Giant coronary artery aneurysms: a case of progressive dyspnoea

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DESCRIPTION

A man in his late 50s with a history of ulcerative colitis and primary sclerosing cholangitis presented with 12 months of progressive shortness of breath. On examination, vital signs were within normal limits, and there was an elevated internal jugular vein and peripheral oedema. Electrocardiogram revealed a sinus rhythm and no sign of ischaemia. Chest X-ray showed gross cardiomegaly, which was not present 11 years earlier (figure 1A).

Transcatheter echocardiography showed normal systolic function, estimated left ventricular ejection fraction of 58% and reduced right ventricle size with normal systolic function. A follow-up stress echocardiogram showed no inducible ischaemia. Further imaging was pursued, and invasive coronary angiography demonstrated a large aneurysmal left main artery, aneurysmal left anterior descending (LAD) arteries, and significant aneurysmal dilatation in the right coronary artery (RCA) with poor opacification with contrast due to its size (figure 1B).

CT coronary angiography was performed and found all coronary arteries were grossly aneurysmal. The RCA aneurysm was the largest, at 110×110×130 mm in size at maximal dilatation, with thrombus formation and compression into the right heart causing shift of the ascending aorta, which in turn caused significant right pulmonary artery (RPA) compression. In addition, the right posterior descending artery branch was aneurysmal and enlarged, the LAD aneurysm was 70×66×80 mm in size, and the left circumflex artery was also aneurysmal (figure 1C).

No other arteries on whole body imaging were found to be aneurysmal. Extensive laboratory investigations looking for inflammatory vasculitides, systemic connective tissue diseases and infection/mycotic aneurysms, including C reactive protein, erythrocyte sedimentation rate, antinuclear antibodies, HLA-B27 and blood cultures, were all unremarkable. On detailed history, there were no obvious preceding illnesses or links to an aetiology. The patient suffered progressive dyspnoea and decompensated heart failure with repeated hospitalisations. The case was discussed at a multidisciplinary meeting with the cardiac surgeons. Due to
the length and extent of the disease, surgical options of grafting or resecting the aneurysm were deemed not feasible, so he was ultimately referred for cardiac transplant. Unfortunately, the patient died while on the waiting list.

Giant coronary artery aneurysms (CAAs) are a rare finding and occur in 0.02%–0.04% of the population.1 CAAs are termed giant if their diameter exceeds the reference vessel diameter by >4 times or if they are >8 mm in diameter and can confer a poor prognosis.2

Studies show the most common artery affected is the right coronary artery (40%–87%) followed by the circumflex artery (40%–20%) and left anterior descending (40%–87%).3 There are many aetiologies to consider for giant coronary artery aneurysms, such as thrombus formation, arterial rupture and external compression on surrounding structures.4

There is limited literature showing association to CAAs.4 Addi- tional differentials to consider in patients presenting with CAA include systemic inflammatory conditions (vasculitis, systemic lupus erythematosus, inflammatory arthropathies, Behcet’s syndrome), connective tissue disorders (Marfan syndrome, Ehlers-Danlos, fibromuscular dysplasia, polycystic kidney disease), infectious causes, drug related (coca, amphet- amines, protease inhibitors), and traumatic (percutaneous coronary intervention) and idiopathic causes. Patients with CAA are often asymptomatic, with the condition picked up incidentally on imaging. However, others may present symptomatic when there are complications from the aneurysms, such as thrombus formation, rupture or external compression on surrounding structures.4

There is no standardised management of CAA, with most evidence coming from case reports. Management is individual-ised and is usually based on the location, size, shape, progression and aetiology. Medical management focuses on mitigating cardiovascular risk factors. There is varied evidence for the use of anticoagulation in the presence of thrombus. Percutaneous management, such as covered stent implantation and coil embolis-ation, are becoming more common. Surgical techniques include aneurysmectomy with or without bypass grafts, depending on the distribution of the aneurysm; other options include aneu- rysmal ligation, resection, marsupialisation or transplant.5 The long-term evaluation of CAA appears to be largely extrapolated from those that arise secondary to Kawasaki disease, for which the American Heart Association has guidelines for follow up, including advanced imaging with which frequency is dependent on clinical status and results of stress testing.6

Learning points

- Giant coronary artery aneurysms are a rare entity with high morbidity and mortality.
- Complications from giant coronary artery aneurysms include thrombus formation, arterial rupture and external compression on surrounding structures.
- There are many aetiologies to consider for giant coronary artery aneurysms with the most common being atherosclerosis in adults and medium vessel vasculitis in children (Kawasaki disease).

REFERENCES