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CASE REPORT
Coronary heart disease

Giant coronary artery aneurysm associated with familial retinal artery macroaneurysm: a case report

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Background The term 'Giant' coronary aneurysm is usually defined as any coronary aneurysm more than 8 mm in maximum diameter. The form of familial retinal arterial macroaneurysms (FRAMs) is a rare autosomal recessive disease that is described by arterial aneurysm formation in the retina. Here, we report an association of coronary artery aneurysms with FRAM in a young male who presented with the acute coronary syndrome.

Case summary A 31-year-old male smoker presented to the emergency department with atypical chest pain for 5 days. Blood investigations showed raised troponin enzymes. Review of his past medical history revealed decreased vision in the left eye, starting at the age of 10 years which progressed to blindness. He was diagnosed to have IGFBP7 mutation which causes eye manifestations in the form of FRAM. Fundoscopy showed bleeding retinal artery macroaneurysms in the right retina and sub-retinal gliosis suggesting laser treatment for the prior retinal arterial aneurysm. Coronary angiogram revealed a large aneurysm in the proximal segment of the left anterior descending (LAD) artery. Cardiac computed tomography scan with contrast was done which showed a 2.28 × 1.64 cm coronary aneurysm at the proximal segment of the LAD artery with peripheral calcification with a narrow neck of about 0.6 cm. After the heart team discussion, he underwent surgical ligation of the LAD and coronary aneurysm with implantation of the saphenous venous graft to distal LAD.

Discussion Coronary artery aneurysms can be a part of multisystem diseases like FRAM. The management should be individualized based on symptoms at presentation, size of coronary aneurysms, and local expertise.

Keywords Case report • Coronary artery aneurysm • Acute coronary syndrome • Familial retinal arterial macroaneurysms (FRAM)

ESC Curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 2.4 Cardiac computed tomography

Learning points

- Coronary artery aneurysms can present as acute coronary syndrome.
- Coronary artery aneurysms can have an association with familial retinal arterial macroaneurysm.
- The treatment of the coronary artery should be individualized based on the size of the aneurysm and mode of presentation.

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Introduction

Coronary artery aneurysm (CAA) is defined as ≥1.5 times dilatation of the luminal diameter of the coronary artery as compared to adjacent segments.[1] Patients with CAA are mostly asymptomatic but it can be associated with adverse outcomes due to thrombosis or rupture. Atherosclerosis is the most common cause but coronary aneurysms can be a cardiac manifestation of the multisystemic disease. Familial retinal artery macroaneurysm (FRAM) is a rare autosomal recessive disorder characterized by retinal arterial aneurysms, which can lead to significant visual impairment due to haemorrhage and fibrosis.[2] Here, we report an association of CAAs with FRAM in a young male who presented with an acute coronary syndrome.

Timeline

Days 1 and 2	• Admission with chest pain • Positive cardiac enzymes • Screening of coronavirus disease-19
Day 3	• Echocardiography • Coronary angiogram
Day 6	• Computed tomography coronary angiogram
Day 7	• Ophthalmology review • Computed tomography brain
Day 12	• Surgery

Case presentation

A 31-year-old male smoker presented to the emergency department with episodes of atypical chest pain radiating to the left arm for 5 days. There was no history of fever, cough, or contact with the coronavirus disease (COVID)-infected patient. On clinical examination, he was vitally stable with a pulse rate of 74 beats/min and blood pressure of 125/74 mm/Hg. He was afebrile and oxygen saturation at room air was 95%. Cardiac examination revealed the normal character of first and second heart sound without additional sounds. A review of his past medical history revealed decreased vision in the left eye, starting at the age of 10 years which progressed to blindness. He was diagnosed to have IGFBP7 mutation which causes eye manifestations in the form of FRAM. His younger brother was also diagnosed to have FRAM and IGFBP7 mutation at the age of 23. The rest of his siblings and parent are asymptomatic.

His electrocardiogram showed normal sinus rhythm with T-wave inversions in lateral leads (*Figure 1*). Blood investigations showed a mildly raised white blood cell count of 13.08 $\times$ 10^9/L (normal 3.9–11 $\times$ 10^9/L). The troponin was raised at 637 ng/L (normal = 34 ng/L) and the repeated level showed 1938 ng/L and it peaked up to 5484 ng/L the next day. Echocardiography showed normal biventricular systolic function with normal valves. There were no regional wall motion abnormalities. Chest X-rays did not show any significant findings. Considering the COVID pandemic, inflammatory markers, and polymerase chain reaction (PCR) for COVID were requested. His c-reactive proteins were high but PCR for COVID was negative. Considering his past ophthalmological history fundoscopy was performed which showed bleeding retinal artery macroaneurysms in the right retina and sub-retinal gliosis suggesting laser treatment for prior retinal arterial aneurysm (*Figure 2*).

Coronary angiogram was performed which showed a large aneurysm in the proximal segment of the left anterior descending (LAD) artery (*Figure 3*, *Videos 1* and *2*). There was no evidence of plaque rupture or thrombus in aneurysm. The rise in cardiac biomarkers can possibly be explained by type 2 myocardial infarction (supply-demand mismatch). Cardiac computed tomography (CT) scan with contrast was done which showed a 2.28 $\times$ 1.64 cm coronary aneurysm at the proximal segment of the LAD artery with peripheral calcification with a narrow neck of about 0.6 cm (*Figures 4* and *5*). Myocardial bridging was also noted in the mid-segment of LAD. There were non-obstructive calcified plaques in the right coronary artery. As part of the work-up, his CT scan of the brain with contrast was done to assess associated intracerebral aneurysms which did not show any significant abnormality.

After the heart team discussion, he was referred for cardiac surgery. Medical therapy was not preferred as he was symptomatic and presented with the acute coronary syndrome, moreover; there was the risk of thrombosis or rupture due to the large (giant) size of the aneurysm. The percutaneous intervention was not chosen due to the size of the aneurysm and the risk of occlusion of a large size diagonal branch originating from the lumen of the aneurysm. He underwent surgical ligation of the LAD and coronary aneurysm with implantation of the saphenous venous graft to distal LAD as the left internal mammary artery was smaller and underdeveloped. On follow-up after 1 year, he is asymptomatic.

Discussion

Although the term 'Giant' coronary aneurysm is still controversial and it is usually defined as any coronary aneurysm more than 8 mm in maximum diameter.[3] Coronary artery aneurysms are a rare entity and reported incidence is between 1.5% and 5%.[4] The incidence of giant coronary aneurysms is unknown as only a few cases are reported in the literature.[5] Most CAA are asymptomatic and diagnosed incidentally on coronary imaging or autopsy. Nevertheless, coronary aneurysms can cause angina pectoris or myocardial infarction due to poor distal flow. It can also lead to coronary thrombosis, dissection, and vessel compression. CAA can be occasionally congenital and mostly acquired. Atherosclerotic coronary artery disease is the most common cause of coronary aneurysm formation. Other aetiologies include trauma, connective tissue diseases (Marfan syndrome and Ehlers Danlos syndrome), infections (bacterial, fungal, syphilis, mycobacterium, and HIV), and inflammatory diseases like Kawasaki disease, Takayasu arteritis, and Behcet's disease.[4]

The CAAs are considered as a part of the multisystemic phenomenon and their association with retinal artery aneurysms is reported in the literature.[6] Retinal arterial macroaneurysms are mostly

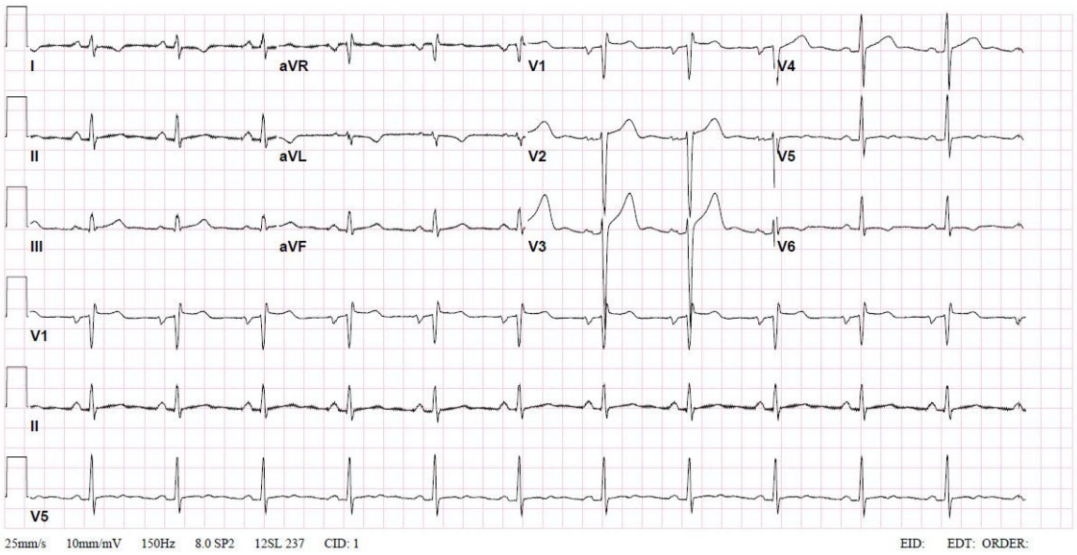


Figure 1 Electrocardiography of patient showing normal sinus rhythm with T-wave inversions in lead 1, aVL, V5–6.

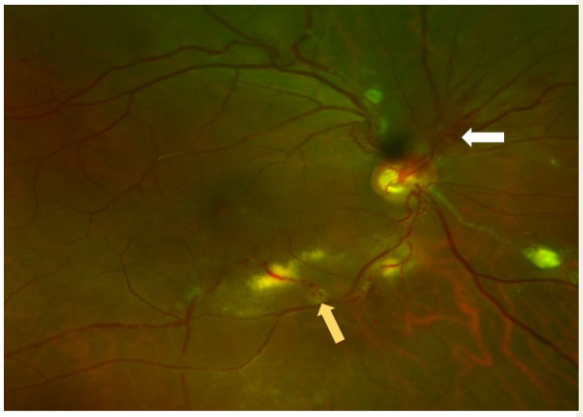


Figure 2 Fundoscopy of the right eye showing bleeding retinal artery macroneurysm (white arrow) and another leaking macroneurysm (yellow arrow).

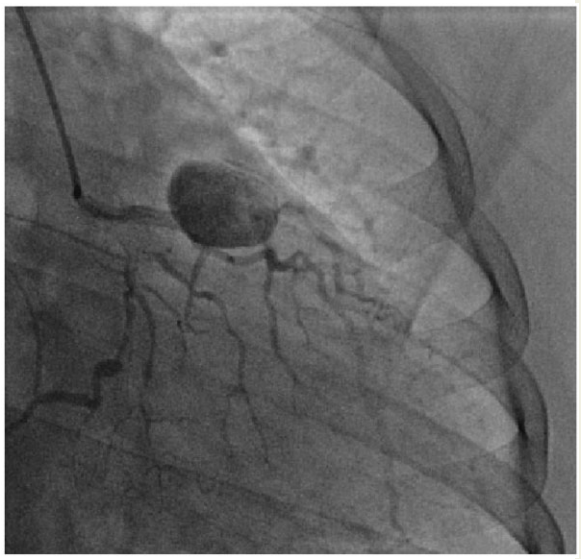
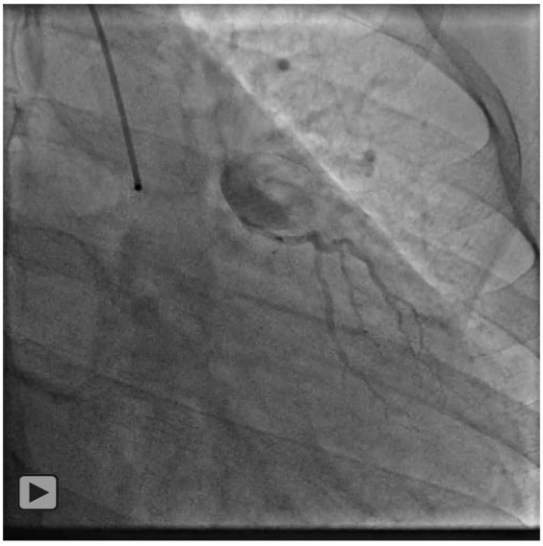


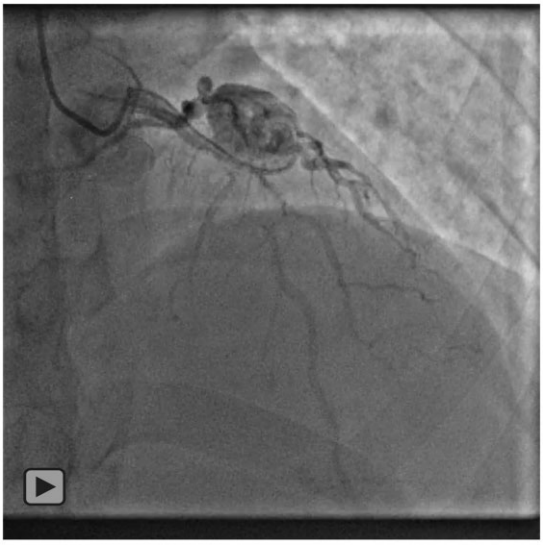
Figure 3 Still image of coronary angiogram showing large aneurysm in the proximal left anterior descending artery.

unilateral, acquired dilatations of retinal arterioles.[7] Clinical presentation varies from initial asymptomatic disease followed by the gradual evolution of visual loss due to thrombosis, fibrosis, and haemorrhage.[8] The form of familial retinal arterial macroaneurysms is a rare autosomal recessive disease that is described by arterial aneurysm formation in the retina. Familial retinal arterial macroaneurysm is a rare progressive disease leading to progressive unilateral or bilateral vision loss. This familial entity is linked to a mutation in IGFBP7, which is translated into defective protein formation in smooth muscles of larger vessels.[9] Although there are case reports of FRAM associated with supravalvular pulmonary stenosis but no such association was found in our patient.[6]

The choice of diagnostic modality for coronary aneurysms depends upon presentation. For screening purposes, non-invasive techniques without radiation or contrast should be used like echocardiography and magnetic resonance angiography. Patients with high clinical suspicion or where further delineation of coronary aneurysms are required, invasive diagnostic tests like cardiac CT or invasive coronary angiography should be utilized. It is important to screen other vasculature of the body as patients with coronary aneurysms can have associated varicose veins, varicocele, or aortic aneurysm.[10,11]



Video 1 Coronary angiogram (ROA cranial view) showing a large aneurysm in the proximal left anterior descending artery.



Video 2 Coronary angiogram (AP cranial) showing large aneurysm in the proximal left anterior descending artery.

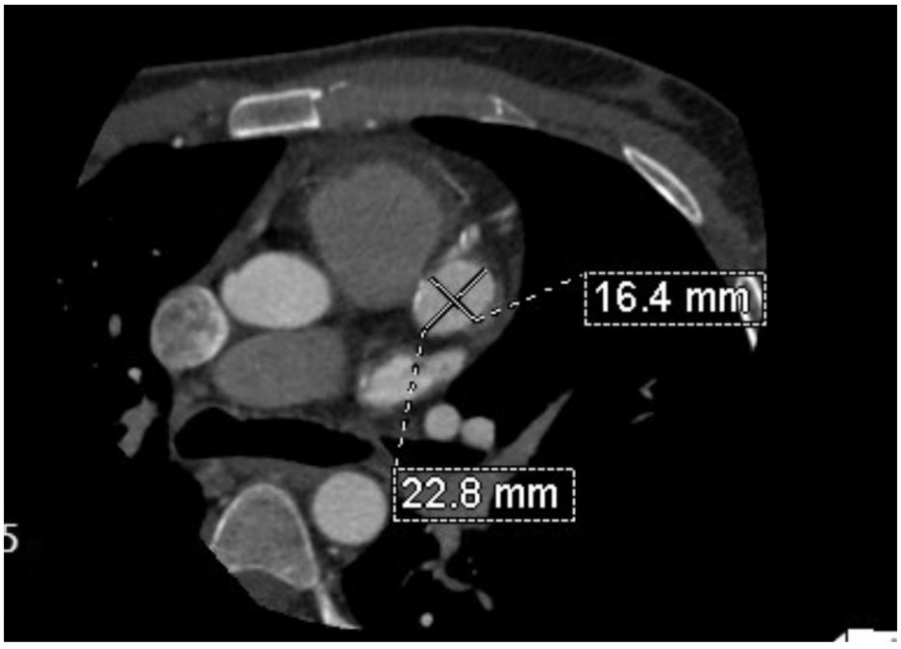


Figure 4 Axial images of computed tomography scan with contrast showing 22.8 mm × 16.4 mm aneurysm with mild calcification in the proximal left anterior descending coronary artery.

Regardless of the aetiology or association of CAAs, there is no agreement for the management. The treatment options include medical therapy with anticoagulation, percutaneous intervention with covered stents, and surgical correction.[12] The surgical approach should be preferred over other options in cases of giant aneurysms, severe coronary stenosis, complications such as fistula formation or compression of adjacent structures, and post-coronary intervention aneurysms.[13,14] Resection or ligation along with bypass grafting is the

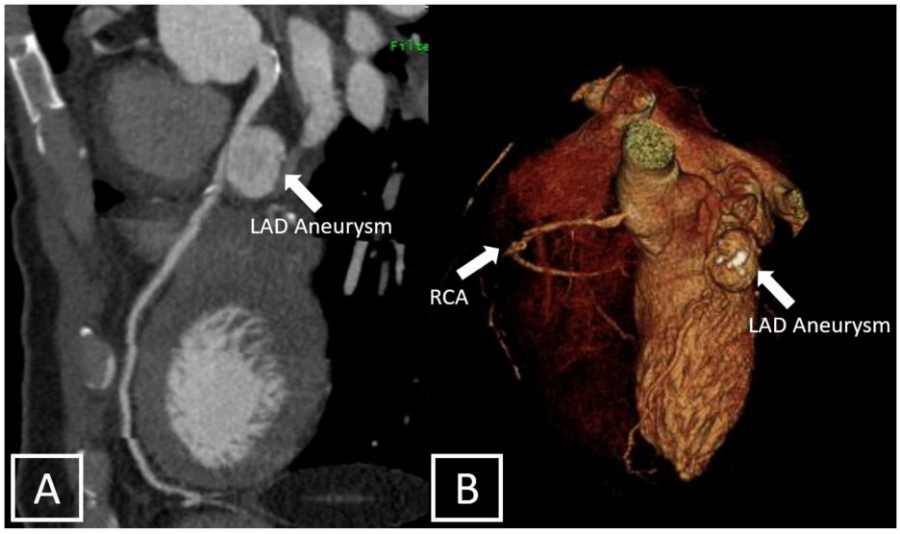


Figure 5 Multiplanar reconstruction image of cardiac computed tomography scan with contrast showing giant coronary aneurysm in the proximal left anterior descending artery in panel *A*. Panel *B* shows three-dimensional volume rendered image of the same aneurysm along with right coronary artery.

preferred, albeit technically challenging treatment option for giant, thrombosed coronary aneurysms.[15] The prognosis of coronary aneurysms is influenced by multiple factors like size, location, numbers, atherosclerotic burden, age, and comorbidities. Since the natural history and outcomes are subjective to variable factors, there is no single treatment for all patients, and management options must be tailored according to each patient.

In summary, CAAs can be a part of multisystem diseases like FRAM. The management should be individualized based on symptoms at presentation, size of coronary aneurysms, and local expertise.

Lead author biography

Dr Muhammad Azam Shah is a qualified cardiologist with a subspecialized qualification in echocardiography and cardiac CT with a keen interest in areas of research and development. After finishing cardiology fellowship from Pakistan, he joined King Fahad Medical City, Riyadh, Saudi Arabia, where he did his subspecialty training in advanced cardiac imaging. He is a Consultant cardiologist and working in the imaging section.

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 authors confirm that written consent for the submission and publication of this case report including images and associated text has been obtained from the patient and family in line with COPE guidance.

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