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MYOCARDIAL BRIDGE AND CORONARY ARTERY FISTULAS IN A PATIENT WITH ANGINA

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Cardiovascular diseases (CVDs) are the leading cause of mortality among the population. At the core of the progression of the coronary heart disease is the atherosclerosis of the coronary arteries, which is found in majority of patients suffering from angina and in patients with myocardial infarction. However, in some cases, coronary angiography reveals, that patients with the mentioned clinical manifestations have their coronary arteries unchanged. This is treated as syndrome X or microvascular angina. Along with that, development or aggravation of the coronary heart disease may be based on the congenital peculiarities in the coronary arteries location and structure, such as muscular bridges and fistulas of the coronary artery. This is confirmed by a number of studies, which indicate the role of the above mentioned pathologies in the occurrence of angina and myocardial infarction. Nevertheless, there is also the opposite view, which is supported by a number of specialists. According to them, the presence of the mentioned peculiarities in the structure of the coronary channel is deemed as the patient-specific norm. Hence, the issue of the surgical treatment of the patients with the aforementioned coronary arteries anomalies remains controversial. The clinical case report of the patient with the symptoms of angina pectoris, in which the coronary angiography did not reveal the stenosis of the coronaries arteries, but located the myocardial bridge and the coronary fistula. The role of the congenital coronary vessels pathology in the angina pectoris is analyzed. The diagnosis guidelines and the tactics of the conservative and surgical treatment of patients with the above mentioned syndromes are discussed.

Keywords: myocardial bridging; coronary artery fistulas; congenital anomalies of coronary arteries; coronary spasm; fractional flow reserve.

МЫШЕЧНЫЙ МОСТИК И ФИСТУЛА КОРОНАРНОЙ АРТЕРИИ У БОЛЬНОЙ СО СТЕНОКАРДИЕЙ

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Сердечно-сосудистые заболевания являются ведущей причиной смерти. В основе развития ишемической болезни сердца лежит атеросклероз коронарных артерий, который обнаруживают у большинства больных, страдающих стенокардией, и у пациентов с инфарктом миокарда. Однако в ряде случаев у лиц с этими клиническими проявлениями коронарные артерии при ангиографии оказываются неизмененными. Это состояние обозначают как синдром X, или микроваскулярную стенокардию. Наряду с этим в основе развития или усугубления течения ишемической болезни сердца могут лежать врожденные особенности расположения и строения коронарных артерий, к которым относят мышечные мостики и фистулы коронарной артерии, что подтверждено рядом исследований, в которых указывается на роль этих патологий в возникновении стенокардии и инфаркта миокарда. Однако существует и противоположное мнение — часть специалистов признает наличие данных врожденных особенностей строения коронарного русла индивидуальным вариантом нормы. В связи с чем в настоящее время остается спорным вопрос хирургического лечения больных с вышеуказанными аномалиями венечных артерий. В статье приведено описание больной со стенокардией, у которой при коронарной ангиографии не было обнаружено признаков стенозирования коронарных артерий, но выявлены мышечный мостик и фистула коронарной артерии; проанализирован вклад врожденной патологии венечных сосудов в развитие стенокардии напряжения, а также обсуждаются диагностика, тактика консервативного и хирургического лечения больных с данными аномалиями.

Ключевые слова: миокардиальный мостик; коронарная фистула; врожденные аномалии коронарных артерий; коронарный спазм; фракционный резерв кровотока.

Cardiovascular diseases, primarily coronary heart disease, and brain disease are the leading causes of premature mortality and disability in people of various ages in industrialized countries. The pathomorphological basis of coronary heart disease is coronary atherosclerosis, which is found in most patients

with angina pectoris and in patients with myocardial infarction. However, in approximately 5%–10% of patients with these clinical manifestations, angiography shows unchanged coronary arteries [2]. This condition is referred to as Syndrome X, or microvascular angina, and it indicates the role of endothelial dysfunction in such cases [7].

Additionally, location and structure congenital anomalies of the coronary arteries can lead to the development of myocardial ischemia. Normally, the main coronary arteries are located on the surface of the epicardium. In some cases, a short section of the artery is immersed in the myocardium (artery "tunneling"), which is called a muscle bridge. Muscle bridges occur in 5%–12% of patients and are more likely to be presented in the left anterior descending artery [9]. The submerged segment of the artery has a normal diameter in diastole, but the diameter tapers sharply during each systole. The coronary arteries are supplied with blood during diastole, that is, it would seem that the muscle bridges do not contribute to the limitation of blood flow to the coronary arteries. However, deep and long muscle bridges can restrict coronary blood flow into the diastole as well. This is due to a number of factors, such as impaired diastolic function i. e., myocardial hypertrophy, endothelial dysfunction and its associated risk factors (arterial hypertension, dyslipidemia, smoking), and coronary artery spasm. In these cases, muscle bridges are associated with angina pectoris, myocardial infarction, rhythm disturbances, and sudden death [5].

Another anomaly of the coronary arteries is fistula i. e., the artery communicates with the heart chambers or large vessels, including vena cava and pulmonary artery. Communication with the left ventricle is often asymptomatic, but significantly large shunts are accompanied by an increase in pressure in the left ventricle, as well as by hypertrophy and dilatation of the left ventricle. In some patients, myocardial ischemia distal to the fistula is observed [12].

Treatment for patients with angina pectoris associated with a myocardial bridge has been described in several medical reference books. A promising research method used to objectify the role of a myocardial bridge in the development of pain is the assessment of the fractional flow reserve (FFR) by coronary angiography. This method is used to assess the risk of myocardial ischemia. The fractional reserve of blood flow is defined as the maximum blood flow in the myocardium in the presence of stenosis divided by the theoretical maximum blood flow in the absence of stenosis. A measurement of $FFR < 0.75$ is considered to be hemodynamically significant. A method in which conductors measure pressure (a pressure sensor mounted in a wire conductor) and a Doppler method for estimating blood flow velocity using spectral analysis are the two methods in which information about hemodynamics based on the fractional reserve of blood flow is obtained [3].

In some patients, pain is also caused by vasospasm. Spontaneous vasospasm is seldom detected during coronarography. Therefore, when variant angina is suspected, performing provocative tests is necessary. The results of testing samples from patients with muscle bridges using these tests have unique characteristics. Thus, an acetylcholine test was performed in 114 patients with chest pain during coronary angiography. The results showed that 41 patients had a myocardial bridge which was located in the middle segment of the left anterior descending artery in all these patients [10]. We found a description of a patient with multiple vasospasms during coronary angiography with intense anginal pain occurring immediately before a planned examination. After intracoronary administration of nitroglycerin, the vasospasm regressed everywhere except for the middle segment of the left anterior descending artery, which is where the myocardial bridge was indicated. After provocation with acetylcholine, vasospasm occurred only at the localization site of the myocardial bridge [8]. Patients with muscle bridges can be treated conservatively or by using invasive methods. Recommended drugs for patients with muscle bridges are calcium antagonists and beta-blockers.

Currently available methods of surgical correction for this pathology include tunnelized segment stenting, coronary artery bypass grafting, supraarterial myotomy, and laser myotomy. According to published data, various results have been obtained for non-drug treatment in patients with muscle bridges. According to Fu Wai Hospital (2005), a somewhat better long-term result of treatment was observed in the group of patients who underwent surgical treatment, which included coronary artery bypass grafting ($n = 8$) and supraarterial myotomy ($n = 7$). Coronary angiography performed 24 months after the intervention showed neither a recurrence of angina pectoris nor restenosis and shunt dysfunction. Additionally, 2/4 patients who underwent balloon angioplasty and stenting experienced recurring angina after 3 and 7 months, respectively. The control coronary angiography showed hyperplasia of the tunnelized artery and systolic compression [11].

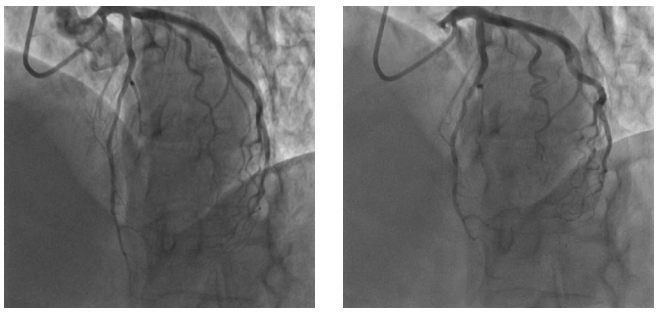
It is interesting that the very presence of this congenital pathology is recognized by some experts as an individual variant of normal coronary artery topography due to its high frequency in patients. Additionally, there is evidence that the risks of adverse outcomes depend on the depth of the muscle bridge. The tunneled segment varies in depth from 0.5 to 10 mm. The length of the muscle bridge varies from 10 to 30 mm [1]. After analyzing the results of an autopsy study on hearts with muscle bridges, A. Morales et al. concluded that, in deep tunnelized segments, myocardial changes are not normal and may be associated with an increased risk of sudden death, including sudden death during physical exertion [6].

The feasibility of surgical intervention for coronary fistula is determined by the hemodynamic significance of the fistula. According to the literature, discussing surgical strategies for closing coronary fistulas, even in children, is necessary because of the risk of complications, including thrombosis, endocarditis, aneurysmically dilated vessel rupture, and myocardial infarction, progressing in the future. Depending on the anatomical features of a coronary fistula, an interventional or surgical technique is used for treatment. Percutaneous fistula occlusion is the optimal treatment for coronary fistula. However, some literature sources indicate that adults can experience complications in the form of device migration, myocardial infarction, fistula recanalization, and thrombus formation. Therefore, in patients with borderline risk, including those with small fistulas, performing regular examination using coronary angiography and echocardiography is advisable for recognizing dilatation of the vessel supplying the fistula [4].

We observed a patient with angina pectoris who showed no signs of coronary artery stenosis but showed a muscle bridge and fistula of the coronary artery on angiography. The patient (76-year-old) was hospitalized as planned, so that a treatment strategy considering persistent chest pain could be determined. Medical history of the patient included hypertension for 10 years, with a maximum blood pressure of 200/100 mm Hg and with the frequently observed value of 110/60 mm Hg associated with antihypertensive therapy. In January 2014, the patient was repeatedly hospitalized and was diagnosed with unstable angina pectoris. The patient described the chest pain as a pressing discomfort behind the sternum and in the left half of the chest that presented throughout the day but more often at night. The duration of the pain varied and sometimes persisted for more than an hour. At night, the pain intensified when the patient turned onto her left side. Nitrates did not have any pronounced effect. She complained of a decrease in exercise tolerance throughout the past two years.

From March 18, 2014 to April 1, 2014, she was treated at a hospital in an emergency for suspected unstable angina. The patient underwent coronary angiography, which revealed a myocardial bridge of the left anterior descending artery with dynamic stenosis of up to 70%, and a small shunt from the pool of the left coronary artery into the left ventricle was revealed using contrast medium (Fig. 1 *a*, *b*). Coronary artery stenosis was not detected. On discharge, additional examinations, including Holter ECG monitoring and stress echocardiography, were recommended to the patient. Disaggregants (acetylsalicylic acid), calcium antagonists (amlodipine), diuretics (torasemide), ACE inhibitors, and beta-blockers were prescribed as components of a permanent medical treatment.

From June 20, 2014 to July 4, 2014, the patient was hospitalized, as planned, for follow-up examination. Stress echocardiography did not show deterioration in the regional contractility of the left ventricular myocardium. However, an insufficient coronary reserve of the left anterior descending artery i. e., a small increase in the maximum diastolic velocity from 42 cm/s to 48 cm/s, which is less



a *b*

Fig. 1. Myocardial bridge of the left anterior descending artery (coronary angiography) diastole (*a*), systole (*b*)
Рис. 1. Мышечный мостик передней нисходящей артерии (коронарная ангиография) в диастолу (*a*) и в систолу (*b*)

than double, was detected. A reliable indicator for hemodynamically significant stenosis is an increase in linear velocity of blood flow in the stenosis zone by a factor of 2 or more. Holter ECG monitoring showed that, during the day, a sinus rhythm with a tendency towards moderate bradycardia and a slightly pronounced supraventricular ectopic activity in the form of episodes of atrial rhythm and solitary extrasystoles were recorded. The registered pauses, which lasted up to a maximum of 1424 ms, were due to sinus arrhythmia, and the number of single ventricular extrasystoles was minimum. No life-threatening rhythm and conduction disturbances were found. After the examination, the patient was consulted by a cardiac surgeon, who concluded that there were no indications for surgical treatment. He recommended continuing conservative therapy. Therefore, dose of 2 tablets of sydnopharm at night was included in the medication regimen. During the therapy, no significant clinical improvement was noted.

A few years later, the patient was again hospitalized because of increased chest pain and a strategy for further treatment was determined. ECG showed sinus bradycardia (medically determined) with a frequency of 46–57 beats per minute, deviation of the electrical axis to the left that blocked the anterior-upper branching of the left branch of the His bundle, and left ventricular hypertrophy with disturbance in the ischemic repolarization processes, including negative T waves in the lateral wall region, septum, and apex of the left ventricle. Based on the results of the treatment strategy during hospitalization and considering the ineffectiveness of antianginal therapy, repeated planned coronary angiography was recommended to the patient. It is possible that, during the three years since the first coronarography, the patient developed stenosis due to atherosclerosis of the coronary arteries, which would explain why the therapy was ineffective. However, severe angina pectoris had been observed in the patient for a long time before the she underwent coronary angiography, which did not show stenosis of the coronary bed. Therefore, it can be concluded that the pain is associated with the myocardial bridge, as hemodynamic coronarography did not show a negative effect on the coronary artery.

This case demonstrates a combination of two congenital anomalies, muscle bridge and coronary artery fistula, in the structure of the coronary arteries without coronary artery stenosis in an elderly patient with angina pectoris, as shown on angiography. Shunting of blood from the pool of the left coronary artery into the left ventricle was insignificant. Additionally, the muscle bridge was of primary importance in the development of angina pectoris. Conservative treatment was ineffective. Repeating coronary angiography and performing diagnostic provocative tests to choose an optimal method of muscle bridge correction is planned for the patient.

REFERENCES

1. Бокерия Л.А., Суханов С.Г., Стерник Л.И., Шатахян М.П. Миокардиальные мостики. — М.: Научный центр сердечно-сосудистой хирургии им. А.Н. Бакулева РАМН, 2013. [Bokeriya LA, Sukhanov SG, Sternik LI, Shatakhyan M.P. Miokardial'nye mostiki. Moscow: Nauchnyy tsentr serdechno-sosudistoy khirurgii im. A.N. Bakuleva RAMN; 2013. (In Russ.)]
2. Кулешова Э.В., Панов А.В. Хроническая ишемическая болезнь сердца // Кардиология. Национальное руководство. Краткое издание. — М.: ГЭОТАР-Медиа, 2018. — С. 409–410. [Kuleshova EV, Panov AV. Khronicheskaya ishemicheskaya bolezn' serdtsa. In: Kardiologiya. Natsional'noe rukovodstvo. Kratkoe izdanie. Moscow: GEOTAR-Media; 2018. P. 409-410. (In Russ.)]
3. Рамракха П., Хилл Дж. Справочник по кардиологии. — М.: ГЭОТАР-Медиа, 2011. — С. 198–199. [Ramrakha P, Hill J. Oxford handbook of cardiology. Moscow: GEOTAR-Media; 2011. P. 198-199. (In Russ.)]
4. Субботин В.М., Белозеров Ю.М., Брегель Л.В. Коронарные фистулы // Российский вестник перинатологии и педиатрии. — 2015. — Т. 60. — № 1. — С. 20–21. [Subbotin VM, Belozerov YM, Bregel LV. Coronary artery fistulas. *Russian Bulletin of Perinatology and Pediatrics*. 2015;60(1):16–22. (In Russ.)]
5. Alegria JR, Herrmann J, Holmes DR, Jr., et al. Myocardial bridging. *Eur Heart J*. 2005;26(12):1159-1168. https://doi.org/10.1093/eurheartj/ehi203.
6. Morales AR, Romanelli R, Tate LG, et al. Intramural left anterior descending coronary artery: Significance of the depth of the muscular tunnel. *Hum Pathol*. 1993;24(7):693-701. https://doi.org/10.1016/0046-8177(93)90004-z.
7. Morrow DA, Boden WE. Stable ischemic heart disease. In Braunwald's heart disease: a textbook of cardiovascular medicine. 10th ed. Philadelphia: Saunders, an imprint of Elsevier Inc.; 2015. P. 1222-1223.
8. Nardi F, Verna E, Secco GG, et al. Variant angina associated with coronary artery endothelial dysfunction and myocardial bridge: A Case Report and Review of the Literature. *Intern Med*. 2011;50(21): 2601-2606. https://doi.org/10.2169/internalmedicine.50.6086.

9. Popma J, Scott K, Bhatt DL. Coronary Arteriography and Intracoronary Imaging. In: Braunwald's heart disease: a textbook of cardiovascular medicine. 10th ed. Philadelphia: Saunders, an imprint of Elsevier Inc.; 2015. P. 412.
10. Teragawa H, Fukuda Y, Matsuda K, et al. Myocardial bridging increases the risk of coronary spasm. *Clin Cardiol*. 2003;26(8):377-383. https://doi.org/10.1002/clc.4950260806.
11. Wan L, Wu Q. Myocardial bridge, surgery or stenting? *Interact Cardiovasc Thorac Surg*. 2005;4(6):517-520. https://doi.org/10.1510/icvts.2005.111930.
12. Winchester DE, Pepine CJ. Nonobstructive atherosclerotic and nonatherosclerotic Coronary Heart Disease. In: Fuster V, Harrington RA, Narula J, Eapen ZJ. Hurst's the Heart. 14th ed. New York: McGrow-Hill Education; 2017. P. 938-939.

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