literature that 8.0 % of patients had intraoperative complications due to the migration of an embolus, namely: transient ischemic attack, chest pain attack, occlusion of the artery of the great circle of blood circulation [5].

Based on the mentioned above, many authors believe that contraindications for obturation therapy are the presence of giant AVFs, a tendency to thrombus formation, the presence of a wide draining vein as well as cases of the draining vein falling directly into the left atrium [4, 5].

Most authors are in favor of surgical resection of the lung in AVML, which allows not only to cure the patient radically but also to avoid complications of the obturation method of closing the shunt. The volume of the operation depends on the volume of the impression of lung tissue and can range from marginal resection of the segment to pulmonectomy. Surgical intervention is especially important for giant shunts, multiple AVMLs, and when the draining vein falls directly into the left atrium [7, 9].
In the presence of bilateral lung pathology M. Ogino et al. [8] recommends staged resections. Over the past 32 years, the clinic of thoracic surgery and invasive diagnostic methods of the National Institute of Phthisiology and Pulmonology named after F.G. Yanovsky NAMN of Ukraine treated 20 patients diagnosed with AVML [1]. There are very few reports on the treatment of bilateral AVML, in the literature so we would like to give a clinical example of just such a case.

Clinical case. Patient P., 17 years old, came to the institute with complaints of weakness, rapid fatigue and shortness of breath during moderate physical exertion. Such complaints had been bothering her for a long time, but no special attention was paid to it. The patient’s mother noted that the child was lethargic, ignorant and is not interested in physical education. However, in general, the girl was satisfactorily fat, with sufficiently developed musculature, but when communicating with her, one got the impression that she was sleepy and somewhat apathetic towards her surroundings. The patient’s mother also drew our attention to the fact that her daughter’s oxygen saturation is low; having been measured, it was 85–88%.

With such complaints, they first turned to their family doctor, who prescribed a chest X-ray examination, after which the patient was recommended to undergo a chest CT scan.

At the time of hospitalization, the patient did not have an X-ray, but it became clear from the description that we are talking about a bilateral infiltrative process in the lungs, possibly of a malignant nature. Therefore, the patient was performed chest CT scan with intravenous reinforcement already at the stage of hospitalization (Fig. 1).

According to chest CT scan AVML was diagnosed with a bilateral lesion (lower lobe on the left and middle lobe on the right).

During the examination, the patient had an elevated hemoglobin level in the blood (160 g/l), while all other indicators were normal. On the first stage the girl was performed a video-assisted (VATS) lower lobectomy on the left. The operative intervention was performed without special features. The postoperative period went smoothly. It should be noted that the patient’s oxygen saturation was 90% and no additional oxygen therapy was necessary.
noted that already on the first day after the operation, oxygen saturation increased to 95% and remained so until the girl was discharged from the hospital. The patient was discharged from the hospital in satisfactory condition after 8 days. The stitches were removed after 14 days. The patient’s mother was informed about the need for surgical intervention on the right lung (it was recommended to perform a VATS middle lobectomy on the right in 2—4 months).

Fig. 2 and 3 presents the appearance of a lung with an arteriovenous malformation.

Fig. 4 presents a macro-preparation of the removed part of the lung in section. Aneurysmatic dilatations of vessels in the type of cavernous bodies are clearly visible in the image.

The patient returned to the institute only 1 year later. CT scan was performed repeatedly with intravenous reinforcement (Fig. 5), where it was established that the left lung is expanded, complete-
ly displacing the left hemithorax, without focal infiltrative changes; the area of malformation remained in the middle lobe of the right lung. It should be noted that the level of hemoglobin in the peripheral blood decreased to 125 g/L. After standard examination, the patient underwent VATS middle lobectomy on the right. The operation and the postoperative period passed without complications. The girl was in satisfactory condition and was discharged from the hospital 7 days later. Oxygen saturation in peripheral blood at discharge was 98%.

Fig. 6 presents the X-ray examination upon discharge. X-ray everything is normal.

Conclusions
AVML is a rare pathology, the timely diagnosis of which allows for radical treatment before the appearance of complications.

References
ВИПАДОК З ПРАКТИКИ

ріовенозної мальформації легень є кровохаркання (легенева кровотеча), гемоторакс, абсцес та інфаркт мозку.

Основою діагностики захворювання є рентгенологічні методи, зокрема спіральна комп’ютерна ангиографія легеневих судин. Лікування артеріовенозної мальформації легень може здійснюватись шляхом емболізації артеріальних судин ділянки мальформації, шляхом резекційних втручань на легені або комбінації двох описаних вище методів.

За останніх 32 роки в клініці торакальної хірургії і інвазивних методів діагностики ДУ «Національний інститут фтизіатрії і пульмонології імені Ф.Г. Яновського НАМН України» перебувало на лікуванні 20 пацієнтів з діагнозом артеріовенозна мальформація легень. У літературі дуже мало повідомлень про лікування двобічної артеріовенозної мальформації легень, тому хочемо привести клінічний приклад саме такого випадку.

У статті наведено клінічний приклад двобічної артеріовенозної мальформації легень.

Ключові слова: артеріовенозна мальформація легень, комп’ютерна ангиографія, хірургічне лікування.

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