Gigantic arteriovenous fistula between left coronary cusp and coronary sinus draining into the right atrium

Syed Raza Shah a, Imtiaz Ismail b, Munis Raza a and Sohail Ikram c

aCardiology Fellow, University of Louisville, Louisville, KY, USA; bInterventional Cardiology Fellow, University of Louisville, Louisville, KY, USA; cFACC, University of Louisville, Louisville, KY, USA

ABSTRACT
Coronary arteriovenous fistulas (CAF) are infrequent anatomic anomalies that establish a direct connection between coronary arteries and cardiac chambers. The reported incidence is extremely low and estimated at 0.002% in the general population. We report a rare case of CAF in a middle-aged man, who was asymptomatic but incidentally found to have a gigantic CAF on a low-dose Computed Tomography scan of his chest. The case was presented to cardiothoracic surgeons. Since the patient was asymptomatic, they recommended medical management and continued close surveillance. The Left Coronary Artery or its branches are extremely uncommon site for CAF. With the advances in technology, the network of veins including coronary sinus has gained further clinical relevance. While technology has helped elucidate many aspects of these rare anomalies, mysteries still remain. With continued research, we can expect more cost-effective and less invasive interventional therapies to be developed in the near future.

1. Introduction
Coronary sinus (CS) is a small tubular structure, just above the posterior left atrioventricular junction, responsible for draining most of the deoxygenated blood from the myocardium. Even though the CS is a major access site for many invasive cardiology procedures, this venous system barely receives any attention as healthcare professionals rarely encounter diseases related to this system.

Coronary arteriovenous fistulas (CAF) are infrequent anatomic anomalies that establish a direct connection between coronary arteries and cardiac chambers. These are rare and may be traumatic, congenital or iatrogenic [1,2]. The reported incidence is extremely low and estimated at 0.002% in the general population [3]. The right coronary artery is the most common site of origin, and CS is rarely the draining site. CAF are reported to range in size from 2 to 5 mm [3]. They are generally asymptomatic but occasionally cause severe symptoms, including dyspnea, chest pain and palpitations, depending on the severity of the shunt [1–3]. They may be treated medically, surgically or by transcatheter closure [3,4]. We report a rare case of CAF in a middle-aged man, who was asymptomatic but incidentally found to have a gigantic CAF on a low-dose Computed Tomography (CT) scan of his chest.

2. Case report
A 65 year-old asymptomatic man came to the cardiology clinic after a low-dose CT scan, which was initially done as a preventative health screening tool for lung cancer screening, showed marked enlargement of his left circumflex coronary artery, measured at 18.9 mm, as an incidental finding (Figure 1). Patient was subsequently referred for a cardiac catheterization for further evaluation (Figures 2–4). There was no family history of congenital heart disease or other inherited abnormalities. The vital signs were normal. Physical examination was unremarkable. EKG showed normal sinus rhythm with no abnormalities. Chest X-rays showed only mild cardiomegaly. Coronary angiography revealed a huge vessel originating from the left coronary cusp that appeared to be draining into the right atrium. It was not clear from the angiogram if this vessel was originating from left coronary artery or directly from the left cusp. Because of its huge diameter, this vessel required a guiding catheter for optimal filling. There were no significant atherosclerotic lesions in the coronary arteries. No significant oxygen step up was noted in the right sided chambers and no pulmonary hypertension was seen. The case was presented to cardiothoracic surgeons. Since the patient was asymptomatic, they recommended medical management and continued close surveillance.
Figure 1. CT showing markedly enlarged left circumflex coronary.

Figure 2. Huge AVF seen during the cardiac catheterization procedure.

Figure 3. Huge AVF seen during the cardiac catheterization procedure.
3. Discussion

The Left Coronary Artery (LCA) or its branches are extremely uncommon site for CAF [4]. Low pressure structures including the right-sided heart chambers, pulmonary artery and CS are the most common draining sites of CAF [4]. Although a left-to-right shunt exists in majority of the patients, the shunt size most often is small and has no clinical significance on myocardial hemodynamics [4,5]. The clinical presentation is generally dependent on the degree of the shunt and the size of the fistula. Literature has shown potential complications, which include myocardial ischemia secondary to embolization or steal phenomenon and sudden cardiac death [6,7].

The closure of the fistula is recommended when it is symptomatic, while the treatment in asymptomatic patients remains controversial. Large coronary fistulas should be closed by transcatheter or surgical treatment, regardless of symptoms, while small to moderate size fistulas should be treated only if they cause symptoms [6–9]. Treatment for clinically significant CAF have focused on surgical management. Since the first successful surgically treated CAF in 1947, several catheter-based interventional techniques have been developed including Tangential Arteriotomy without the need for cardiopulmonary bypass (CPB), Transcatheter Embolization with various types of devices, and use of Covered Stents [7–10]. Despite the increasing use of these modalities, the intracardiac closure of the fistula by CPB with or without coronary artery bypass graft remains the standard method [8,11].

With the advances in technology, the network of veins including coronary sinus has gained further clinical relevance. While technology has helped elucidate many aspects of these rare anomalies, mysteries still remain. With continued research, we can expect more cost-effective and less invasive interventional therapies to be developed in the near future.

Disclosure statement

No potential conflict of interest was reported by the authors.

ORCID

Syed Raza Shah http://orcid.org/0000-0002-7404-1513

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