

CASE REPORT

Extensively Thrombosed Ectatic Circumflex Coronary Artery Fistula Presenting as Acute Coronary Syndrome

Nooraldaem Yousif^{1,#}, Mohammady Shahin^{1,#}, Robert Manka¹ and Slayman Obeid^{1,*,#}

¹Department of Cardiology, University Heart Center, University Hospital Zurich, Zurich, Switzerland

Abstract: Background: Coronary artery fistula (CAF) is an abnormal communication between the termination of a coronary artery or its branches and a cardiac chamber, a great vessel or other vascular structure. Symptomatic patients with large CAF should undergo surgical or percutaneous closure of the fistula at the drainage site while still the debate on closing asymptomatic CAF and re-opening symptomatic occluded CAF is ongoing.

Case Summary: We are reporting a 30-year-old male patient with no previous medical history presented as non-ST segment elevation myocardial infarction. Coronary angiography showed an entirely thrombosed ectatic circumflex artery with a suspicion of thrombosed coronary arterial fistula. In view of the ongoing ischemia in the setting of acute coronary syndrome; we tried to open percutaneously but all efforts were to no avail.

Discussion: In this case report, we are sharing our experience in the management of this challenging case in view of the rarity of such peculiar clinical condition and the unfavourable presentation along with the lack of clear-cut Guideline and Consensus whether to/not to open such huge and immensely thrombosed symptomatic coronary artery fistula as well as the dilemma of choosing the best long-term medical treatment between antiplatelets vs anticoagulants in such young patient.

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1. INTRODUCTION

Coronary artery fistula (CAF) is an abnormal communication between the termination of a coronary artery or its branches and a cardiac chamber, a great vessel or other vascular structure [1]. Most of the cases are congenital while acquired cases are mainly seen after cardiac surgery with an overall estimated prevalence of 0.002% in the general population and 0.05-0.25% in patients who undergo invasive coronary angiography (ICA) [2]. However, the incidence of CAF is highly speculative since a significant proportion is clinically silent and detected incidentally [2, 3]. CAF terminating in the right heart chambers accounts for about 60% of the cases [3]. The vast majority of patients are asymptomatic with a benign clinical course while minority of patients may become symptomatic and may present with myocardial ischemia due to possible persistent or episodic steal of blood flow to the fistulous tract from the normal coronary branches, leading to ischemia and rarely to infarction; another explanation can be due to stenosis and eventually occlusion secondary to thrombosis, ulcerations and atherosclerosis associated with fistulous tracts [1-3].

Symptomatic patients with large CAF should undergo surgical or percutaneous closure of the fistula at the drainage site while still the debate on closing asymptomatic CAF and re-opening symptomatic occluded CAF is ongoing [4]. We present a case of fully thrombosed RCX-Fistula in a 30 year old patient presenting as NSTEMI with severe chest pain.

2. CASE PRESENTATION

A 30-year-old male patient (F.H.) with no previous medical history presented to the emergency room at 2:00 am after 3 days of persistent ongoing typical chest pain. He reported similar episodes of angina pectoris within the last two and half years, *i.e.* at 27 years of age, of same quality, lasting between 10-15 seconds, with the longest pain being intermittent over a 3-day period. However, for the last 6 month he has been pain free.

Physical exam was unremarkable except for elevated blood pressure of 170/90 mmHg in both arms. The electrocardiogram showed minimally diffuse ST segment depressions of 0.5mm in the lateral leads (I, aVL, V3-V6) (Fig. 1a). Blood tests revealed an elevated creatinine phosphokinase (CPK: 581U/I, normal range: <196U/I), and elevated high sensitivity troponin T (hsTNT: 0.954 ng/ml, normal range: <0.014 ng/ml), while myoglobin and CK-MB were in the normal range (Fig. 1b).

*Address correspondence to this author at the Interventional cardiologist, University Heart Center, University Hospital Zurich, Zurich, Switzerland; Tel: +41 77 960 76 76; Fax: +41 44 255 01; E-mail: Slayman.Obeid@usz.ch

[#]Equal contribution



Fig. (1). a. The electrocardiogram showed minimally diffuse ST segment depressions of 0.5mm in the lateral leads (I, aVL, V3-V6). b. Normalization of ECG on third day of admission, 48 hours after continuous administration of the glycoprotein IIb/IIIa inhibitor Abciximab.

Accordingly, the patient was diagnosed with Non-ST-Elevation Myocardial Infarction (NSTEMI), loaded with 500mg of acetyl salicylic acid, a bolus dose of heparin (5000 IU) and underwent a coronary angiography, which showed an occlusion of what seemed a markedly aneurysmatic circumflex artery with a diameter of up to 6.2mm just after the bifurcation of the posterolateral branch. (Fig. 2a, Moving Image 1). Due to the large thrombus burden, wiring of the circumflex ended up being a frustrating and cumbersome procedure even with balloon back-up (Fig. 2b). After successful placement of the wire in the distal circumflex, multiple attempts to manually aspirate and thereafter dilate the blood vessel with a compliant 4.0x20mm balloon failed to open the occluded artery (Fig. 2c and Moving Image 2 and 3). Hence, the procedure was stopped and a loading dose of prasugrel of 60 mg was given along with a continuous intravenous infusion of the glycoprotein IIb/IIIa inhibitor eptifibatide.

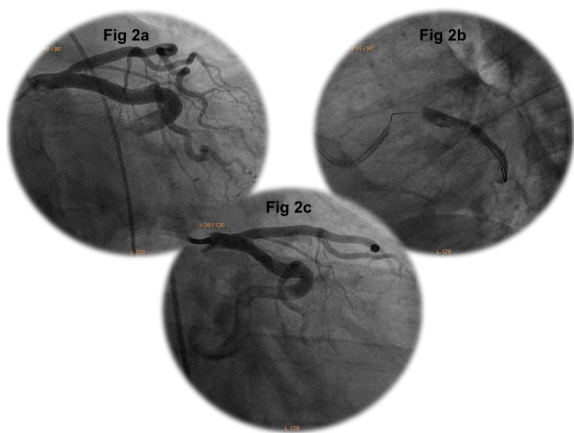


Fig. (2). a. Coronary angiogram showing occluded aneurysmatic circumflex artery. b. Large thrombus burden, wiring of the circumflex failed initially even with balloon back-up. c. After successful placement of the wire in the distal circumflex, multiple attempts to manually aspirate and dilate the blood vessel failed to open the occluded artery.

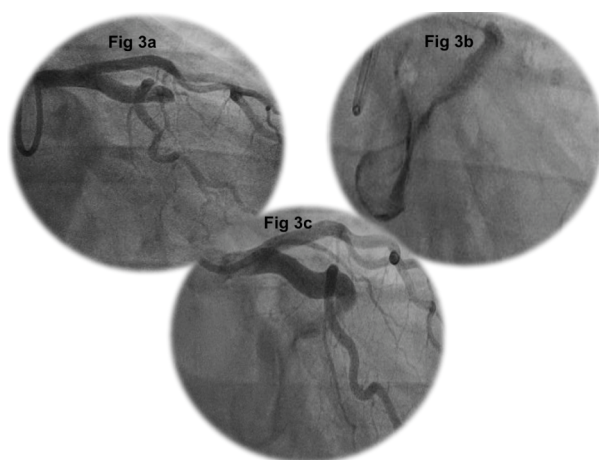


Fig. (3). a. Second look angiography documented persistent occlusion of the circumflex artery. b. Injection through the microcatheter showed a completely thrombosed circumflex artery along its runoff. c. Manual aspiration and repeated balloon again failed to open the artery.

The two operators on site re-evaluated the available options for further management. i) Conservative treatment with oral anti-coagulation, ii) Conservative treatment with dual antiplatelet therapy alone, iii) Continuous infusion of glycoprotein IIb/IIIa inhibitor and a second trial to attempt opening the vessel percutaneously.

Twelve hours later, a second look angiography documented a persistent occlusion of the circumflex artery (Fig. 3a, Moving Image 4). Hence, a second attempt to open the occluded artery was undertaken. With the use of a Fielder FC wire (Terumo, Japan) and the help of a Finecross Microcatheter (Terumo, Japan) we were able to place the guidewire in a very distal portion of this hugely aneurysmatic and tortuous vessel. Injection through the microcatheter showed a completely thrombosed circumflex artery along its runoff (Fig. 3b, Moving Image 5). Manual aspiration and

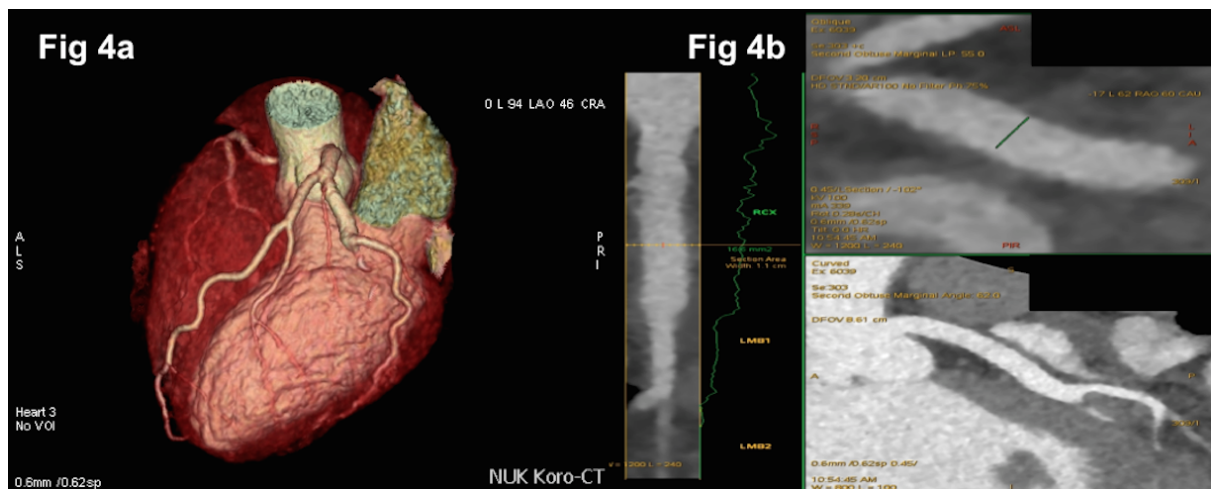


Fig. (4). a and b. Coronary CT scan with i.v. contrast performed on the third day of admission, 48 hours after continuous administration of the glycoprotein IIb/IIIa inhibitor Abciximab documented a persistently occluded circumflex artery.

repeated balloon dilatations with NC Quantum Maverick and Maverick balloon (Boston Scientific, 300 Boston Scientific Way Marlborough) again failed to open the artery (Fig. 3c, Moving Image 6) At this point, a bolus dose of intracoronary Abciximab (Reopro[®]; Janssen-Cilag Zug, Schweiz) was administered distally through the Finecross[®] and a continuous intra-venous perfusion was planned.

Echocardiography done after the procedures during the index hospitalization revealed a normal left ventricular ejection fraction of 57% with hypo-kinesis of the infero- and anterolateral wall as well as a right ventricle of normal size and function (TAPSE=21mm) with no evidence of pulmonary hypertension Furthermore, a coronary CT scan with i.v. contrast performed on the third day of admission, 48 hours after continuous administration of the glycoprotein IIb/IIIa inhibitor Abciximab and of normalization of the ECG (Fig. 1b), continued to documented a persistently occluded circumflex artery with no hint to whether the artery ended as a distal coronary fistula. (Fig. 4a and 4b).

Subsequent to an interdisciplinary discussion regarding the treatment strategy in a thrombosed circumflex-arterial fistula, the asymptomatic patient was discharged 4 days after an uneventful hospitalization on 100 mg of acetylsalicylic acid and 10 mg of prasugrel for 12 months.

After discharge, the patient underwent an extensive hematological work-up, which showed no evidence of thrombophilia or of a hypercoagulable state (Table 1). Additionally, cardiac MR showed a mildly depressed systolic LV function with inferior and inferolateral, basal akinesia. T2 weighted images confirmed myocardial edema (recent infarction) (Fig. 5) as well as late gadolinium enhancement exposed transmural scarring (Fig. 6).

3. DISCUSSION & CONCLUSION

Coronary artery fistulas (CAF) accounted for less than 3% of congenital cardiac anomalies [5]. Although most are congenital; acquired cases are infrequently seen following trauma from angiography, endomyocardial biopsy, open heart surgery and pacemaker implantation. Although CAF

carries a potential risk of cardiac morbidity and mortality; Myocardial infarction caused by CAF is an extremely rare event and the majority of cases were reported in order to answer the traditional question whether to/not to close symptomatic CAFs [2-5]. Only very scarce cases in the literature were discussing the emerging dilemma whether to/not to open occluded symptomatic CAF; thus we ended up with no definitive guideline how to optimally tackle these cases [6]. In our patient we couldn't figure out the final destination of this aneurysmatic RCX-Fistula due to the distally occluded

Table 1. Hematological work-up.

Factors	Result
Factor V laden mutation	-ve
Anti-thrombin mutation	-ve
Protein C	-ve
Protein S	-ve
Congenital Dysfibrinogenemia	-ve
Protein electrophoresis	NI
Antiphospholipid syndrome	
Lupus anticoagulant(LA1,LA2)	-ve
Anti-Cardiolipin	-ve
Anti-beta 2 glycoprotein	-ve
Autoimmune disease	
ANA	-ve
ANCA	-ve
Anti DNA	-ve
Myeloproliferative neoplasia(JAK2-V617F)	-ve
Paroxysmal nocturnal hemoglobinuria	-ve

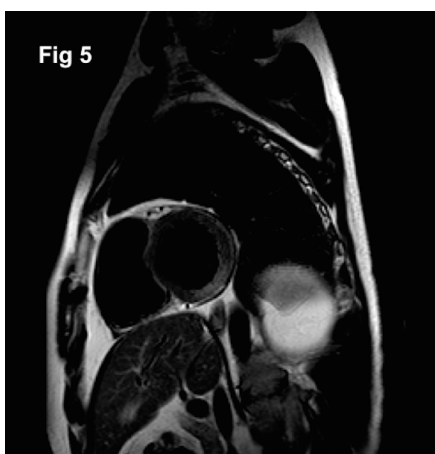


Fig. (5). T2 weighted images confirmed myocardial edema (recent infarction).

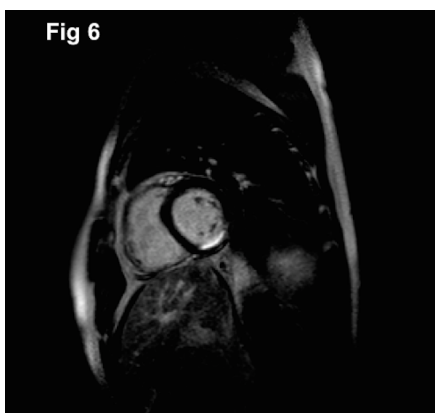


Fig. (6). Late gadolinium enhancement showed transmural scarring.

segment, On the other hand this explains nicely the absence of murmur during the initial clinical examination as the high thrombotic burden obstruct the drainage of the Fistula to a cardiac chamber, but its indubitable that the Fistula was used to provide a supply to the myocardium as this is the only rationale that explains the acute infarct.

The pursue behind Interventions for coronary fistula with objective evidence of myocardial ischemia and steel syndrome is to provide long-term event-free survival either by transcatheter interventions (Coiling, Embolisation, Occluder Devices) or surgical closure utilizing external placcation on the beating heart or by intracardiac closure using cardiopulmonary bypass with or without cardiac arrest [7, 8]. A paradoxical scenario is seen in our patient as the first presentation of this occluded coronary fistula was an ongoing ischemia in the setting of acute coronary syndrome, so the dilemma was whether to open or not! Eventually, we tried (fortunately unsuccessfully) to revascularize the occluded segment overlooking the risk of manipulation within the thrombosed fistula which may have caused the risk of systemic embolization as the thrombus was likely to protrude and dislodge into heart chambers [9].

In view of the rarity of such peculiar finding and the challenging presentation along with lack of clear Guidelines and Consensus whether to/not to open such huge and im-

mensely thrombosed coronary artery fistula as well as the dilemma of choosing the best long-term medical treatment between antiplatelets vs anticoagulants in such young patient.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Not applicable.

HUMAN AND ANIMAL RIGHTS

No Animals/Humans were used for studies that are the basis of this research.

CONSENT FOR PUBLICATION

Not applicable.

FUNDING

None.

CONFLICT OF INTEREST

The authors declare no conflict of interest, financial or otherwise.

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SUPPLEMENTARY MATERIAL

Supplementary material is available on the publisher's web site along with the published article.

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