ESC European Society of Cardiology

European Heart Journal - Case Reports (2020) **4**, 1–5
doi:10.1093/ehjcr/ytaa253

CASE REPORT
Coronary heart disease

Coronary artery ectasia presenting with ST-elevation myocardial infarction in a young indigenous man: a case report

John Lee , Satish Ramkumar *, Nancy Khav , and Benjamin K. Dundon

Monash Cardiovascular Research Centre, MonashHeart, Monash Health, 246 Clayton Road, Clayton, Melbourne, VIC 3169, Australia

Received 5 January 2020; first decision 4 March 2020; accepted 13 July 2020; online publish-ahead-of-print 25 August 2020

Background Coronary artery ectasia (CAE) is often an incidental finding on angiography, however, patients can present with acute coronary syndrome due to a large thrombus burden requiring treatment with percutaneous coronary intervention or with emergency surgery.

Case summary A 26-year-old Indigenous Australian male was admitted with anterior ST-elevation myocardial infarction associated with an out of hospital ventricular fibrillation arrest. Coronary angiography demonstrated thrombotic occlusion of the proximal left anterior descending (LAD) artery with heavy thrombus burden and prominent vascular ectasia of all three coronary arteries. He was managed with surgical thrombectomy and coronary artery bypass graft of his LAD.

Discussion This is the first case of triple CAE in an Indigenous Australian. The case highlights the lack of consensus approach in the management of CAE due to paucity of prospective studies.

Keywords Case report • Coronary artery ectasia • Acute coronary syndrome • • Anticoagulation • Percutaneous coronary intervention

Learning points

- Coronary artery ectasia (CAE) is a rare finding in angiography and occur due to disease processes that affect vessel wall integrity.
- Understand the pathophysiology and presentations of CAE.
- Understand the paucity of prospective studies in management of CAE.

Introduction

Coronary artery ectasia (CAE) is an aneurysmic abnormality of the artery characterized by diffuse dilatation, with a luminal diameter 1.5× wider than that of adjacent normal segments. It differs to coronary artery aneurysms which is a focal dilatation of an artery. Coronary artery ectasia is found in 2.7–2.8% of angiograms and occurs from disease processes that impair vessel wall integrity.[1,2] It is commonly associated with atherosclerotic disease (50%) but it can be a congenital malformation (20–30%) or occur secondary to connective tissue disorders, inflammatory diseases, and infections (10–20%).[3] Coronary artery ectasia most commonly presents with stable angina (61–66%) and can be complicated by acute coronary syndrome (ACS).[4,5] Treatment of ACS in isolated CAE can be challenging as no consensus

* Corresponding author. Tel: +61 433126231, Email: satish.ramkumar@monashhealth.org
Handling Editor: Dejan Milasinovic
Peer-Reviewers: Gavin Paul Raphael Manmathan, Domenico D'Armario, and Milenko Zoran Cankovic
Compliance Editor: Rahul Mukherjee

approach exists and evidence is scarce.[3,6] The principles, however, are of relieving the obstructive lesion, antiplatelet therapy, optimization of cardiovascular risk factors, and potential consideration of long-term anticoagulation to prevent recurrent thrombus formation.[3,6,7]

Timeline

Day	Events
Day 1	Index episode—16:15
	Patient suffers out of hospital ventricular fibrillation (VF) arrest
	Initial bystander cardio-pulmonary resuscitation commences with automated electronic defibrillator advising a shock
	Paramedic arrival—17:15
	Ambulance officers diagnose anterolateral ST-elevation myocardial infarction on electrocardiogram. Administer heparin intravenous (IV) 4000 units and tenecteplase IV 50 mg
	30 min from VF arrest to thrombolysis
	Patient is flown out from rural setting to tertiary centre. Flight begins at 17:49
	Arrival to the Emergency Department—19:13
	Patient arrives at the emergency department at 19:13
	Coronary angiography—20:30
	Patient seen by cardiology team and taken to the catheterization lab where triple vessel coronary artery ectasia is demonstrated
	Due to unsuitability for percutaneous coronary intervention and thrombus aspiration, patient referred for urgent coronary artery bypass graft (CABG)
	CABG—23:00
	Patient taken to theatre by cardiothoracic team and consented for urgent coronary artery bypass grafting. Intra-aortic balloon pump (IABP) inserted via right femoral artery prior to theatre with no complications
	Total procedure time of 3 h
Day 2	Patient is admitted to intensive care unit (ICU) post-operatively
	Noradrenaline is weaned
Day 3	IABP is removed in ICU
	Uncomplicated post-operative course
	Vasculitic, connective tissue and thrombophilia screen is sent
Day 4	Patient discharged to ward bed
Day 5	Vasculitic, connective tissue and thrombophilia screens return and are unremarkable
	Rheumatology consultation obtained: no stigmata of Marfan's syndrome or Ehlers–Danlos syndrome, no childhood rashes, previous rheumatic fever, Kawasaki disease, no localizing infective symptoms
Day 10	Patient had an uncomplicated post-operative recovery and was discharged home with ongoing cardiology and cardiothoracic surgery follow-up

Case presentation

A 26-year-old male Indigenous Australian was transferred to our tertiary hospital with an anterior ST-elevation myocardial infarction associated with an out of hospital ventricular fibrillation (VF) arrest. He developed central chest pain whilst playing a competitive football match prior to the cardiac arrest. Initial bystander cardio-pulmonary resuscitation was commenced and he received one 150 joule direct current shock from an automated electrical defibrillator for VF with return of spontaneous circulation. His initial vital signs in the ambulance included a heart rate of 78 b.p.m. (regular) and blood pressure of 125/88 mmHg with no clinical signs of cardiac failure. The patient's cardiovascular risk factors included an active smoking history and a family history of ischaemic heart disease; the patient's uncle underwent coronary artery bypass graft (CABG) surgery. There was no family history of sudden cardiac death.

The initial electrocardiogram (ECG) performed by the paramedics demonstrated sinus rhythm with anterolateral ST-elevation (*Figure 1*). Upon diagnosis of ST-elevation ACS, the patient was thrombolysed with 50 mg of tenecteplase as the patient was more than 2 h away from a primary percutaneous coronary intervention (PCI) capable site. In addition to thrombolysis, 4000 international units of intravenous heparin and oral aspirin 300 mg were also administered. Post-thrombolysis, due to <50% ST-segment resolution and ongoing chest pain, the patient proceeded to our tertiary centre for rescue PCI.

Bloods tests on admission revealed a haemoglobin of 150 g/L, platelets of 26 600/μL, and a troponin I of 5.08 μg/L. A bedside echocardiogram demonstrated severely reduced left ventricular ejection fraction of 15% with severe hypokinesis extending from the mid-anterior wall to the apical and inferoapical segments (see Supplementary material online, *Video S1*).

Coronary angiography demonstrated thrombotic occlusion of the proximal left anterior descending (LAD) artery with heavy thrombus burden and prominent vascular ectasia of all three coronary arteries (see *Figures 2 and 3* and Supplementary material online, *Videos S2 and S3*). The left circumflex and right coronary artery demonstrated thrombolysis in myocardial infarction 3 flow with no stenosis despite significant ectasia. Thrombus aspiration was not performed due to the perceived high risk of systemic cardioembolic events. Percutaneous coronary intervention was not attempted due to significant vascular ectasia preventing stent apposition within the grossly dilated vessel wall. Due to continuing chest pain, ST-segment elevation and persistent thrombotic vessel occlusion, the patient was referred for urgent CABG surgery. Long-term LAD graft patency (following possible future thrombus resolution) was discussed, should non-obstructive coronary artery disease (CAD) be identified at surgery. On balance, given the patient's young age and desire to minimize further myocardial injury, surgery was agreed as the most appropriate immediate therapeutic option.

Prior to surgery, an intra-aortic balloon pump (IABP) was inserted, with a noradrenaline infusion running at 2 μg/min. Then, the left inferior mammary artery was harvested and anastomosed to the distal LAD successfully. Post-pump trans-oesophageal echocardiogram revealed reduced left ventricular ejection fraction of 40–45% with significant improvement in the anterior wall contractility. The patient remained in the intensive care unit (ICU) for 48 h post-surgery where

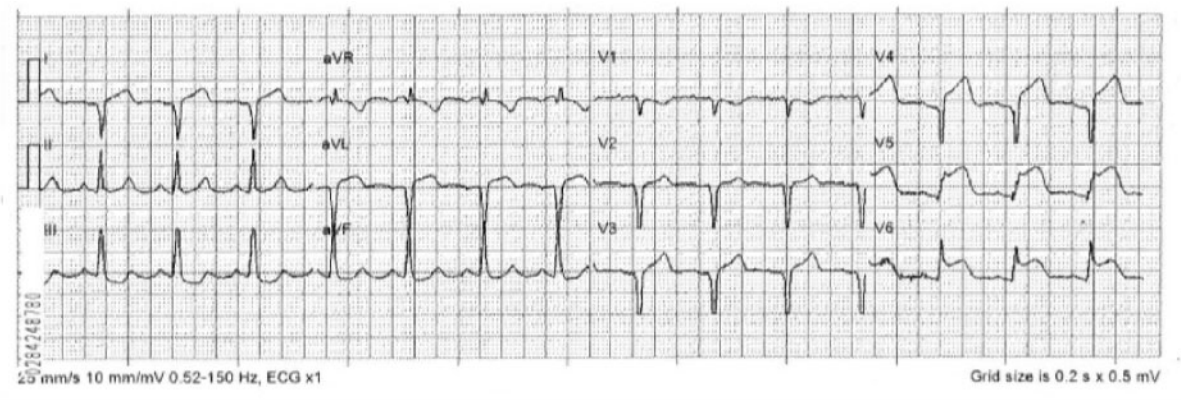


Figure 1 Initial electrocardiogram demonstrating anterolateral ST-elevation myocardial infarction with reciprocal changes in the inferior leads.

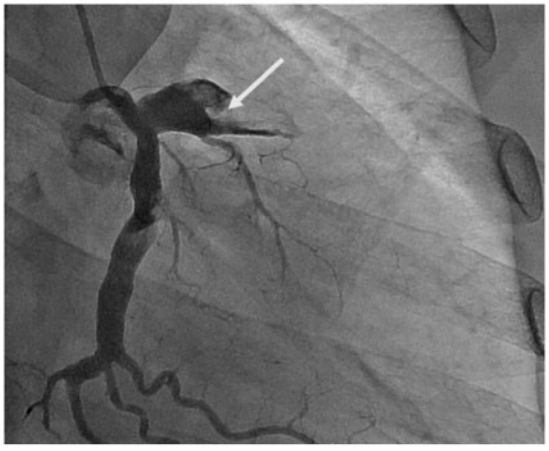


Figure 2 Coronary angiogram demonstrating ectatic left main coronary artery and an arrow demonstrating proximal left anterior descending artery with thrombotic occlusion.

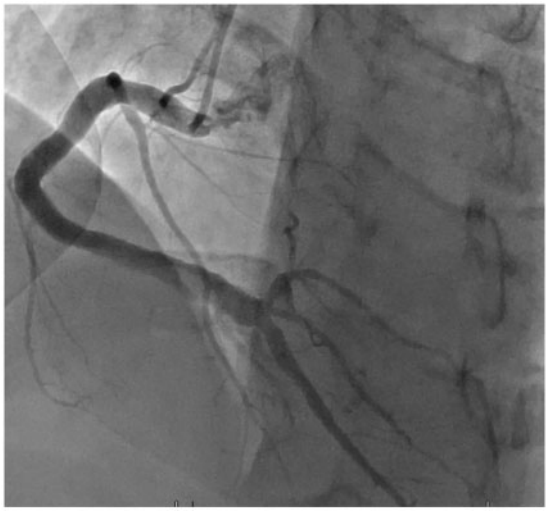


Figure 3 Coronary angiogram demonstrating ectatic right coronary artery.

a therapeutic heparin infusion was administered for the 48 h. Prior to discharge to the ward, the noradrenaline infusion was ceased and the IABP removed. Post-operatively in ICU, the rheumatology team consulted and alternative causes for the CAE such as vasculitic, connective tissue disease, and autoimmune conditions were ruled out.

The patient experienced an uncomplicated post-operative course and was discharged home after 10 days in hospital. The final ECG demonstrated normal sinus rhythm with no abnormalities (*Figure 4*). On discharge the patient was prescribed dual antiplatelet therapy (DAPT) of aspirin 100 mg daily and ticagrelor 90 mg twice daily to continue for a duration of 1 year given the presentation of ACS. For management of the heart failure with reduced ejection fraction, perindopril 2.5 mg daily, bisoprolol 1.25 mg daily, and spironolactone 12.5 mg daily were prescribed with view for uptitration to maximal tolerated dose. Atorvastatin 80 mg was prescribed for aggressive secondary prevention of ACS. The patient was referred for ongoing outpatient cardiology and cardiothoracic surgery clinic review.

Discussion

The gold standard of diagnosis of CAE remains coronary angiography coupled with intracoronary imaging modalities such as intravascular ultrasound or optical coherence tomography. Both provide excellent morphological information of the ectatic artery and can help distinguish the pathology behind the CAE, especially if it is associated with previous stent implantation.[3,8,9] Non-invasive imaging techniques can also diagnose CAE. Computed tomography (CT) coronary angiography via multi-detector CT is now more readily available and

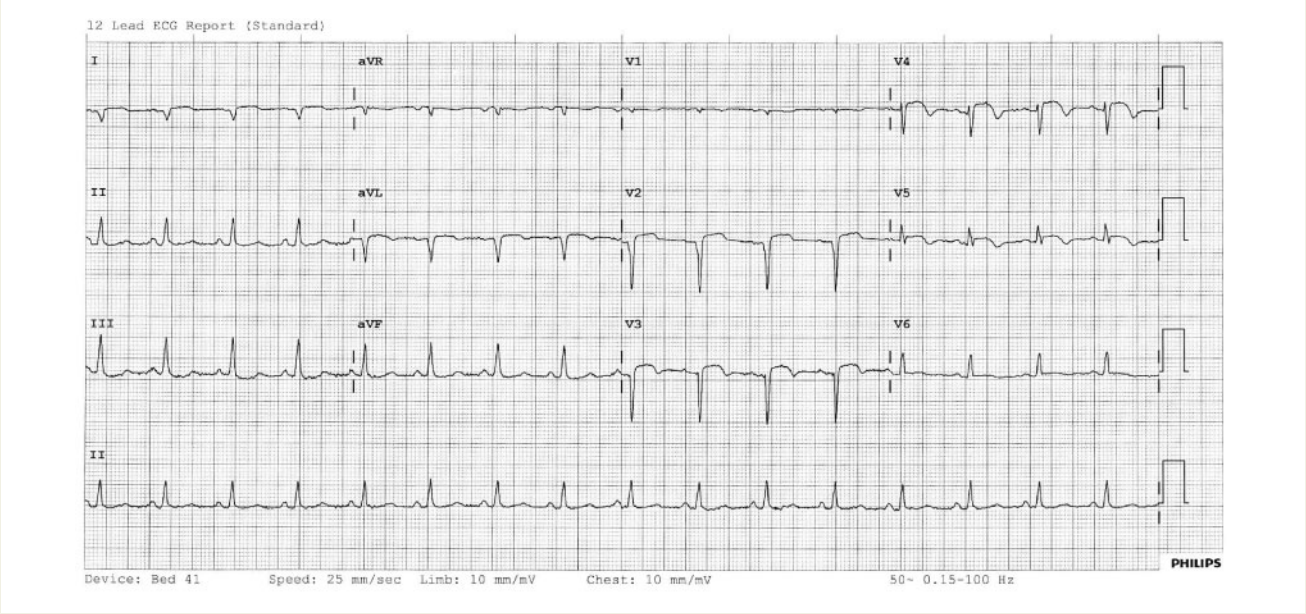


Figure 4 Patient's final electrocardiogram prior to discharge home demonstrating anterior Q waves consistent with recent LAD territory infarct.

provides adequate representation of lumen size, morphology and may also demonstrate associated stenotic segments and plaques.[3,8,10]

There is no consensus approach in the management of isolated CAE, as in this case. The evidence is derived largely from case reports and studies limited by population size and definitive management and long-term prognosis remain unclear.[3,6,11] In the absence of compelling evidence favouring therapeutic strategies specific to isolated CAE, the management needs to be tailored to each patient with consideration of the comorbidities and angiographic findings.[11]

During the presenting acute myocardial infarction, treatment options in the catheterization lab include manual aspiration thrombectomy (MAT), use of a stent retriever, intracoronary glycoprotein IIb/IIIa inhibitors, intracoronary thrombolysis, plain old balloon angioplasty, or mesh-covered stent implantation if the ectatic segment is short enough.[11] Percutaneous coronary intervention to these ectatic lesions, however, have unique technical challenges. Coronary artery ectasia lesions have high thrombus burdens that may be refractory to MAT or intracoronary delivery of medications.[11] There is also a higher risk of embolic stroke with MAT. Furthermore, in the case of stenting, sizing and landing zone assessment is difficult and multiple overlapping stents may be needed to adequately traverse ectatic segments.[11] These challenges are reflected by the lower rates of procedural success, higher rates of no-reflow, and distal embolization in PCI for acute myocardial infarction in CAE.[12,13] Where PCI fail, emergency surgery is an option. Operative interventions include ligation or resection of the ectatic artery, aneurysmal thrombectomy, or bypass grafting across the ectatic segment.[11] Surgical approaches are individualized to the coronary lesion as the ideal approach has not been studied.

In terms of medical therapy, administration of aspirin has been recommended due to the observation of concomitant obstructive coronary lesions in the majority of patients with CAE and myocardial infarction.[7] The role and specific benefits of DAPT in long-term secondary prevention are unknown.[11] Initial studies have recommended the use of long-term oral anticoagulation on the premise of an increased risk of thrombosis in aneurysmic or ectatic segments. To date, however, there has only been a single retrospective study addressing this. In this study, patients who were treated with vitamin K antagonists who had a percentage of time in therapeutic range (%TTR) ≥60% had a lower occurrence of major adverse cardiac events than those with %TTR <60% or without anticoagulation therapy.[14] The population in that particular trial, however, were elderly patients with multiple cardiovascular risk factors, unlike our presented case. The potential role of novel oral anticoagulants in the primary or secondary prevention of ACS due to CAE thrombosis in affected patients is also yet to be determined.

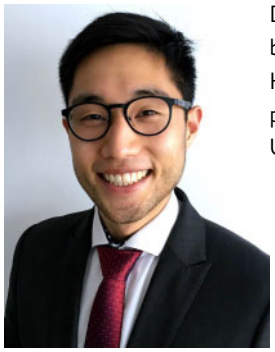
It is noteworthy that nitrates, a mainstay of therapy in conventional CAD, may exacerbate myocardial ischaemia in CAE due to the coronary steal phenomenon.[6] The use of angiotensin-converting enzyme (ACE) inhibitors have been speculated as ACE gene polymorphisms have been noted to be associated with the presence of CAEs, but their utility is not yet proven. Statins are also thought to potentially be of benefit by suppressing matrix metalloproteinases that cause dilatation of the arterial wall, but this has not been demonstrated in prospective trials.[7] Due to the rare nature of the condition, and the absence of randomized controlled clinical trials, it has been recommended that affected patients should have regular outpatient follow-up to assess for new symptoms and side effects of medications. In the absence of alternative evidence, CAE patients should be treated to standard secondary prevention targets if underlying CAD is present.

Conclusion

We present the first documented case of triple vessel CAE in a young Indigenous Australian, who was, in the absence of randomized

controlled trials regarding management of CAE-associated ACS, ultimately treated with emergency CABG surgery after failed thrombolysis.

Lead author biography



Dr John Lee is a resident completed his basic physician training in Monash Health in Melbourne, Victoria. He completed his Doctor of Medicine at the University of Melbourne.

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.

References

1. Giannoglou GD, Antoniadis AP, Chatzizisis YS, Damvopoulou E, Parcharidis GE, Louridas GE. Prevalence of ectasia in human coronary arteries in patients in northern Greece referred for coronary angiography. *Am J Cardiol* 2006;**98**:314–318.
2. Sultana R, Sultana N, Ishaq M, Samad A. The prevalence and clinical profile of angiographic coronary ectasia. *J Pak Med Assoc* 2011;**61**:372–375.
3. Mavrogeni S. Coronary artery ectasia: diagnosis and treatment. *Hellenic J Cardiol* 2010;**51**:158–163.
4. Malviya A, Jha PK, Mishra A. Isolated coronary artery ectasia: clinical, angiographic, and follow up characteristics. *Indian Heart J* 2017;**69**:619–623.
5. Zografos TA, Korovesis S, Giazitzoglou E, Kokladi M, Venetsanakos I, Paxinos G *et al.* Clinical and angiographic characteristics of patients with coronary artery ectasia. *Int J Cardiol* 2013;**167**:1536–1541.
6. Boles U et al. Coronary artery ectasia; clinical updates and management options in acute presentation. *Med Res Arch* 2017;**5**. ISSN 2375-1924.
7. Devabhaktuni S, Mercedes A, Diep J, Ahsan C. Coronary artery ectasia—a review of current literature. *Curr Cardiol Rev* 2016;**12**:318–323.
8. Abou Sherif S, Ozden Tok O, Taşköylü Ö, Goktekin O, Kilic ID. Coronary artery aneurysms—a review of the epidemiology, pathophysiology, diagnosis, and treatment. *Front Cardiovasc Med* 2017;**4**:24.
9. Fam JM, van der Sijde JN, Karanasos A, Zhang B, van Geuns R-J, Regar E. Use of intracoronary imaging in ST elevation myocardial infarction with coronary artery aneurysm and very late stent thrombosis. *Int J Cardiol* 2015;**197**:296–299.
10. Farrag A, Faramawy AE, Salem MA, Wahab RA, Ghareeb S. Coronary artery ectasia diagnosed using multidetector computed tomography: morphology and relation to coronary artery calcification. *Int J Cardiovasc Imaging* 2013;**29**:427–433.
11. Kawsara A, Núñez Gil IJ, Alqahtani F, Moreland J, Rihal CS, Alkhouli M. Management of coronary artery aneurysms. *JACC Cardiovasc Interv* 2018;**11**:1211–1223.
12. Bogana Shanmugam V, Psaltis PJ, Wong DTL, Meredith IT, Malaiapan Y, Ahmar W. Outcomes after primary percutaneous coronary intervention for ST-elevation myocardial infarction caused by ectatic infarct related arteries. *Heart Lung Circ* 2017;**26**:1059–1068.
13. Ipek G, Gungor B, Karatas MB, Onuk T, Keskin M, Tanik O *et al.* Risk factors and outcomes in patients with ectatic infarct-related artery who underwent primary percutaneous coronary intervention after ST elevated myocardial infarction. *Catheter Cardiovasc Interv* 2016;**88**:748–753.
14. Doi T, Kataoka Y, Noguchi T, Shibata T, Nakashima T, Kawakami S *et al.* Coronary artery ectasia predicts future cardiac events in patients with acute myocardial infarction. *Arterioscler Thromb Vasc Biol* 2017;**37**:2350–2355.