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CASE REPORT

A RARE CASE OF A PATIENT WITH A MYOCARDIAL BRIDGE IN COMBINATION WITH BICUSPID VALVE AND SUBAORTIC STENOSIS

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Abstract. *Combined congenital heart defects in adults are very rare, and knowledge of their diagnostic-therapeutic approach is of interest to specialists. Subaortic stenosis (SAS) is a heart disease of unclear etiology and variable clinical manifestation. In some cases, SAS is described in combination with a congenital bicuspid aortic valve, but a real rarity is the combination of the listed lesions with the non-obstructive coronary stenosis caused by a myocardial bridge. A middle-aged woman with chest pain and dyspnea during minimal physical exertion, that subsides after rest, was referred for surgery. Two-dimensional transthoracic echocardiography demonstrated left ventricular hypertrophy with preserved systolic function without anomalies of cardiac wall motion. A dynamic gradient was observed in the left ventricular outflow tract reaching up to 90 mmHg. Bicuspid aortic valve was presented with high-grade regurgitation. The picture of myocardial ischemia was complemented by selective coronary angiography demonstrating a rarely presented myocardial bridge over the left anterior descending coronary artery. The diagnostic approach and surgical corrections performed are the subject of this report.*

Key words: *myocardial bridge, bicuspid aortic valve, subvalvular aortic stenosis*

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INTRODUCTION

The coronary myocardial bridge is a congenital anomaly of the course of one or more coronary arteries first described by Reyman in 1737. Historically, myocardial bridges were considered as benign lesions, as it was assumed that systolic compression of the coronary artery would not lead to ischemic changes due to the prevailing diastolic coronary blood flow. The affected epicardial coronary artery passes under a bridge of myocardium with a limited area, subsequently coming back out again above the myocardium. A 'muscle loop' is formed over the coronary artery, which is tightened during systole. In diagnostic terms, the frequency of myocardial bridges varies between 0.5% to 12% of people undergoing selective coronary angiography [1, 2]. However, published autopsy studies shows a significantly higher frequency between 5% to 86%, respectively [1, 2]. In the majority of cases, the myocardial bridge is asymptomatic. Patients are born with it and never realize that they have such a lesion. Contemporary reports of unstable angina, arrhythmia, myocardial infarction and sudden death highlight the hemodynamic importance of this disease in a subset of patients [3].

In turn, the bicuspid aortic valve represents the most common congenital heart defect involving approximately 1% of the total population [4]. It occurs twice as often in men than in women, and its presence predisposes to degenerative changes of the aortic valve and is associated with dilatation of the ascending aorta. Bicuspid valves can progress to both valvular stenosis and valvular regurgitation, depending on the presence of excess cusp tissue. Subvalvular aortic stenosis (subaortic stenosis or SAS) is a rare disease in the adult population by itself, with different etiologies and clinical manifestations. SAS usually represents a progressive disease characterized by significant obstruction of the left ventricular outflow tract (LVOT), left ventricular hypertrophy, and aortic valve destruction with subsequent regurgitation. Fixed subaortic stenosis (SAS) may be due to a discrete fibrous membrane, myocardial hypertrophy, or a combination of both [6].

In this article, a case of a patient with a rare combination of multiple congenital heart defects discovered on the basis of angina pectoris has been described. Part of interest is the diagnostic approach, that arranges the puzzle of the ischemic picture, and the applied surgical techniques that solve the described problems.

CLINICAL CASE

A 61-year-old woman with chest pain and dyspnea on moderate exertion was admitted for surgical treatment. The patient had high-grade aortic insufficiency with bicuspid aortic valve, subvalvular aortic stenosis-muscular type, heart failure III class of NYHA, as well as arterial hypertension. Cardiac catheterization was used to confirm the diagnosis. Ventriculography was performed, which showed no segmental wall motion abnormalities of the ventricles. Coronary vessels tracing revealed the presence of a myocardial bridge in the middle third of the left anterior descending (LAD) coronary artery. The presence of dynamic strangulation of the LAD (non-obstructive type) dependent on the phases of the cardiac cycle with more than 90% compression of the lumen was observed (Fig. 1). No significant stenosis of LAD, as well as the rest of the coronary vessels were observed. Preoperative sinus rhythm of 68 beats/min, dull heart sounds on auscultation and a weak systolic murmur in the aortic position were notable upon physical examination. The performed laboratory tests were without particular deviations from the norm, except for a slight increase of leukocytes (Leu) – 12.9 × 10^9/L; Fibrinogen F-I – 4.61 g/l, and increased blood glucose level – 8.1 mmol/l. From the two-dimensional transthoracic echocardiography at admission were measured: aorta ascendens-35 mm; interventricular septum – 14 mm; left ventricular lateral wall – 11 mm and preserved ventricular function. Presence of dynamic gradient in an outflow tract of the left ventricle up to 90 mm Hg was registered. The mitral valve was without regurgitation. The aortic valve was bicuspid with high-grade regurgitation.

General anesthesia was administered according to a standard protocol for aortic valve disease surgery. After careful preparation of the operative field, a mediansternotomy was performed. The surgical intervention was performed under conditions of normothermic extracorporeal blood circulation – temperature up to 36°C. After aortic cross-clamping, cardiac arrest was delivered by infusing 1000 ml of cold crystalloid antegrade cardioplegia through the coronary ostium. The bicuspid aortic valve and the prominent part of the hypertrophic septum were excised. A mechanical prosthesis SJM Regent 19 mm was implanted using single U-shaped etibond 2/0 sutures with felt, and an adequate function of the prosthesiswas achieved.

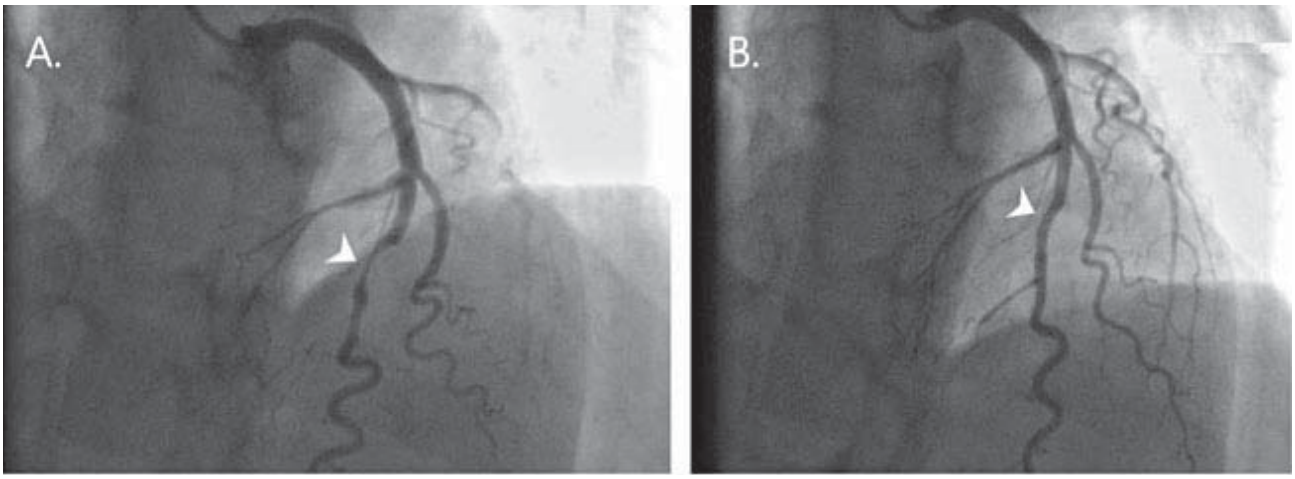

Fig. 1. Selective coronary angiography A. systolic and B. diastolic phase

Passed to the previously targeted muscle band in the middle segment of the LAD. The left anterior descending coronary artery was decompressed through a supra-arterial myotomy about 20 mm long. A Beaver® blade knife (standard coronary artery dissection techniques) was used, with the deepest level of myotomy penetration reaching 3 mm in the middle of the LAD. The entire course of the artery was traced, with no areas of compression observed. Weaning from cardiopulmonary bypass was smooth and without complications. The patient was admitted to intensive care unit without inotropic support, attached to invasive pulmonary ventilation. This was followed by extubation on the 16th hour and discharge from intensive care on the 4th postoperative day. A control transthoracic echocardiography revealed preserved LV systolic function. A mechanical prosthesis was scanned in the aortic position with gradients of 28/13 mm Hg, without regurgitation (Fig. 2).

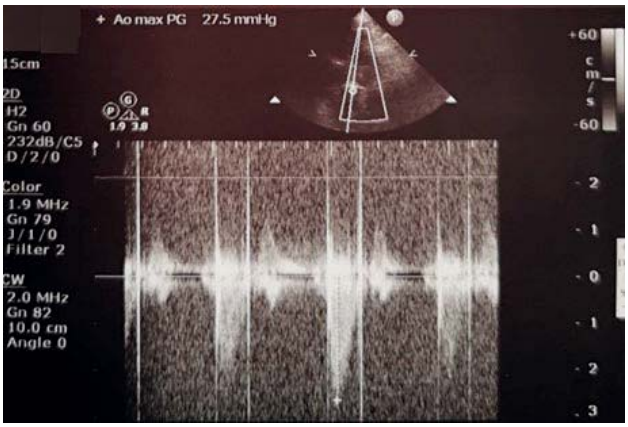

Fig. 2. Control transthoracic echocardiography

The patient was discharged on the 10th postoperative day in good condition. At one-year follow-up, no new ischemic episodes were reported and no dynamics were observed in the gradient across the prosthesis on the follow-up echocardiogram.

DISCUSSION

In the majority of patients, subvalvular aortic stenosis is detected by chance during evaluation for other heart defects. It is observed significantly more often in males and is responsible for 65% to 75% of cases [7, 8], with a male-to-female ratio of 2:1. The prevalence of SAS is 6.5% of all congenital heart disease in adults [9] and in more than 50% of cases it is seen in patients with other cardiovascular malformations [10, 11]. Bicuspid aortic valve, membrane ventricular septal defect, aortic coarctation or patent ductus arteriosus most often come in combination with subaortic stenosis [12, 13]. This disease usually tends to progress slowly. Without appropriate surgical intervention, SAS can progress to left ventricular hypertrophy and dysfunction, aortic regurgitation, endocarditis, arrhythmias, and death [14].

From the above it seems clear that subaortic stenosis is often seen in combination with a bicuspid aortic valve. This valvular abnormality is the most common congenital heart defect. It occurs in approximately 1% of the general population, with a male to female ratio of 2:1 [4]. The bicuspid aortic valve consists of two cusps, in the majority of cases they are asymmetrical. Fusion between the right and the left coronary cusp most often occurs, and this anomaly is associated with aortic coarctation. In other cases, the fusion is between the non-coronary and right coronary cusp, which is associated with valvular abnormalities such as aortic stenosis and aortic regurgitation. In the reported case, this congenital heart disease has degenerated into a high degree of regurgitation [15]. It is a well-known fact that coronary perfusion occurs mainly during diastole. A decrease in the patient's aortic diastolic pressure decreases coronary blood perfusion. Moderate myocardial hypertrophy due in this particular case to subvalvular stenosis leads to increased oxygen consumption. Insufficiency coronary perfusion and increased oxygen consumption of the left ventricle predispose the myocardium to ischemic injury.

The completeness of the clinical picture is achieved with the diagnosis of the myocardial bridge. It should be noted here that an ECG is not a reliable or convincing diagnostic tool for diagnosing MB. The majority of patients with symptomatic MB show normal ECG results. Of the non-invasive methods used in the diagnosis of myocardial bridging, multislice computed tomography (MSCT), stress single-photon emission computed tomography and stress echocardiography are used. The MSCT defines bridges as segments surrounded by myocardium [16]. In selective coronary angiography, the diagnosis is made by the dynamic change in the diameter of the vessel between systole and diastole in the myocardial bridge area. A significant "milking effect" is present when there is a ≥70% decrease in lumen diameter during systole and a persistent ≥35% decrease in minimum lumen diameter during mid-to-late diastole [17]. The systolic narrowing of the bridge can be accentuated by intracoronary injection of nitroglycerin by vasodilation of adjacent coronary segments excluding a bridge [18].

Schwarz's classification (Table 1) can serve as a guide to lead the therapeutic approach in patients with a myocardial bridge [19]. Patients with Schwarz type A did not require treatment, while patients with types B and C showed significant symptomatic im-

Table 1. Schwarz Classification for Myocardial Bridges and Treatment [19]

Schwarz Type	Criteria	Objective Signs of Ischemia	Treatment
A	Incidental finding on angiography	−	None
B	Ischemia on stress test	+	BB or CCB
C	Altered intracoronary hemodynamics (quantitative coronary angiography/coronary flow reserve/Doppler)	+/−	BB or CCB and/or cardiac surgery

BB = beta-blocker; CCB = calcium channel blocker.

provement with beta-blockers or calcium channel blockers at 5-year follow-up. Patients with Schwarz type C refractory to drug therapy should be considered for surgical repair of the myocardial bridge.

The methods of surgical correction include myotomy or coronary artery bypass graft (CABG). In a typical case of myotomy, the goal is to decompress the coronary vessel. Potential complications of myotomy include perforation of the myocardial wall, ventricular aneurysm formation and postoperative bleeding. Problems with CABG are mainly related to graft obstruction. In a study of 77 patients, Zhang summarized that supra-arterial myotomy is preferred in patients with isolated myocardial bridge and that acceptable results can be achieved when choosing supra-arterial myotomy in combination with CABG or other cardiac surgery in patients with a myocardial bridge in combination with other heart disease [20].

The multicomponent aspect of the case increases the requirements in the diagnostic and treatment plan. A good selection of operative techniques is the basis of a positive outcome for the patient. Incomplete repair of the lesions would result in persistent ischemia and eventual fatal outcome for the patient.

CONCLUSION

Multicomponent ischemic diseases are a challenge both in light of the diagnostic approach but also in light of the therapeutic completeness of the healing process. The risk of acute myocardial infarction increases in direct proportion to the number of cardiac lesions due to their cumulative involvement in the ischemic process. The combination of morphological damage due to congenital valvular lesion, subvalvular stenosis and coronary stenosis during systole poses a serious danger to the health and life of the patient. Their different mechanism of action can be seen as pieces of a puzzle. Surgical resection of subvalvular stenosis and prosthetic aortic valve are classic procedures that do not cause doubt in the choice of therapeutic approach. However, the choice of operative procedure for myocardial bridging is of interest. It is our opinion that regardless of the often chosen coronary bypass, the non-obstructive coronary component would create conditions for competing blood flow, and this in turn to graft obstruction. Due to the presented facts, supraarterial decompression myotomy was chosen.

Disclosure summary: *The authors have nothing to disclose.*

Abbreviations:

SAS – Subvalvular Aortic Stenosis
LAD – left anterior descending coronary artery
LVOT – left ventricular outflow tract
MB – Myocardial Bridge
SCAG – selective coronary angiography
ECG – electro cardiography
TTE – transthoracic echocardiography
CMR – Cardiovascular Magnetic Resonance Imaging

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