

An unpleasant surprise in the setting of primary percutaneous coronary intervention: diffuse and severe vessel ectasia with acute thrombosis of the distal right coronary artery in a patient with acute inferior myocardial infarction

Diego Milazzo, Giuseppe Caramanno, Piero Innocente, Rita La Mantia, Ignazio Vaccaro

Catheterization Unit, Division of Cardiology, San Giovanni di Dio Hospital, Agrigento, Italy

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Coronary artery ectasia is defined as a > 1.5-fold dilation of the coronary artery compared to the diameter of adjacent normal segments. It must be distinguished from discrete aneurysms that appear in areas adjacent to coronary artery stenosis. It is usually considered a variant of coronary atherosclerosis. Dilated segments are thought to modify the rheology of blood, sluggish or turbulent flow predisposing to myocardial ischemia and its sequelae, including myocardial infarction and sudden death. We report the case of a 52-year-old man, light smoker, with arterial hypertension and family history of coronary artery disease, who was referred to our coronary care unit for an inferior ST-elevation acute myocardial infarction and presented with severe and diffuse vessel ectasia and right coronary thrombosis at coronary angiography.

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Address:

Dr. Diego Milazzo
Servizio di Emodinamica
U.O. di Cardiologia
A.O. San Giovanni di Dio
Contrada Consolida
92100 Agrigento
E-mail: D.Milazzo@lycos.com

Case report

A 52-year-old man, light smoker, with arterial hypertension treated with angiotensin-converting enzyme (ACE) inhibitors and family history of coronary artery disease, was referred to our coronary care unit symptomatic for typical persistent ischemic chest pain (symptom onset-admission time about 2 hours) associated with ST-segment elevation in the ECG inferior leads. On physical examination there were no signs of hemodynamic impairment; the systemic arterial pressure was 120/80 mmHg and heart rate was 58 b/min.

According to our protocol, the patient was given i.v. unfractionated heparin (5000 IU), i.v. acetylsalicylic acid (250 mg), i.v. omeprazol and i.v. abciximab in bolus + infusion, according to the patient's weight.

Our interventional cardiologist team was immediately activated and an urgent coronary angiography with a percutaneous coronary intervention (PCI) was performed (door-to-balloon time about 30 min).

All three major coronary branches were severely aneurysmal, with turbulence and slow flow despite normal arterial pressure

(Figs. 1 and 2); the right coronary artery was occluded in the distal tract, with evidence of extensive endoluminal thrombosis (Fig. 3). A quantitative coronary angiography analysis documented these diameters in the proximal tracts of the coronary tree (anterior descending artery 13 mm, left circumflex artery 6 mm, right coronary artery 10 mm).

Despite passing the guide-wire (Hitorque BMW, Guidant) relatively easily through the site of occlusion and multiple balloon inflations (Aqua 2.0 × 20 mm, Cordis; Mercury 3.0 × 20 mm, Abbott), no reperfusion was obtained (Figs. 4 and 5).

During the procedure the patient's symptoms were spontaneously disappearing, and on the basis of his good hemodynamic status, with evolution of the ECG ischemic alterations toward a limited necrosis pattern without involvement of the right ventricle both at ECG and at echocardiography, and of challenging coronary anatomy, we decided to stop the procedure.

We preferred not to use a thromboaspirator device due to the aneurysmal condition, often correlated with frailty of the vessel wall, and due to distal site of occlusion.



Figure 1. Left coronary artery, left anterior oblique projection. CX = left circumflex artery; DA = anterior descending artery.

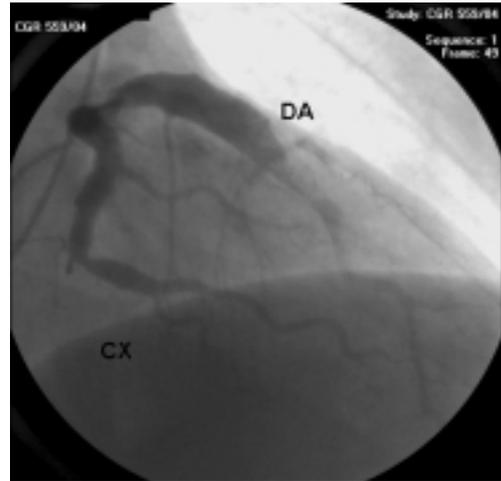


Figure 2. Left coronary artery, right anterior oblique projection. CX = left circumflex artery; DA = anterior descending artery.

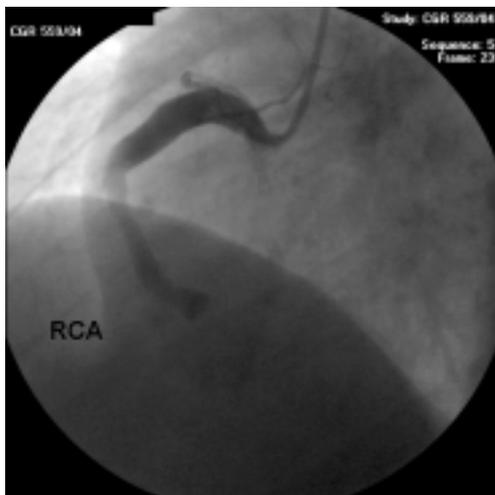


Figure 3. Right coronary artery (RCA), left anterior oblique projection.

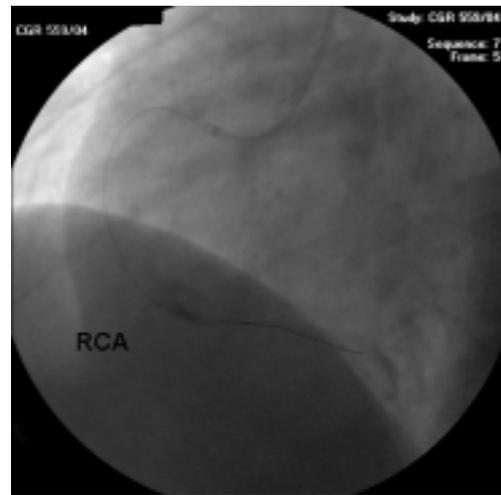


Figure 4. Right coronary artery (RCA), left anterior oblique projection at balloon inflation.

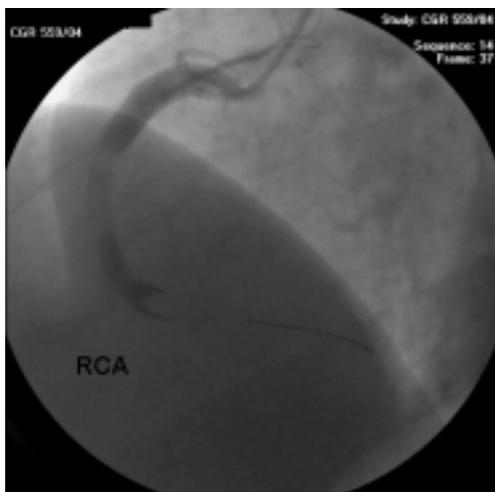


Figure 5. Right coronary artery (RCA), left anterior oblique projection, final result.

During the hospital stay, the patient was anticoagulated with unfractionated heparin and then switched to oral anticoagulants, in association with clopidogrel.

On discharge, after 9 days, the patient was asymptomatic and in good hemodynamic conditions; the ECG showed QS in D3 and small Q wave in D2 and aVF; an ultrasound evaluation showed inferior akinesia with ejection fraction of 50%, normal size of the aortic root and of the thoracic and abdominal aorta.

At 5-month follow-up the patient is asymptomatic and keeps on taking oral anticoagulants, clopidogrel, ACE-inhibitors and statins. An ergometric test documented no signs of ischemia.

Discussion

Coronary artery ectasia is defined as a > 1.5-fold dilation of the coronary artery compared to the diameter

of adjacent normal segments and its prevalence ranges between 0.3 to 5.3% depending on series¹. It must be distinguished from discrete aneurysms that appear in areas adjacent to coronary artery stenosis.

It is usually considered a variant of coronary atherosclerosis¹ but has also been described as an isolated congenital lesion² or in association with syphilis³, congenital heart disease⁴, scleroderma⁵, polyarteritis nodosa⁶, Ehlers-Danlos syndrome⁷, bacterial infections⁸, Kawasaki syndrome⁹, vascular trauma¹⁰, and also exposure to herbicides¹¹.

Male gender, smoking, systemic hypertension and familial hypercholesterolemia¹² seem to be associated with an increased prevalence of coronary artery ectasia, while diabetes mellitus, a condition characterized by increased arterial stiffness, was found to be inversely associated with it¹³, thus prompting someone to consider coronary ectasia as an extreme form of positive remodeling¹⁴.

Dilated segments are thought to modify the rheology of blood, sluggish or turbulent flow predisposing to myocardial ischemia and its sequelae, including myocardial infarction and sudden death, even when ectasia is not associated with coronary artery stenosis (22% of cases in a recent large Spanish series)¹⁵.

In the majority of cases, coronary ectasia affects one vessel, mostly the right coronary artery, but in about one third of cases it is diffuse to all three major coronary branches¹⁵.

The underlying pathophysiologic mechanism is unclear; it has been proposed that the action of different risk factors based on genetic predisposition leads to initial endothelial damage, activating a series of inflammatory mediators (macrophages, metalloproteinases, etc.) that cause degeneration of the medial layer of the vessel.

Polymorphism of HLA class II genes may be involved in this process, as HLA-DR B1 and DQ B1 genotypes have been reported more frequently in patients with coronary artery ectasia¹⁶. Moreover, an insertion/deletion polymorphism of the ACE gene, the DD genotype, has been associated with this condition¹⁷.

Recently several studies have suggested a role for inflammatory processes, documenting a significant increase in the levels of C-reactive protein¹⁸ and of interleukin-6¹⁹ in patients with coronary artery ectasia.

The prognosis of patients with coronary artery ectasia is controversial. One of the first studies reported a 13% mortality rate over 3 years in patients treated medically, due to sudden rupture of a coronary artery or thrombosis and myocardial infarction²⁰, but, more recently, several studies have documented a good prognosis of ectasia itself if not associated with significant coronary stenosis²¹.

No recommendations exist to determine the optimal therapeutic regimen, so it should be based on known pathophysiology and extrapolated from treatment of

obstructive atherosclerotic coronary artery disease. The most common treatments, as reported in the literature, are antiplatelet therapy with aspirin, with or without ticlopidine or clopidogrel, anticoagulant therapy with warfarin, or a combination of antiplatelet and anticoagulant agents.

In our case we preferred to use clopidogrel and warfarin (INR between 2.0 and 3.0) due to the comparable, if not superior, efficacy of clopidogrel with respect to aspirin, and due to its slightly superior safety²².

In the occurrence of thrombosis and myocardial infarction in a case of coronary artery ectasia, current reperfusion options, such as intravenous or intracoronary thrombolysis, PCI with or without stenting, thrombectomy and thromboaspiration with several devices, and distal protection with filters, have proven to be of poor efficacy, probably due to the greater thrombotic burden that increases the risk of distal embolization and no-reflow phenomenon, thus prompting the search for new approaches²³.

A recent report documented a case of thrombosis of an ectatic right coronary artery treated successfully by means of a modified 6F Judkins right diagnostic catheter as a thromboaspiration system²⁴. However this intriguing technique needs further evaluation, due to the risk of major complications, such as collapse of the coronary artery, injury of the arterial wall or even embolism of large thrombotic fragments in the aorta.

In our case, we started aggressive antiplatelet and anticoagulant therapy, with aspirin, clopidogrel, abciximab and unfractionated heparin; we tried to obtain mechanical reperfusion by PCI within reasonable limits of safety, as the risk of dissection or rupture of the arterial wall and of proximal extension of thrombosis was high in such a peculiar scenario.

After the failure of many attempts, we preferred to stop the procedure, as the patient's symptoms were disappearing, with evolution of the electrocardiographic ischemic alterations toward a limited necrosis pattern without involvement of the right ventricle, having considered the good hemodynamic status with no signs of hypotension, congestive heart failure or electrical instability.

We decided not to proceed to thromboaspiration due to the extension and site of thrombosis and due to the challenging coronary anatomy, as coronary artery ectasia is often correlated with frailty of the vessel wall and, in our opinion, it was hazardous to reach the site of occlusion with a quite stiff device, in a very distal point of the vessel.

Intravenous or intracoronary "rescue" thrombolysis with half-dose alteplase, in a patient already being treated with abciximab, could be an attractive option, but currently this treatment is neither codified by guidelines nor supported by consistent clinical studies in this specific condition, after failed PCI in an aneurysmal coronary artery with a massive thrombotic burden; indeed the indication was questionable, given the reduced

expected benefit of fibrinolytics due to the extension of the thrombosis, the risk of major bleeding associated with combination therapy with abciximab, the distal site of occlusion with small amount of injured myocardium and, finally, the absence of right ventricular involvement and other markers of elevated risk that could justify a more aggressive approach²⁵.

The appropriateness of our therapeutic strategy has been confirmed by an uneventful hospital course, with only a modest reduction in left ventricular systolic function, and by the patient's good conditions both at discharge and at follow-up, with absence of inducible ischemia at 5 months.

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