

Acute coronary syndrome or steal phenomenon – a case of right coronary to right ventricle fistula

Efekt podkradania czy ostry zespół wieńcowy, przetoka pomiędzy prawą tętnicą wieńcową a prawą komorą – opis przypadku

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Abstract

We report a case of 53-year-old male admitted to the hospital with severe chest pain and elevation of troponin I levels, who was transferred to our department for a coronary angiography. Contrast injection into the right coronary artery (RCA) demonstrated no evidence of coronary artery disease. However, a double outlet coronary artery fistula which drained into the right ventricle was visualised. With standard 2-dimensional echocardiographic imaging abnormal flow was detected in the inflow part of the right ventricular free wall. Three-dimensional colour Doppler imaging clearly demonstrated the presence of double outlet RCA – right ventricle coronary fistula with sufficient quality.

Key words: coronary artery fistula, myocardial ischaemia, 3D echocardiography

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Background

Coronary artery fistulas are not a frequent finding on coronary angiography in adults although they represent the most common haemodynamically significant congenital coronary abnormality [1, 2]. Coronary fistulas are classified as abnormalities of termination of the coronary artery with abnormal drainage into the cardiac cavities or great vessels. A coronary artery fistula is a sizable communication between a coronary artery, which bypasses the myocardial capillary bed, and enters either a chamber of the heart (coronary-cameral fistula, CCF) – usually the right ventricle (RV) (60%) – or any segment of the systemic or pulmonary circulation (coronary arteriovenous fistula, CAF) [1-7]. The aetiology of coronary artery fistulas may be congenital, traumatic or iatrogenic, i.e. after coronary intervention or valve replacement [3, 4]. Diagnosis of CAF or CCF is based on angiography. Echocardiographic examination may be helpful for diagnosis, usually demonstrating the drainage site and sometimes the dilated fistula itself; moreover, it is used for the functional assessment of the fistula. We report a detailed anatomical definition of right coronary artery

– right ventricular fistula with double outlet into the right ventricle obtained with state-of-the-art three-dimensional colour Doppler mapping.

Case report

A 53-year-old male was admitted to our department with severe chest pain lasting for 5 hours, concomitant elevated troponin I and CK-MB mass. ECG showed normal sinus rhythm 75 beats/min, and ST elevations in leads V₁ to V₃ stable during the hospitalisation period. The first bedside echocardiogram (a limited study) revealed hypokinesis of the basal segments of the inferior-posterior wall and intraventricular septum. The biochemical panel of the patient is shown in Table I. The patient was symptomatic and considering the clinical presentation his preliminary diagnosis was suspected acute coronary syndrome. Thus, percutaneous coronary intervention was attempted. The coronary angiography revealed perfectly patent coronary arteries with no luminal irregularities. No percutaneous intervention was performed. However, a distal segment of the right coronary artery (RCA) was feeding a fistula draining into the RV chamber (Figure 1).

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Table I. Laboratory findings

WBC [μ l]	8.2×10^3
RBC [μ l]	5.2×10^5
HGB [g/dl]	15.8
HCT [%]	48.8
PLT [μ l]	348×10^3
Sodium [mmol]	144
Potassium [mmol]	4.29
Magnesium [mg/dl]	2.18
Glucose [mg/dl]	95
Urea [mg/dl]	37
Creatinine [mg/dl]	1.03
Total cholesterol [mg/dl]	218
Cholesterol LDL [mg/dl]	92
Cholesterol HDL [mg/dl]	60.8
Triglycerides [mg/dl]	196
CK [U/l]	124
Troponin I [ng/ml]	0.42*
CRP [mg/l]	2.46
D-dimer [ng/ml]	227
Fibrinogen [mg/dl]	482

Abbreviations: WBC – white blood cells, RBC – red blood cells, HGB – haemoglobin, HCT – haematocrit, PLT – platelets
* significant elevation

On day 2, complete echocardiographic examination allowed the detection of a pathological flow entering the inflow part of the RV (Figure 2). In Figure 3 we visualised two jets corresponding to blood drainage into the RV. Spectral Doppler recording of the fistula shows flow from the fistula into the RV with predominant diastolic component of 1 m/s (Figure 3). However, the optimal visualisation of

abnormal flow due to the fistula was obtained with colour Doppler real time three-dimensional echocardiography (3D) (Figure 4). The 3D examination directly showed the localisation of fistula ostia and the colour-coded jet of fistula/chamber blood flow with acceptable quality. After the angiography there was uncertainty regarding the number of communications between the RCA and RV chamber. Thus detailed examination with 3D echocardiography revealed two ostia entering the inflow part of the free wall of the RV (Figure 4).

The Qp/Qs ratio was 1:1. Thus we assumed that the fistula had no significant influence on the pulmonary circulation. However, considering the clinical presentation we postulated that the fistula, although small, might in certain conditions produce a 'steal' phenomenon that contributed to the patient's inferior wall ischaemia and chest pain. During hospitalisation CK-MB mass and troponin I normalised. A submaximal ECG exercise test performed on the third day was symptom-free, without any significant changes in ECG at workload of 10 METs (equal to age-predicted submaximal workload). Considering the lack of evidence that the fistula significantly impaired the coronary circulation, the patient was disqualified from coil or stent graft implantation. On the 6th day of hospitalisation he was discharged home with final diagnosis of coronary cameral fistula, arterial hypertension and hypercholesterolaemia. Medical treatment included bisoprolol, ramipril, atorvastatin and acetylsalicylic acid.

Discussion

Coronary artery fistulas are usually congenital malformations [1]. However, they may also develop after chest trauma or as complications of invasive cardiac procedures [2-5]. The haemodynamic significance of

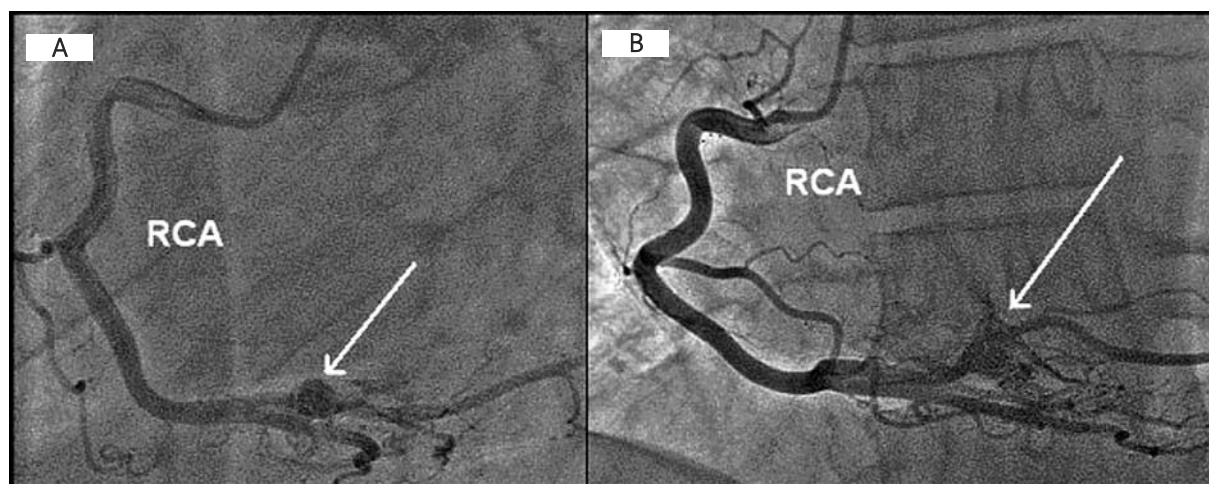


Figure 1. Angiography of the right coronary artery with visible coronary-ventricular fistula draining into the right ventricle (arrow). **A** – view of the right coronary artery (RCA) in LAO 50°, **B** – view of the RCA in cranial 30° position

coronary artery fistulas depends on their origin, size and the receiving chamber [2, 6, 7]. The most common artery developing a CAF is the RCA (55 to 60% of cases). The left coronary artery is not so commonly involved (up to 35% of CAFs). The most prevalent receiving chamber of coronary artery fistulas (45%) is the RV, followed by the right atrium (25%) and the pulmonary artery (20%) [1, 7]. The volume of the shunt depends on the fistula size and gradient between the aortic and receiving chamber pressures.

Echocardiography is helpful to define both coronary artery anomalies [8, 9] and fistulas. However, there are very few reports regarding the use of 3D echocardiography for such diagnosis. We were able to demonstrate that real-time 3D colour Doppler imaging was superior to standard mode due to unrestricted viewing plane manipulation. This allowed us to view the multiple drainage sites of the fistula.

If the coronary artery fistula is small and, consequently, shunting does not compromise antegrade flow in the feeding artery, the patient is usually asymptomatic. However, in some cases a significant coronary artery steal phenomenon occurs and ischaemia of myocardial segments perfused by the coronary artery distal to the fistula may develop [2-7]. Potentially, a haemodynamically significant fistula with a left-to-right shunt may lead to congestive heart failure, pulmonary artery hypertension, or myocardial ischaemia due to the steal phenomenon with concomitant cardiac arrhythmias [10, 12]. Uncommon complications caused by this clinical entity include endocarditis, embolisation with thrombotic material from the aneurysmal fistula, and potential rupture of the aneurysm. Haemodynamically insignificant fistulas are clinically silent and when not associated with other abnormal findings usually do not require further treatment.

In children a spontaneous closure of a fistula has been reported but it is seldom noted in adults [10, 11]. A spontaneous closure may be more common in iatrogenic, e.g. biopsy- or intervention-related CCF. In some cases, however, closure of the fistula is required. Mavroudis et al. [11] recommend elective coil occlusion in patients who fulfil the following criteria: absence of multiple fistulas, a single narrow drainage site, absence of large branch vessels, and safe accessibility to the coronary artery supplying the fistula. In our patient the fistula originated from RCA branches not meeting the above criteria. Therefore, since the patient remained asymptomatic on pharmacological therapy, we decided not to occlude this fistula. Stent graft occlusion of the feeding vessel might represent another treatment option. In our patient an early ECG stress test which was symptom-free without ST-T changes specific for ischaemia facilitated the decision of conservative treatment.

The question remains concerning the origin of symptoms and the rise of troponin I and CK-MB in our patient. Up to 1.9% of coronary angiograms performed in patients with myocardial infarction may reveal normal

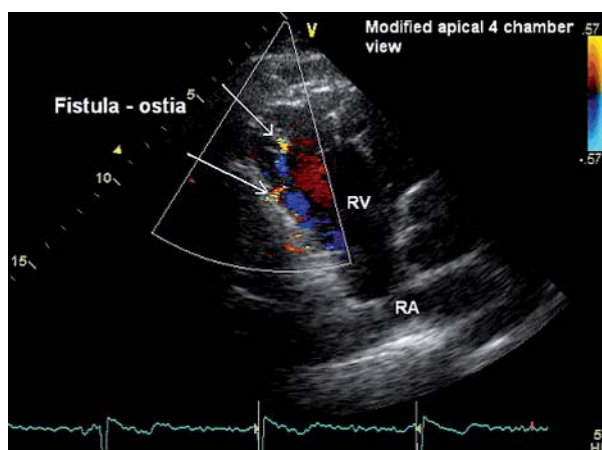


Figure 2. Modified 4-chamber long axis view. Numbers 1 and 2 indicate abnormal diastolic flow located in mid portion of right ventricular (RV) free wall

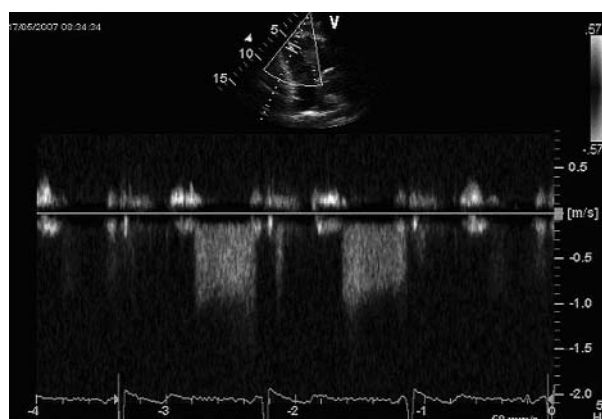


Figure 3. Pulsed spectral Doppler of fistula flow sample at the apically located orifice – diastolic part of spectrum directed away from the probe, confirming coronary origin

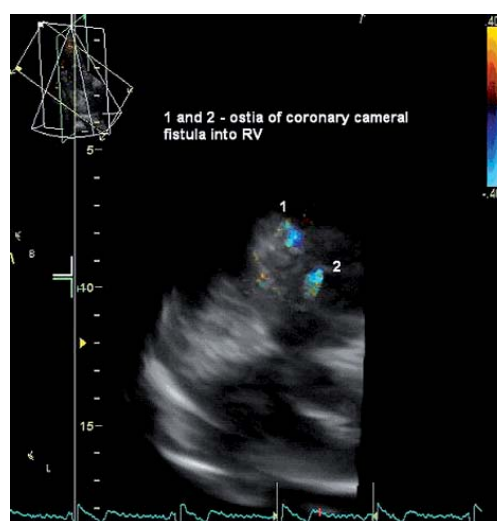


Figure 4. Real time three-dimensional imaging with 3D colour Doppler flow imaging. Two ostia draining into right ventricle chamber are marked with numbers 1 and 2

coronary arteries. A possible explanation may be coronary thrombosis with spontaneous reperfusion or coronary spasm unrelated or related to congenital abnormality. Common features besides the young age of these patients included predominance of female gender, smoking as a risk factor, and the lack of pre-infarction symptoms. Coronary thrombosis with subsequent recanalisation of the infarct-related vessel has been proposed as a possible mechanism [12, 13]. Minimal intraluminal atherosclerosis missed by conventional angiography has also been considered a possible contributing factor. Coronary artery spasm is another potential mechanism that has been noted to play a role in some case reports [14]. However, in the majority of cases and also in our patient, the proof for its existence was difficult to gather.

Conclusion

In conclusion, we present a case of a patient admitted with acute coronary syndrome and double-orifice RCA to right ventricle coronary fistula as the only coronary abnormality detectable in coronary angiography. We were able to demonstrate a unique additional value of real time 3D echocardiography in detection of fistula drainage into the RV and localisation of its ostia.

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