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CASE REPORT
Coronary heart disease

Two presentations of acute coronary syndrome with progression of giant right coronary artery aneurysm; a case report

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Background Giant coronary aneurysms are a rare finding on coronary angiography. Given its very low prevalence, little is known about optimal management of this coronary pathology.

Case summary In this case report, we review the two presentations of a patient with acute coronary syndrome during a 6-year period. With regard to the second presentation, we review the investigations that demonstrate the progression of a coronary aneurysm in the right coronary artery as well as the Heart Team evaluations that resulted in surgical treatment of the coronary aneurysm.

Discussion Following perspectives on prevalence and risk factors, we emphasize upon the available data with regard to interventional options in coronary aneurysms and describe the considerations with regard to interventional treatment in patients with giant coronary aneurysms. Finally, we discuss the available literature with regard to antithrombotic regimens in patients with coronary aneurysms.

Keywords case report • coronary aneurysm • acute coronary syndrome • coronary artery bypass graft

ESC Curriculum 3.1 Coronary artery disease • 7.5 Cardiac surgery

Learning points

- Clinical decision-making on the treatment of coronary artery aneurysms is guided by observational studies, whereas data on giant coronary artery aneurysms are limited by case reports.
- As for the giant coronary aneurysms, this case adds data to the available literature that bypass surgery with aneurysm ligation is a plausible modality for coronary blood flow restoration and thrombotic risk reduction.
- The role of anticoagulation therapy in patients with surgically treated coronary aneurysms is not well-defined and warrants randomized evaluations.

Introduction

A coronary artery aneurysm (CAA) is an abnormal dilatation in the diameter of a localized segment of a coronary artery. A dilatation of 1.5 times the adjacent non-affected coronary artery is generally used as definition for CAA.[1] Definitions for giant coronary artery aneurysm (GCAA) include but are not limited to a four-fold increase in the reference diameter or a diameter exceeding 20 millimeters.[2,3] A GCAA is very incidental finding with a prevalence of only 0.02% in a population with a clinical indication for invasive coronary angiography.[2]

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A CAA may lead to myocardial infarction, which can occur in the absence of atherosclerotic disease due to distal microembolization or prolonged depression of coronary flow.[3,4] A CAA does not necessarily cause symptoms, though giant aneurysms are believed to be symptomatic more frequently.[3] We report the case of a patient with progression of a GCAA leading to myocardial infarction, who underwent successful ligation and coronary bypass surgery.

Timeline

Day	Event
January 2015 (first presentation)	Admission with clinical and electrocardiographical signs of ST-elevation myocardial infarction; balloon dilatation and thrombus aspiration performed. Aneurysms in all epicardial coronary arteries
June 2021 (second presentation)	Admission with clinical and biochemical signs of non-ST elevation myocardial infarction
0	Coronary angiography shows progression to a giant aneurysm in right coronary artery
1	Heart Team (1): options include percutaneous coronary intervention with covered stent or surgical ligation of aneurysm and coronary artery bypass grafting
3	Additional imaging with transthoracic echocardiography and computed tomography of coronary arteries with 3D-reconstructions
5	Heart Team (2): preference for surgical ligation of aneurysm and coronary artery bypass grafting with venous graft
11	Transfer to cardiothoracic centre
12	Uncomplicated surgical ligation of aneurysm and coronary artery bypass grafting with venous graft
15	Post-operative discharge from Intensive Care Unit
19	Discharge from cardiology ward
67	Outpatient clinic visit: no complications occurred following discharge

Clinical case

A 74-year-old Caucasian woman without allergies or cardiovascular risk factors was hospitalized due to inferior ST-elevation myocardial infarction in 2015. Coronary angiography showed an aneurysmatic aspect of all coronary arteries. The right coronary artery (RCA) was occluded distal to a large aneurysm (see Supplementary material online, *Supplementary Video S1*). Balloon dilatation and aspiration of red thrombus was performed. Due to distal embolization, tirofiban was administered, which resulted in relief of chest pain and resolution of the ST-segment elevation. Upon discharge, medical treatment included dual antiplatelet therapy (DAPT), statin, and beta blocker. In the subsequent years, there were no anginal symptoms.

Six years after the aforementioned presentation, our patient called the emergency services because of typical retrosternal chest pain with radiation to the left arm. Upon hospital presentation, the heart rate was 61/min and the blood pressure was 146/90 mmHg. Physical examination including auscultation of heart and lungs was unremarkable. Sublingual administration of nitrates did not result in resolution of the anginal symptoms. Electrocardiography showed sinus rhythm with a right bundle branch block (*Figure 1A*) with similar ST-segments as compared to prior electrocardiography (*Figure 1B*). High-sensitive troponin I was elevated (130 ng/L, upper limit of normal 47 ng/L). Due to persisting chest pain despite intravenous nitroglycerin, transfer to the catheterization laboratory for urgent coronary angiography took place. The aneurysm in the RCA progressed to a giant aneurysm with limited contrast flow in the distal bed of the RCA (see Supplementary material online, *Supplementary Video S2* and *Figure 2*).

Evaluation with transthoracic echocardiography showed a left ventricular ejection fraction of 57% with an akinetic, aneurysmatic basal inferior segment, and surrounding hypokinetic segments (see Supplementary material online, *Supplementary Videos S3 and S4*). Computed tomography of the coronary arteries showed an aneurysm dimension of 26 × 30 millimeters (*Figure 3*).

Following evaluation in the Heart team, a surgical intervention was preferred to a percutaneous approach. Ligation of the aneurysm and distal revascularization with a venous graft was performed without complications. Medication upon discharge, seven days post-operative, included single antiplatelet therapy with acetylsalicylic acid, as well as statin treatment and beta blockade. On the first outpatient clinic visit, there were no symptoms of angina pectoris.

Discussion

The prevalence of coronary aneurysms varies across registries from 0.35% in the international coronary artery aneurysm registry (CAAR) to 4.9% in the more historical data from The Coronary Artery Surgery Study (CASS).[1,2] In the CAAR, the prevalence of GCAA was 0.02%, the majority being located in the RCA and left anterior descending artery.[2,5]

Aetiology and risk factors

Atherosclerosis is thought to be the cause of CAA in about half of the cases, which was also the presumed cause for the GCAA in our patient. A potential pathophysiologic mechanism might be the increased intraluminal pressure in atherosclerotic vessels. Other, less frequently reported causes include congenital aneurysms (20–30%), Kawasaki disease, connective tissue disorders, infectious causes, and iatrogenic causes.[3–5]

Treatment and outcome

The optimal management of CAAs is not well-defined, given the paucity of data with regard to natural history and long-term outcomes. Furthermore, the low prevalence hampers acquisition of randomized data. Therefore, clinical decision-making is guided by predominant observational data.

In a comparison of anticoagulation therapy to DAPT, data from a retrospective cohort showed no differences in outcome.[5] In patients with CAA, stenting in the setting of an acute myocardial infarction might be deferred due to the increased risk of no-reflow and incomplete stent apposition with consequent restenosis.[6–8]

After a median follow-up of 62 months in 458 patients with CAA, favourable outcome on cardiovascular and cerebrovascular events was observed among patients who underwent surgical intervention as compared to medical management.[5] Importantly, these findings should be interpreted with caution given the retrospective and observational design of the study. Within the CAAR population, no differences in adverse outcome (infarction, embolism, bleeding or mortality) were reported between the surgical and percutaneous study groups. Interestingly, as compared to drug eluting stent implantation, rates of restenosis were significantly higher in patients treated with a bare metal stent.[2] The use of a covered stent has also been described

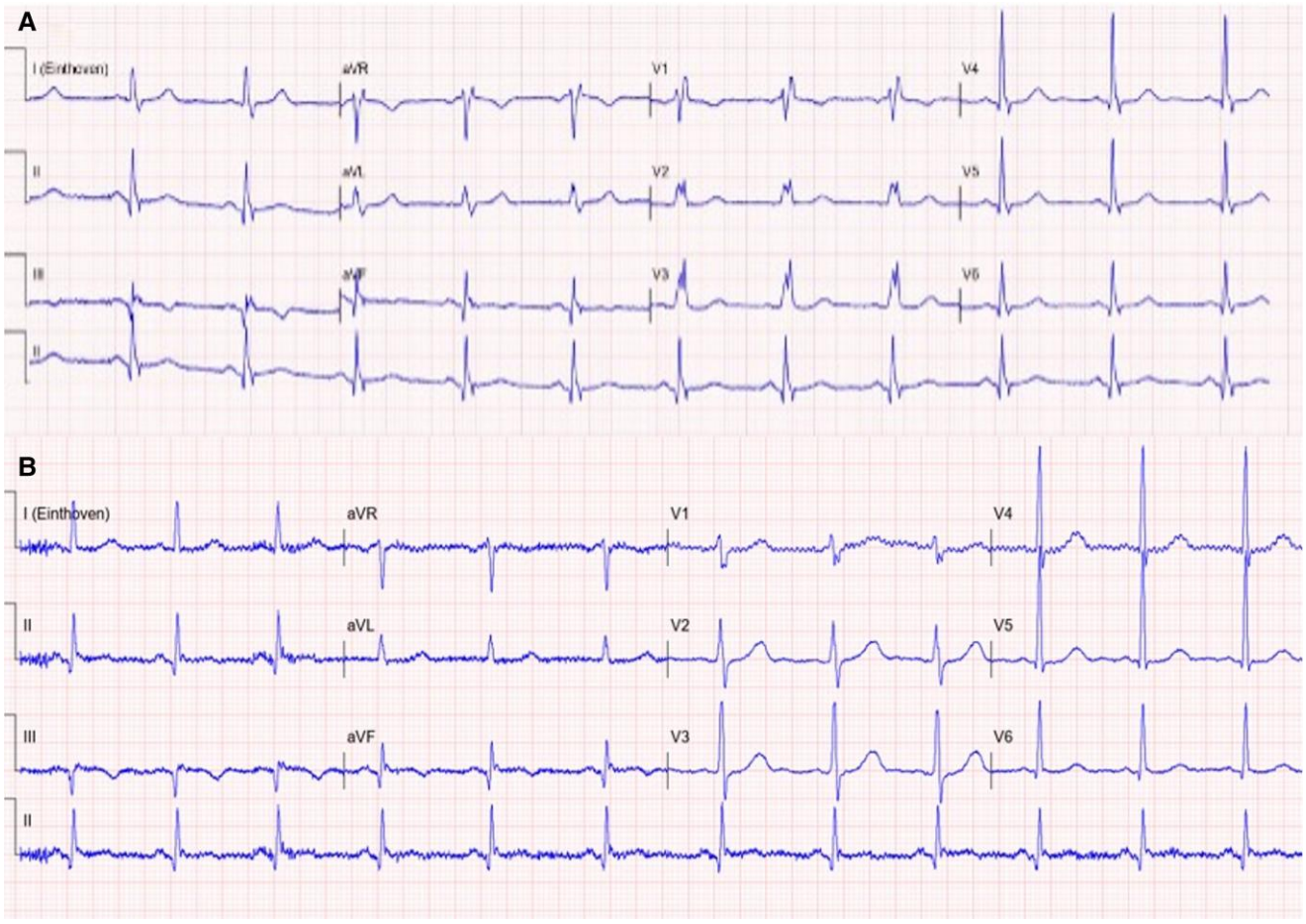

Figure 1 (*A*) Electrocardiography at the time of index presentation (2021). As compared to a prior registration (*B*), no new ST-segment elevation or T-wave inversion was observed. (*B*) Electrocardiography after percutaneous intervention (2015). The ECG has been derived from the medical record, patient provided informed consent.

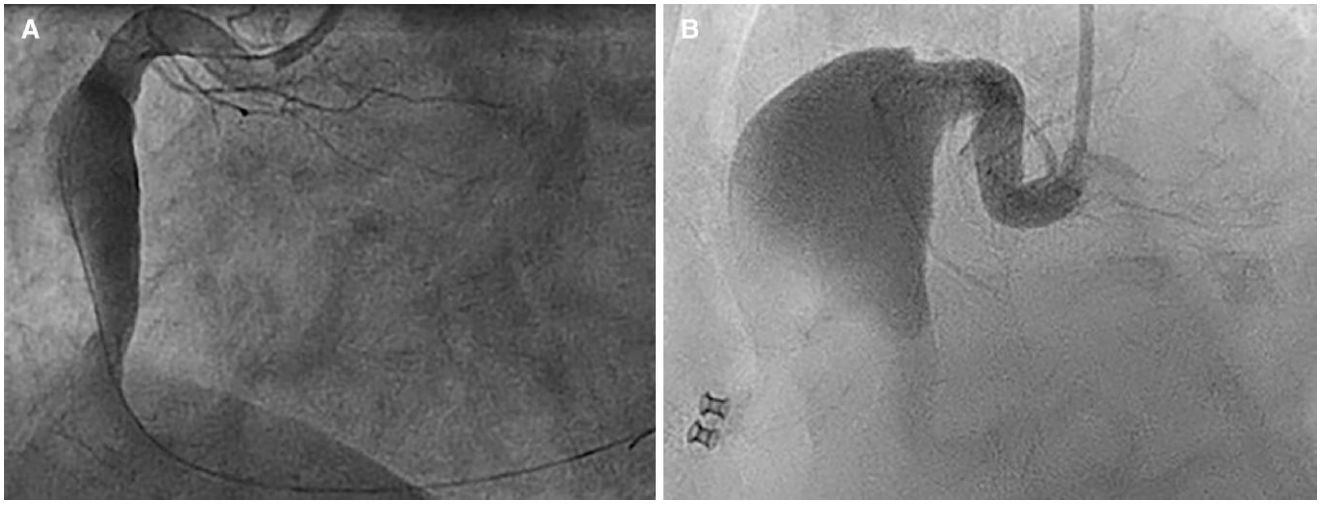

Figure 2 Angiographic images of the right coronary artery. (*A*) Coronary aneurysm on coronary angiography performed because of admission with ST-elevation myocardial infarction in 2015. (*B*) Coronary aneurysm on angiography performed because of non-ST segment elevation myocardial infarction in 2021, which showed progression of the aneurysm as compared to 2015.

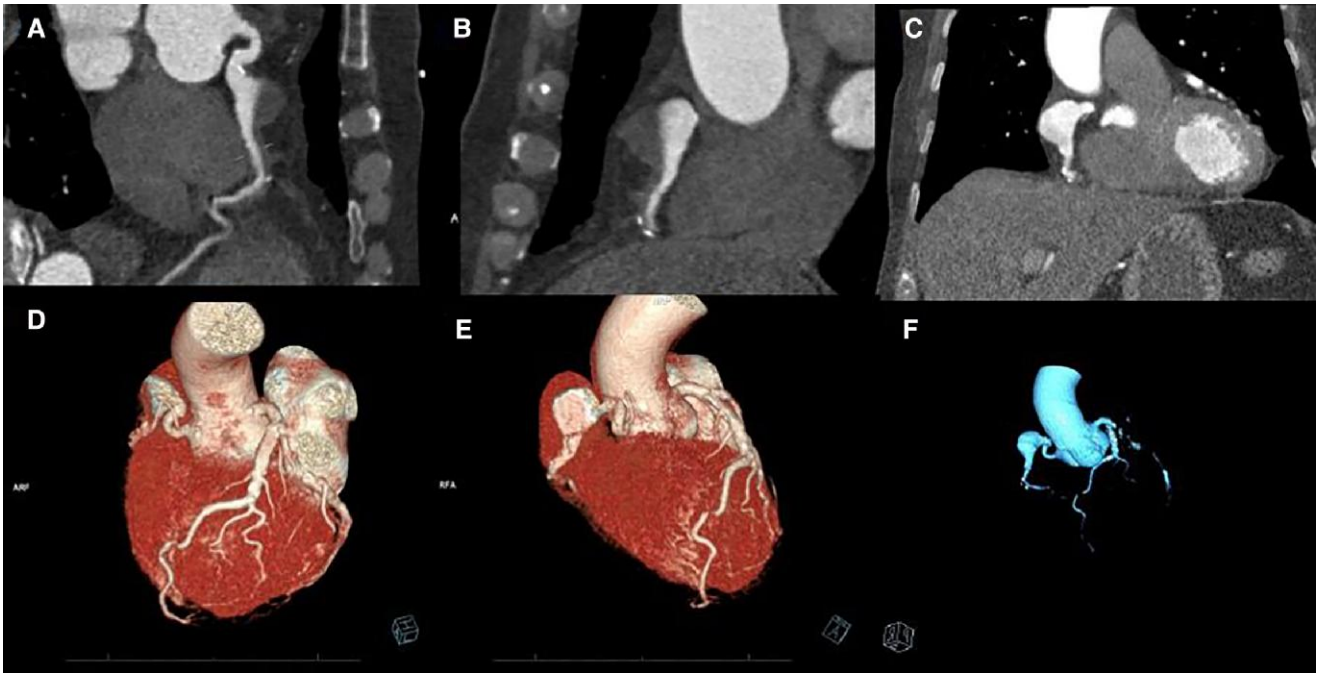

Figure 3 (*A–C*) Computed tomography imaging with incomplete contrast filling in the aneurysm. (*D–F*) 3D reconstructions of the aneurysmal pathology.

in patients with CAA to achieve aneurysm sealing resulting in acceptable outcomes.[9]

As for GCAA, available data concerning invasive treatment and antithrombotic strategies are derived from case presentations. From an empiric point of view, treatment decisions should include aneurysmal size, symptoms, and location of the aneurysm. Complications including thrombosis and distal embolization are believed to be higher in GCAA.

Surgical intervention, including ligation or resection of the aneurysm with concomitant bypass grafting, has been suggested as preferred strategy for GCAA.[10,11] In our patient, a saphenous vein was used as bypass graft for the RCA. The preferential use of an arterial graft as bypass for the RCA is not as straightforward as it is for the left anterior descending coronary artery. Interestingly, a systematic review and meta-analysis from the United Kingdom showed a non-significant trend towards better performance of venous graft in the right coronary system.[12]

Finally, the use of oral anticoagulation therapy, including the direct oral anticoagulants, has been described in a small number of cases.[13,14] Most insights have been derived from the CAAR population. In a propensity-matched (with or without oral anticoagulation) sample, the composite endpoint of myocardial infarction, unstable angina, and aneurysm thrombosis was significantly lower in the anticoagulation group (8.7% vs. 17.2%) after a median follow-up of three years.[15] Importantly, only 8% of the studied population was diagnosed with a GCAA. In addition, only 10% underwent bypass surgery and over 60% of the patients were treated with medical therapy. Therefore, generalizability to patients who underwent invasive treatment is limited. Our patient was discharged with single antiplatelet therapy following bypass surgery. In general, anticoagulation therapy might be considered in patients who are not suitable for invasive therapy, whereas the beneficial effects seem rather questionable in patients in whom the aneurysm is ligated or resected.

Conclusion

As of yet, insights with regards to coronary artery aneurysms are derived from observational studies, whereas data on the rare occurrence of giant coronary artery aneurysms are limited to case reports. Appreciating these limitations, invasive treatment of coronary artery aneurysms has not been related to increased adverse events. With regards to giant coronary artery aneurysms, surgical treatment may be preferred from a technical point of view. The presented case provides additional data to the available literature that cardiothoracic surgery with ligation and bypass grafting is a plausible consideration in patients with giant coronary aneurysms. There is no robust evidence that oral anticoagulation therapy results in lower rates of adverse events among patients who underwent invasive treatment. Especially in case of aneurysm ligation, routine use of oral anticoagulation therapy warrants randomized trials to establish the best therapeutic strategy.

Lead author biography

Stijn van Vugt is resident cardiology from the Radboud University Medical Center, Nijmegen, the Netherlands. In his third year of residency, he was trained in the Jeroen Bosch Hospital, 's-Hertogenbosch, the Netherlands. During his internship on the catheterization room, he encountered the described case.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written informed for submission and publication of this case report has been obtained from the patient in line with COPE guidance.

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