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Editorial

EDITORIAL

Coronary artery aneurysms—a truly rare entity or simply unrecognized so far?

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Coronary artery dilatations are characterized by an enlarged coronary artery lumen more than 1.5 times the diameter of normal adjacent segments or the diameter of the patient's largest coronary artery. They are divided into coronary artery ectasia and aneurysms. Coronary ectasias are diffuse and involve 50% or more of the length of the artery, while coronary artery aneurysms (CAAs) are more focal, involving <50% of the total length of the vessel. In children, according to the AHA classification a diagnosis of a CAA is based on a coronary artery Z score ≥2.5. CAAs are characterized as giant when the maximal diameter exceeds 20 mm in adults and 8 mm or a coronary artery Z score ≥10 in children [1, 2]. The biggest CAA described had a maximum diameter of 180 mm [3].

Aneurysmal coronary artery disease is a relatively uncommon entity found in 0.3–5% of adult patients undergoing invasive coronary angiograms. Giant CAAs are very rare, with an estimated prevalence of 0.02–0.2% [4], however, they have been reported in 5.9% of patients with congenital coronary artery fistulas [5]. The underlying aetiology for CAAs varies geographically and with age; atherosclerosis accounts for around half of the cases identified in the Western world, while Kawasaki disease is more common worldwide with development of CAAs in 12–25% of untreated patients. Other causes include connective tissue disease, congenital disease, vasculitis/inflammation and trauma/iatrogenic [6, 7]. Giant CAAs may present with angina, especially when they are related to atherosclerotic cardiovascular disease, or may be an incidental finding in asymptomatic patients. Because of the rarity of giant CAAs, the exact natural progression is unknown, and treatment is not well established. Surgery is usually performed in patients with obstructive coronary artery disease, angina, arrhythmias or other complications, while asymptomatic patients especially those with untreated Kawasaki disease can be offered antiplatelet or antithrombotic treatment [4, 8, 9].

The case report by Ab Hamid and Joshi [10] published in the Oxford Medical Case Reports, describes the very rare presentation of an asymptomatic 71-year-old patient with giant CAAs in all three coronary arteries. Cardiomegaly on chest X-ray for pneumonia prompted further investigation with echocardiography and then cardiovascular magnetic resonance (CMR) imaging and computed tomography coronary angiogram (CTCA). Interestingly, despite their size and the presence of circumferential intramural thrombus the giant CAAs caused no obstruction to adjacent cardiac chambers, no evidence of cardiac ischaemia on stress echocardiography and there was no myocardial infarction on CMR late gadolinium imaging. The authors concluded that the giant CAAs in their patient could reflect either undiagnosed childhood Kawasaki disease or they might be congenital due to coexisting multiple dilated venous structures in the chest. Nevertheless, atherosclerosis is the most common aetiology of CAAs in the general population, and in this patient atherosclerosis could well be the explanation as coronary artery calcifications were evident on the CT coronary angiogram.

This case illustrates the important role of advanced modern imaging modalities in diagnosing disease entities that were previously considered rare. CTCA has shown high accuracy for diagnosing obstructive coronary artery disease [11], and it has

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been proposed in the United Kingdom NICE guidelines [12] as the non-invasive, cost-effective test of choice, that can act as a gate keeper for invasive catheterization. This is projected to result in an increase in the number of cardiac CT scans across the UK by 700% [12] and as a consequence an increased rate of recognition of coronary artery dilatations. CTCA can provide information about the number, the exact location, the size of the aneurysms, the layers of the vessel wall involved, the presence of intraluminal thrombi and the associated complications.

CMR has further changed the dynamic of non-invasive imaging by providing simultaneous assessment of ventricular function, wall motion, myocardial perfusion, and tissue characterization in a single examination without potentially harmful ionizing radiation. It has gained an important role in the evaluation and management of patients with coronary artery disease [13]. Although CMR coronary angiography remains technically challenging, it can precisely delineate CAAs in the proximal segments of coronary arteries and assess for complications such as intraluminal thrombosis, myocardial ischaemia or infarction. Furthermore, MR angiography can provide a radiation-free evaluation for aneurysms in other arterial beds [14]. CMR thus offers a valuable tool for the initial assessment, risk stratification and long-term surveillance of patients with CAAs, especially in young patients with previous Kawasaki disease where the cumulative risk of life-time radiation exposure is high [15].

To conclude, the increasing use of advanced non-invasive modalities to image the coronary arteries and to assess cardiac function will lead to increased recognition of entities such as coronary artery aneurysms that were previously thought to be uncommon. As a result, more data on the true burden, the disease course, treatment options and outcomes are needed to make therapeutic decisions especially in asymptomatic patients.

CONFLICT OF INTEREST STATEMENT

No conflicts declared.

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