Cardiology in the Young

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Chest pain due to coronary artery compression in an adult with CHD

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Brief Report

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Abstract

A 47-year-old with repaired ventricular septal defect and pulmonary valve stenosis as a child presents with chronic intermittent chest pain. CT evaluation for transcatheter pulmonary valve replacement revealed right coronary artery compression between a sternal wire and dilated right ventricle. Removal of the sternal wire resulted in improved symptoms.

History of presentation

A 47-year-old female with a history of ventricular septal defect and congenital pulmonary stenosis underwent ventricular septal defect patch closure and pulmonic valve repair at age 5 years. She had no further interventions and had limited congenital cardiac follow-ups. She presented to our adult CHD clinic to re-establish care. She reports long-standing and progressive dyspnea with exertion (NYHA class II), palpitations, exercise intolerance,and near syncopal events. She had chronic intermittent pressure, 5/10, reproducible left-sided chest pain since she was in her early twenties. She otherwise denied orthopnea or paroxysmal nocturnal dyspnoea. Her physical exam was pertinent for a well-appearing, though somewhat obese, female of her stated age. She had a well-healed midline sternotomy scar and mild 1+ pedal oedema of the left lower extremity. Auscultation revealed a normal first heart sound, single second heart sound, and ejection click with a 3/6 systolic ejection murmur, as well as a 2/4 early diastolic murmur.

Past medical history

Review of available medical records confirmed ventricular septal defect and congenital pulmonary valve stenosis, followed by repair as a child. She also related a remote diagnosis of chronic pericarditis for which she previously had been on azathioprine and colchicine. Finally, she had a history of deep venous thrombosis in the left lower extremity status post treatment but with continued mild left-sided pedal oedema, and obesity s/p gastric bypass surgery two years prior to presentation.

Chest pain was thought to be due to chronic stable angina related to undiagnosed coronary artery disease or due to acute intermittent exacerbation of chronic pericarditis.

A 12-lead electrocardiogram demonstrated sinus rhythm, a right bundle branch block with a QRS duration of 148 milliseconds, and non-specific ST segment changes. Transthoracic echocardiogram demonstrated low normal left ventricular systolic function, moderate-severe right ventricular dilation with qualitatively mildly reduced right ventricular systolic function, evidence of biventricular diastolic dysfunction, dysplastic pulmonary valve leaflets with non-coaptation resulting in free pulmonary insufficiency, mild-moderate tricuspid regurgitation, mildly dilated main and branch pulmonary arteries, and no pericardial effusion or thickening.

She underwent Holter monitoring, which demonstrated multiple non-sustained runs of supraventricular tachycardia and multiform premature ventricular complexes.

Functional retrospective electrocardiogram gated cardiac CT scan was obtained for possible transcatheter pulmonary valve replacement, demonstrating severe right ventricular dilation (RV end-diastolic volume 218 ml/m^2) and mildly depressed systolic function (RV ejection fraction 48%), normal left ventricular size and function (LV end-diastolic volume 96 ml/m^2 and ejection fraction 65%), and normal pulmonary valve annulus with non-functional leaflets. Surprisingly, the right coronary artery was found to be torturous, taking an unusual course away from the right atrioventricular groove anteriorly towards the sternum before travelling inferiorly directly between the sternum and the dilated anterior right ventricle surface. The right coronary artery measured 1 mm (>50% narrowing) as it coursed between the anterior right ventricle surface and a lower sternal wire (Fig. 1a, b), which was concerning for right coronary artery distal flow compromise between dilated right ventricle and sternum.

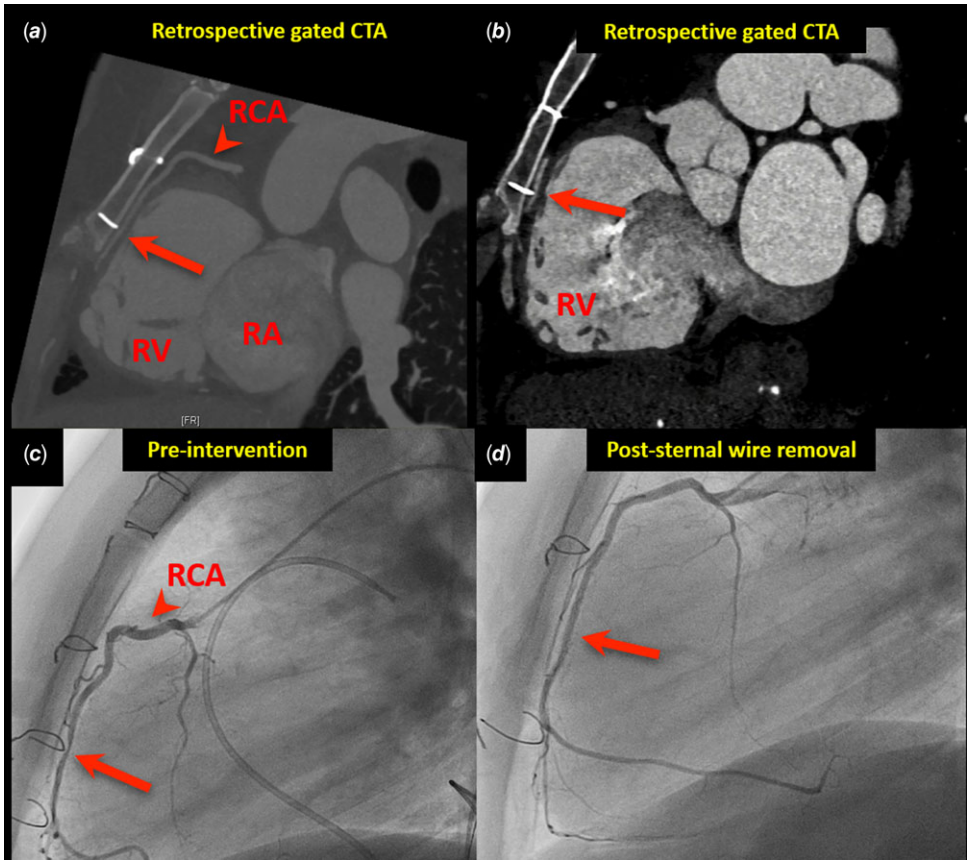

Figure 1. (***a***) and (***b***) Shows saggital and double oblique projections of retrospective gated cardiac CT angiography demonstrates significant stenosis of the mid-right coronary artery as it courses between the dilated right ventricle and a sternal wire. (***c***) Selective angiography of the right coronary artery that demonstrates 48% stenosis of the mid-right coronary artery as it courses between the dilated right ventricle and the sternal wire. (***d***) Selective angiography after the removal of the offending sternal wire demonstrates resolution of RCA stenosis. RA = Right atrium; RV = Right ventricle; RCA = Right coronary artery.

Management

Given the findings of the investigations, she underwent cardiac catheterisation with selective angiography of the coronary arteries. This confirmed 48% narrowing of the mid-right coronary artery as it coursed between the dilated RV and sternal wire (Fig. 1c). She then underwent surgical removal of the compressing sternal wire. Repeat angiography showed no residual right coronary artery stenosis (Fig. 1d). She also underwent electrophysiology study the same day, which demonstrated an intra-atrial re-entrant tachycardia that was successfully mapped and ablated.

Discussion

This case demonstrates an atypical cause of a long-standing chest pain along with dyspnoea on exertion and exercise intolerance. Concurrently, it demonstrates the diagnostic difficulties associated with evaluating adults with CHD, as it is often necessary to evaluate for sequelae of the underlying CHD and repair, as well as acquired heart disease. While it is not uncommon for severe right ventricle dilation due to pulmonary insufficiency and arrhythmia to cause similar symptoms, coronary compression from a sternal wire is an uncommon cause that has only been previously reported once in an adult that underwent mitral valve replacement not due to CHD.[1] There are case reports of meandering right coronary arteries away from right atrioventricular groove in a patient with Tetralogy of Fallot and ascending aortic dilatation, respectively.[2,3] The real incidence of this particular association is not known; it is estimated about 1% of Tetralogy of Fallot subjects have a large conal branch from right coronary artery, thus making this association even rarer.[4] Congenital cardiologists and surgeons are usually cognizant of the right coronary anomalous course or prominent branches, especially if they need a congenital heart surgery or a redo sternotomy, to avoid injury to the right coronary artery.[2] Patients with severe right ventricle dilatation can experience coronary compression and symptoms as in our patient. These anomalous coronary courses are considered normal variants and can often be an afterthought in the patient's care until they become an issue such as in this case. Therefore, as more and more patients born with CHD are surviving well into adulthood, we need to keep atypical causes of chest pain such as in this case in mind.

Follow-up

Upon follow-up, her chest pain had markedly improved. She underwent successful transcatheter pulmonary valve replacement with Alterra adaptive pre-stent and 29 mm Sapien 3 valve placement. This procedure was uncomplicated; she had transient post-procedural arrhythmia that was medically managed with sotalol. Her functional status continues to improve on further follow-up.

Conclusion

We report an unusual case of right coronary artery anomalous course away from the right atrioventricular groove directly between the dilated right ventricle and sternal wire resulting in compression and long-standing reproducible chest pain that went unrecognised for over two decades. Knowledge of these unusual coronary variants is important for appropriate diagnosis and management. This case highlights the difficulties in taking care of adults with CHD, as they often suffer from many sequelae related

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to their underlying CHD and multiple interventions. This is further complicated by the remote nature of the interventions, associated anatomical variants such as in this, and the fact that these patients are not immune to acquired heart diseases that commonly afflict the general population.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951123004274.

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Competing interests. None.

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