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CASE REPORT
Cardiac imaging

A pebble clogging a river: a case report of thrombosed coronary aneurysmal ectasia

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Background Coronary artery ectasia (CAE) is a rare anomaly that can present at any age. Predisposing risk factors include Kawasaki disease in a younger population and atherosclerosis in the older generation. We present a unique case of the management of a young woman diagnosed with multivessel CAE with aneurysmal changes in the setting of acute coronary syndrome and subsequently during pregnancy.

Case summary A 23-year-old woman presented with acute onset chest pain. Electrocardiogram revealed no ischaemic changes; however, troponin I peaked at 16 ng/mL (reference range 0–0.04 ng/mL). Echocardiogram showed apical dyskinesis with preserved left ventricular ejection fraction. Coronary angiography showed multivessel CAE along with significant thrombus burden in an ectatic lesion of the left anterior descending artery. Since the patient was haemodynamically stable, conservative management with dual antiplatelet therapy and anticoagulation was started. On follow-up, coronary computed tomographic angiogram illustrated resolution of the coronary thrombi and echocardiogram showed improvement to the apical dyskinesis. It was presumed that Kawasaki disease was the most likely aetiology of her disease. Subsequently the patient reported that, contrary to medical advice, she was pregnant, adding another layer of complexity to her case.

Discussion Coronary artery ectasia can be discovered as an incidental finding or can present with an acute coronary syndrome. Management is challenging in the absence of randomized trials and large-scale data. Treatment options include medications, percutaneous intervention, and surgical revascularization. Close surveillance is required in these patients to assess progression of disease. Here we discuss treatment options during acute coronary syndrome and pregnancy.

Keywords Coronary artery ectasia • Coronary aneurysm • Acute coronary syndrome • Coronary angiography • Three-dimensional coronary imaging • Case report

Learning points

- It is prudent to not dismiss chest pain in any young woman with no risk factors.
- We hope to bring attention to the importance of medical therapy until failure with coronary artery ectasia.
- Disease management is case-oriented when dealing with this rare diagnosis.

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Introduction

Coronary artery ectasia (CAE) is defined as dilation of an arterial segment to a diameter at least 1.5 times that of the adjacent normal coronary artery. The key difference between CAE and coronary aneurysm is the diffuse involvement of the affected artery. Predisposing risk factors include Kawasaki disease in younger populations and atherosclerosis in the older generation. The prevalence of CAE is unknown, but it is thought to vary between 0.3% and 4.9% of patients receiving coronary angiograms.[1,2] We present a unique case of a young woman diagnosed with multivessel CAE with aneurysmal changes in the setting of acute coronary syndrome.

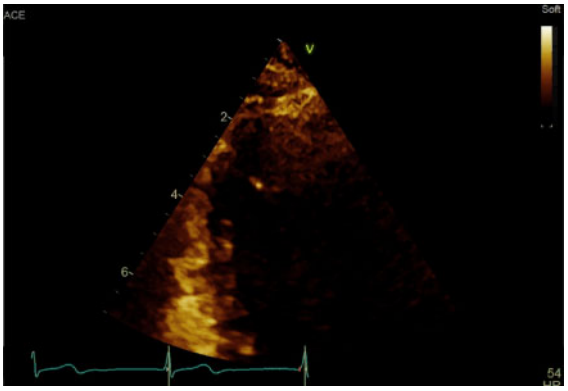
Timeline

July 2018	Patient presented to emergency room with chest pain, troponin elevation, and regional wall motion abnormality on echocardiogram. Coronary angiography showed triple vessel coronary ectasia with aneurysmal changes to the left anterior descending artery with large thrombus. Patient was started on warfarin, clopidogrel, and aspirin
August 2018	One-month follow-up. Computed tomography (CT) coronary angiogram showed complete resolution of thrombosis. Diffuse coronary ectasia and aneurysmal changes seen on CT. Three-dimensional reconstruction obtained
September 2018	Contrary to medical advice, the patient became pregnant; warfarin was discontinued
April 2019	Aspirin and clopidogrel were discontinued 5 days prior to delivery, and the patient delivered a healthy baby. Post-partum echocardiogram showed preserved ejection fraction with normal strain patterns

Case presentation

A 23-year-old woman presented to the emergency department with sudden onset of non-pleuritic chest pain that radiated to her arms. She was diaphoretic. The patient had no family history of heart disease. She did not smoke or use alcohol or illicit drugs. She had been shopping in a local store when the pain suddenly started. The patient was haemodynamically stable, and there were no pertinent cardiovascular findings on physical examination.

In the emergency department, her electrocardiogram showed sinus bradycardia (Supplementary material online, *Figure S1*) with no significant changes concerning for ischaemia. Initial serum troponin I was 1.2 ng/mL (reference range 0–0.04 ng/mL). The patient was admitted, and serial troponins were performed. Her troponin peaked at 16 ng/mL. She was started on a heparin drip. A transthoracic echocardiogram (TTE) showed apical dyskinesia (*Video 1*) with hyperdynamic motion of the spared myocardium resulting in a preserved ejection fraction of 65% and reduced apical strain (*Figure 1*).



Video 1 Echocardiogram on presentation showing apical dyskinesia.

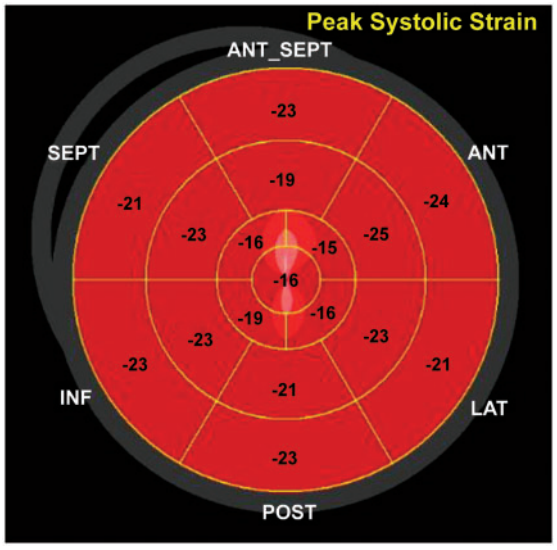


Figure 1 Longitudinal strain bull's-eye plot depicting reduced apical strain. The bull's-eye map provides a view of the left ventricular strain pattern. Apex strain is reduced owing to apical dyskinesis secondary to the thrombosed left anterior descending artery.

Coronary angiography revealed triple vessel coronary ectasia with significant thrombus burden in the occluded left anterior descending artery (LAD) (*Figure 2* and *Video 2*).

Aspiration thrombectomy or intracoronary thrombolysis was contemplated, but, because she was haemodynamically stable and there were no signs of worsening ischaemia, conservative management was recommended. The patient was bridged to warfarin, in addition to dual antiplatelet therapy with aspirin and ticagrelor. Low-dose metoprolol was started. The patient had low blood pressures throughout the admission so we deferred initiation of an angiotensin-converting enzyme inhibitor; she was intolerant to statin therapy due to myalgias. She was discharged on the above regimen. Prior to discharge, the patient was able to return to her baseline activities of daily living with no chest pain or dyspnoea on exertion. It was decided to monitor

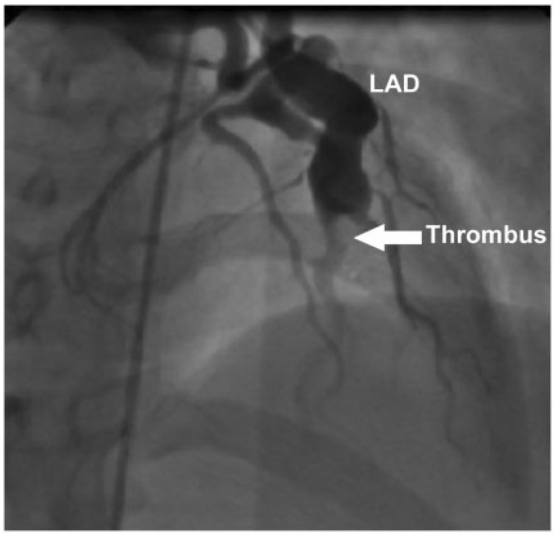
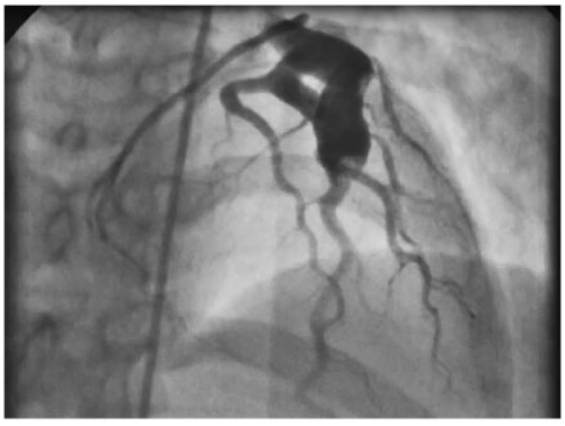


Figure 2 Coronary angiogram of coronary aneurysmal ectasia. Coronary angiography shows ectatic changes to the left anterior descending artery. Thrombus formation is in the mid-left anterior descending artery. LAD, left anterior descending artery.



Video 2 Coronary angiogram reveals diffuse coronary artery ectasia with a thrombus in the aneurysmal left anterior descending artery.

her closely and follow-up in an outpatient setting to pursue further imaging. Given her age, Kawasaki disease was thought to be the likely aetiology of her CAE.

One month after discharge, the patient underwent computed tomography (CT) angiography of the coronary arteries, which confirmed prior findings of multiple coronary artery ectatic changes. The largest diameters noted were 10 mm in the mid-LAD (*Figure 3A*), 9 mm in the proximal LAD, 6.5 mm in the left marginal branch (*Figure 3B*), and 7.5 mm in the right coronary artery (*Figure 3C*). Previously identified coronary thrombi resolved as well. Three-dimensional reconstruction of the coronary vessels was done (*Figure 4*). Repeat TTE continued to show a preserved ejection fraction of 65%, as well as resolved apical dyskinesia and improved apical strain (*Figure 5* and *Video 3*).

Soon after, she reported that, contrary to medical advice, she was pregnant. Warfarin was stopped owing to the teratogenicity of the drug, and she was continued on aspirin, metoprolol, and ticagrelor. As there are no standard guidelines, we decided to de-escalate to dual antiplatelet therapy rather than reintroduce triple therapy with a new agent. Throughout her 37-week pregnancy, the patient did not have any reoccurrence of chest pain, dyspnoea, or palpitations. At 21 weeks of gestation, the patient had a TTE that continued to show a stable ejection fraction and strain pattern. The patient stayed on dual antiplatelet therapy until 5 days before delivery. She delivered a healthy child at 37 weeks via vaginal delivery under epidural anaesthesia with no complications. Two days post-delivery the patient had an echocardiogram that continued to show resolved strain pattern (Supplementary material online, *Figure S2*) and left ventricular function. Further coronary imaging will be pursued in the future.

Discussion

Coronary artery ectasia affects men (2.2%) more than women (0.5%).[2] Atherosclerosis is the most common factor responsible for CAE in older adults, whereas Kawasaki disease is the likely underlying aetiology in younger populations. Coronary artery ectasia is thought to be the result of degradation of extracellular matrix in the media resulting in luminal size increase.[2] The gold standard for diagnosis is coronary angiography; most cases are found incidentally. Other diagnostic modalities include magnetic resonance angiography or coronary CT angiography.[3,4] There are four classifications of CAE: Type 1 is diffuse ectasia with aneurysmal changes in two or more vessels, Type 2 is diffuse ectasia in one vessel and discrete ectasia in a second, Type 3 is diffuse ectasia in one vessel, and Type 4 is discrete ectasia in one vessel. Our patient is a classic example of Type 1 CAE.

Patients with CAE can develop thrombi owing to sluggish blood flow through the enlarged artery.[4] Currently, there are no clear guidelines for treatment of this disease process, and treatment should be individualized for each case. Treatment modalities may include percutaneous intervention, surgery, or medication. One recent case report successfully treated a non-ST-elevation myocardial infarction with thrombolysis.[5] In our case, we initially had plans to pursue either aspiration thrombectomy or coronary thrombolysis, but owing to the facts that the patient was symptomatically improving, had a preserved ejection fraction, and showed no further signs of ischaemia, we decided to continue with conservative management and bridge the patient to warfarin. Bail-out administration of glycoprotein IIb/IIIa inhibitors was another viable option.

Percutaneous intervention in patients with CAE or coronary aneurysm is challenging owing to difficulty with coaptation of the stent to the vessel wall. Outcomes data have been limited to small case series, which showed lower procedural success and higher incidences of no re-flow, repeat revascularization, and long-term mortality.[6] One case of a 24-year-old man who presented with ST-elevation myocardial infarction secondary to CAE was reported; in this case, percutaneous coronary intervention was not able to establish Thrombolysis in Myocardial Infarction 3 flow and the patient ultimately had to be treated with medical therapy.[7] There are two techniques that have

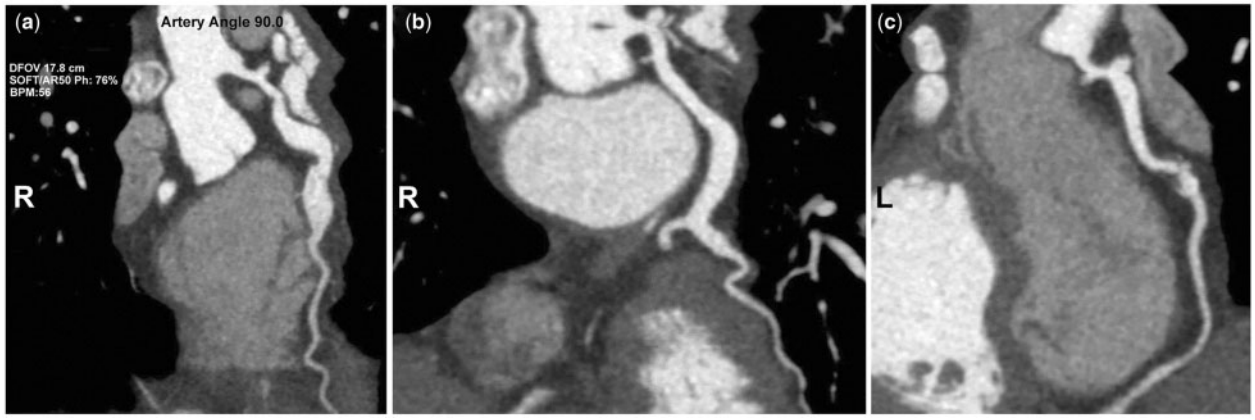


Figure 3 Multiple computed tomography views of coronary aneurysmal ectasia. (*A*) Computed tomography of the left anterior descending artery shows resolution of the previously identified thrombus. (*B*) Computed tomography of the left marginal branch. (*C*) Computed tomography of the right coronary artery.

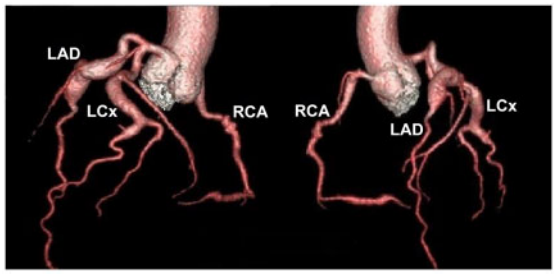


Figure 4 Three-dimensional imaging of coronary aneurysmal ectasia. Three-dimensional reconstruction of the coronary arteries shows ectatic changes to the right coronary, left anterior descending, and left circumflex arteries. LAD, left anterior descending artery; LCx, left circumflex artery; RCA, right coronary artery.

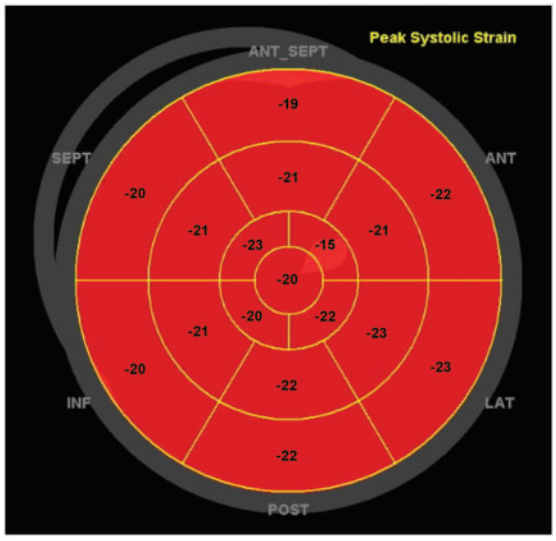


Figure 5 Longitudinal strain bull's-eye plot at 1-month follow-up depicts improved apical strain.

overcome these challenges. Covered stent or double open-cell stent can be used for revascularization if the proximal right coronary artery has aneurysmal disease.[8] If a large side branch originates near the aneurysm, the patient may benefit from stent-assisted coiling.[8]

When an ectatic lesion enlarges and becomes aneurysmal, surgical resection of the aneurysm is an appropriate alternative.[9] Patients who should be considered for surgery include those who have obstructive lesions or significant ischaemia despite the use of medications.[10] If surgery is pursued, there are multiple surgical techniques to correct aneurysms, including, but not limited to, ligation with bypass grafting, marsupialization with interposition graft, and resection.[8]

Our case presented a second degree of complexity once the patient informed us she was pregnant. We had difficult decisions to make about how to manage anticoagulation with no clear guidelines. We decided to discontinue warfarin because of its teratogenicity. With pregnancy, blood volume, heart rate, and cardiac output all increase and blood pressure fluctuates. All of these physiological changes increase the risk of coronary rupture and/or spontaneous dissection. As there are no guidelines, we made the decision to de-escalate to dual antiplatelet therapy rather than continue with triple therapy. In one case study, a patient was treated with propranolol, aspirin, and enoxaparin throughout her 37-week pregnancy.[11] Her pregnancy was complicated with worsening New York Heart Association class as well as recurrent episodes of chest pain and palpitations; ultimately, she delivered a healthy baby at 37 weeks' gestation.[11] In our case, the patient did not have any adverse symptoms throughout her pregnancy while on aspirin, ticagrelor, and metoprolol. In a study done on 13 women with Kawasaki disease and coronary artery lesions, 10 patients delivered via vaginal delivery with epidural anaesthesia and 3 via caesarean section with no complications, endorsing the decision to deliver via vaginal delivery with epidural anaesthesia in our case.[12]

Video 3 Echocardiogram at 1-month follow-up shows resolved apical dyskinesia.

Currently, the recommendation is to treat with medical management until failure.[4] The mainstay of medical management is antiplatelet therapy as well as avoiding medications with vasodilating properties.[4]

Conclusion

Coronary artery ectasia is a rare disease that currently has no set guidelines for treatment. Here, we present a case of a young female with significant ectatic burden who was successfully treated with dual antiplatelet therapy and anticoagulation. She subsequently had a safe and healthy pregnancy with delivery of a healthy baby while on beta blocker and dual antiplatelet therapy. We elected for medical therapy owing to the lack of significant left ventricular dysfunction and the quick resolution of physical symptoms.

Lead author biography

Dr Akshar Jaglan is an Internal Medicine resident who has an interest in cardiology and complex cases. He received his bachelor's degree from Saint Louis University and his doctorate in osteopathic medicine from Campbell University School of Osteopathic Medicine. He is a third year Internal Medicine resident at Aurora Sinai/Aurora St. Luke's Medical Centers, University of Wisconsin School of Medicine and Public Health, Milwaukee, WI, USA, and aims to be a fellow in cardiology in the near future.

Supplementary material

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

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

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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