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Tilea I[1], Moraru L[2], Raicea V[2], Tilea Brindusa[3], Bocicor Andreea Elena[4], Suciu H[2]

Bicoronary-pulmonary fistulae and severe mitral valve regurgitation – uncommon cause of myocardial ischemia – a case report

[1]Cardiac Catheterization Laboratory, Centre of Cardiovascular Diseases, University of Medicine and Pharmacy at Tîrgu Mureş, Romania
[2]Cardiovascular Surgery Clinic, Centre of Cardiovascular Diseases, University of Medicine and Pharmacy at Tîrgu Mureş, Romania
[3]Cardiac Rehabilitation Clinic, Emergency County Clinical Hospital Tîrgu Mureş, University of Medicine and Pharmacy at Tîrgu Mureş, Romania
[4]Infectious Disease Clinic, County Clinical Hospital Tîrgu Mureş, University of Medicine and Pharmacy at Tîrgu Mureş, Romania

ABSTRACT
Bicoronary - pulmonary artery fistulae are rare conditions. Their association with mitral valve prolapse is even rarer and randomly reported. This association is important to be recognized in clinical practice because of the differential diagnose problems. Closing the coronary fistulae and mitral valve replacement during the same surgical procedure is probably the optimal management of these patients. We report a case involving the correction of congenital bicoronary-pulmonary artery fistulae and mitral valve replacement within the same surgical procedure in a 56 years old female patient with angina and clinical signs of left ventricular failure associating the fistulae to severe mitral regurgitation due to mitral valve prolapse. Past medical history revealed autoimmune thyroiditis, atrial fibrillation, mitral and tricuspid valve regurgitation. At admission physical examination revealed stable vital signs, irregular tachycardia with significant pulse and a mitral regurgitation systolic murmur. ECG showed atrial fibrillation, no ischemia. Echocardiography revealed severe mitral regurgitation, prolapse of anterior and posterior mitral leaflets, moderate tricuspid valve regurgitation, and mild pulmonary hypertension. Coronary angiogram showed no significant lesions of the epicardial vessels but high flow congenital bicoronary-pulmonary fistulae (right coronary artery and left coronary artery to main pulmonary artery). Surgical correction of the congenital bicoronary-pulmonary fistulae was performed simultaneously with mitral valve replacement in the same session. Post-surgical evolution was uneventful. Post-procedural ECG showed atrial fibrillation with a controlled heart rate, postoperative echocardiography showed normal functional and normal positioned prosthetic mitral mechanical valve, and rather normal left ventricle function. Coexistence of bicoronary-pulmonary fistulae and mitral valve insufficiency due to prolapse in a symptomatic patient with angina pectoris is a very rare clinical entity. Solving both abnormalities within the same surgical procedure was the optimal management for this patient.

Keywords: bicoronary-pulmonary fistulae, mitral regurgitation, mitral valve prolapse, angina pectoris.

Moraru Liviu

Cardiac Surgery Clinic, Centre of Cardiovascular Diseases, Emergency County Clinical Hospital Tîrgu Mureş
50 Gh. Marinescu st., zip code 540136, Tîrgu Mureş, Romania
Tel: +40.723.256970
E-mail: dr.liviu.moraru@gmail.com

Background

Bicoronary-pulmonary artery fistulae are rare congenital conditions. The majority of these fistulas arise from the right coronary artery and the left anterior descending coronary artery; the circumflex coronary artery is rarely involved. Coronary artery fistula, first reported by Krause in 1865 [1] was thought to be a rare abnormality of the coronary arteries. It has been reported more frequently in recent years because of the increase in coronary artery procedures performed. The incidence in the adult population is around 0.1 to 1% [2,3,4] and it may be either congenital or acquired. Only 5% of the fistulas are dual. However, patients with ischemic chest pain owing to dual or multiple coronary fistulas without significant coronary artery stenosis are rather rare [5,6,7]. Bilateral coronary artery to pulmonary artery fistulas is an uncommon congenital anomaly. These fistulas have a clinical and embryological significance [1].

Case presentation

A 56 years old female was referred to Cardiology Department with the suspicion of ischemic heart disease, presenting chest pain, shortness of breath, palpitations, fatigue. At the moment of the admittance she had been diagnosed 2 months before with severe mitral insufficiency due to mitral valve prolapse. She has also been previously diagnosed with tricuspid valve insufficiency, atrial fibrillation, and left cardiac failure. She also had previous medical history of autoimmune thyroiditis and multi-nodular goitre with hypothyroidism.

At admission the patient presented the clinical signs of left ventricular failure – progressive shortness of breath, fatigue to a minimum level of physical activity and she also presented angina type chest pain and palpitations – irregular tachycardia. Physical examination revealed stable vital signs, blood pressure at 120/70 mmHg, irregular tachycardia, pulse deficit with a heart rate of 126 bpm and a peripheral irregular pulse rate of 100 bpm, and a mild systolic murmur at mitral valve, irradiating towards the axilla. Laboratory test results pointed to an elevated level of cholesterol (6.44 mmol/l) and high TSH level (4.98 uUI/ml). Rest ECG showed atrial fibrillation, high heart rate (100 bmp), and normal QRS axis, without ST-T changes that could suggest ischemia (Figure 1).

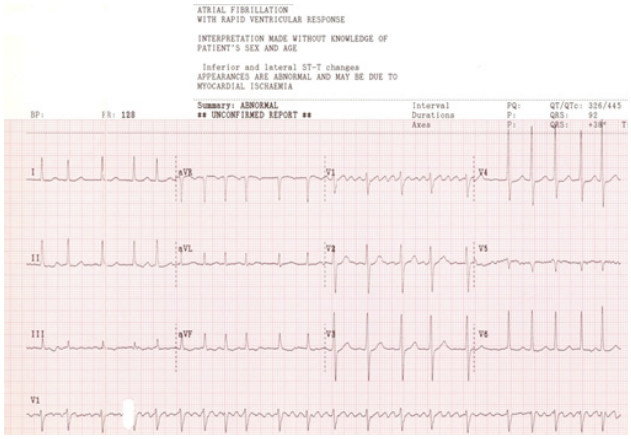

Figure 1 – Preoperative EKG

Echocardiography showed normal left ventricular dimensions and function, mild left atrial enlargement (46 mm), 3rd degree mitral regurgitation due to prolapse of anterior and posterior mitral leaflets, mild aortic valve regurgitation, moderate tricuspid valve regurgitation, mild pulmonary hypertension with systolic pulmonary artery pressure (PAPs) estimated to 40-45 mmHg. Coronary angiography showed no significant atherosclerotic stenosis of the epicardial coronaries but a high flow congenital fistula between the right coronary artery (Figure 2 a,b) and left anterior descending artery (Figure 3 a,b) on one side and the main pulmonary artery on the other side. A medical team involving cardiologist, cardiovascular surgeon and angiographist decided on the surgical resolution of both the congenital fistula and mitral valve replacement.

Surgical procedure was performed and intraoperative exploring of the heart revealed multiple tracts of fistulae situated in the outflow tract of the right ventricle and between the aorta

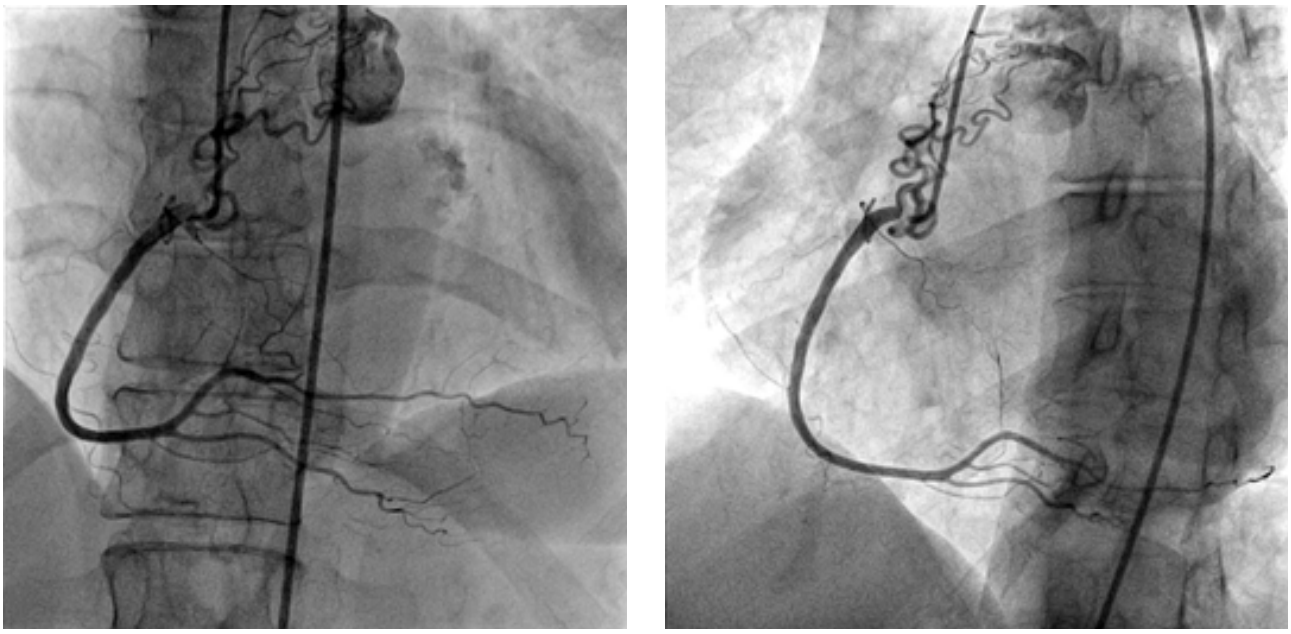

Figure 2 – Congenital coronary fistula between right coronary artery and main pulmonary artery (a, b)

and the pulmonary artery. First surgical step was the correction of the congenital bicoronary-pulmonary fistulae by intrapulmonary suture of the drainage orifices and epicardial suture of the communication vessels. The procedure continued with mitral valve replacement using a prosthetic mechanical valve ”St. Jude Medical” 29 and the deliming of posterior mitral annulus.

Postoperative evolution was uneventful. Post-procedural ECG showed atrial fibrillation with a heart rate of 75 bpm, normal QRS axis (Figure 4), and postoperative echocardiography showed normal

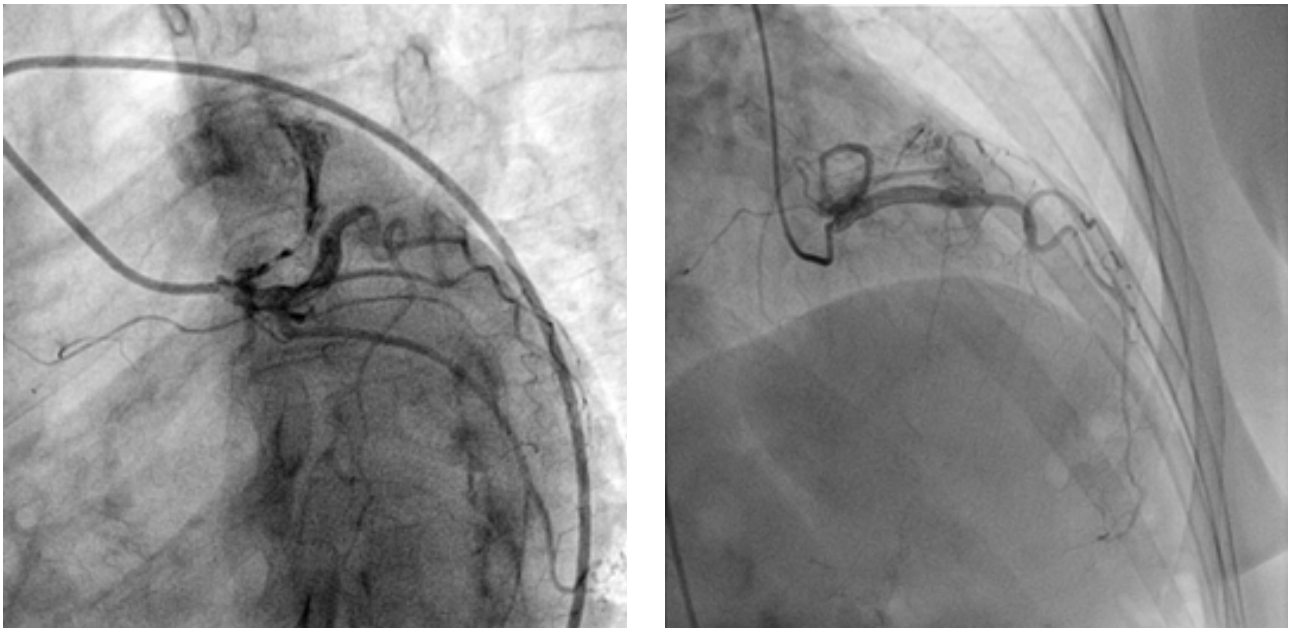

Figure 3 – Congenital coronary fistula between left coronary artery and main pulmonary artery (a, b)

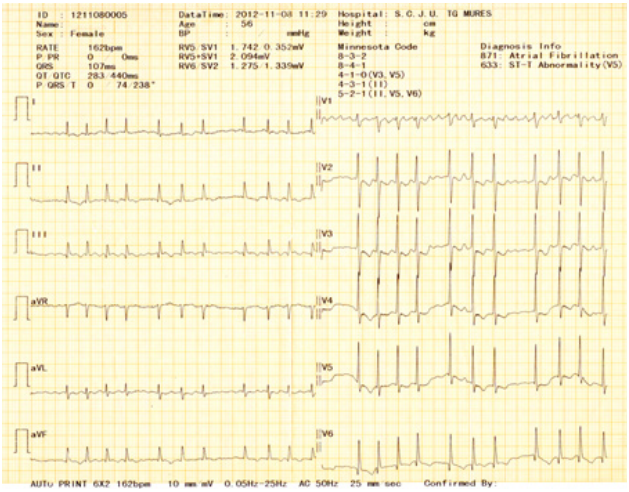

Figure 4 – Postoperative EKG

functional and normal positioned prosthetic mitral mechanical valve SJM no. 29, with a trans-prosthetic gradient 6/2,5 mm Hg, EF 55% (Figure 5, a,b).

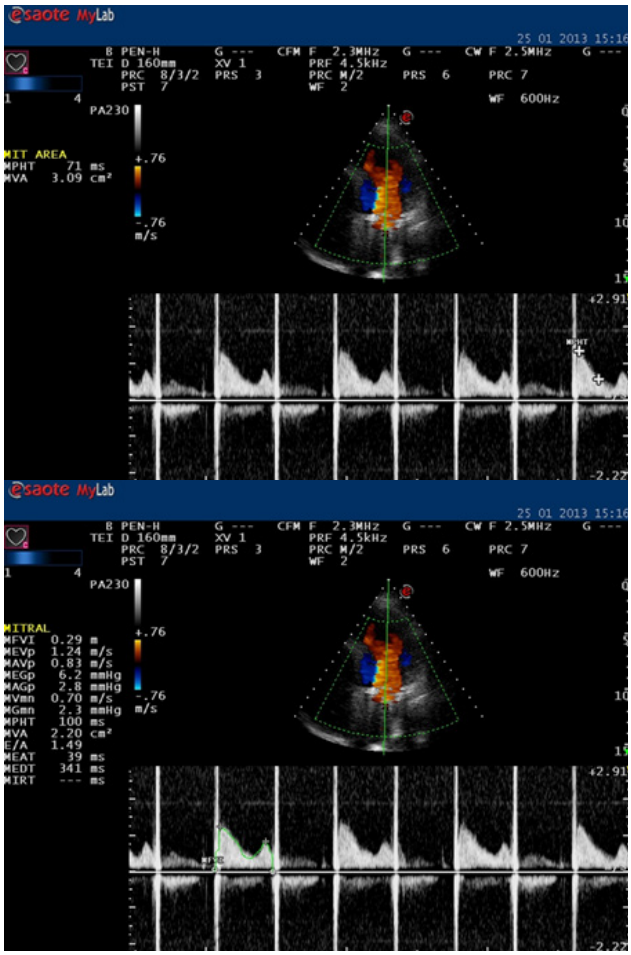

Figure 5 – Postoperative echocardiography showed – functional and normal positioned prosthetic mitral mechanical valve SJM no. 29 (a, b)

Discussion

We report a rare case of combination of right coronary and left anterior descending artery coronary fistula draining into main pulmonary artery associated to mitral insufficiency due to mitral valve prolapse, in a 56 years old female patient who presented in the emergency room with acute pain chest, signs of left ventricular failure and palpitations.

A variety of coronary arteries abnormalities were described. If they associate mitral valve prolapse they can form particular rare anatomic entities. First case of coexistent bicoronary-pulmonary fistulae and Barlow`s syndrome (mitral valve prolapse) in a non-symptomatic patient was cited in 1983 [8,9]. Left ventricular angiography revealed a contractility abnormality and Hammer et al described congenital changes in collagen III and AB patterns showing that coexistence of mitral valve prolapse and coronary pulmonary fistulae are not accidental [9].

In our patient the differential diagnosis of angina pectoris implicates the difference between the mitral insufficiency due to mitral valve prolapse and the mitral valve insufficiency of ischemic etiology. Since the basic ECG showed no sign of ischemia and the echocardiography practically revealed the mitral valve prolapse as the cause of mitral insufficiency we considered angina pectoris as a consequence of the coronary theft due to the bicoronary pulmonary fistula and the clinical signs (mitral systolic murmur) of mitral insufficiency due to mitral prolapse.

Conclusions:

Particularity of our case report was the coexistence of bicoronary pulmonary fistulae and mitral valve insufficiency due to prolapse in a symptomatic patient with angina pectoris. The simultaneously surgical management of both abnormalities was the best option in the clinical management of this patient.

Acknowledgement

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