Contents lists available at ScienceDirect

International Journal of the Cardiovascular Academy

journal homepage: www.elsevier.com/locate/ijcac

Short communication

Surgical management of a rare circumflex coronary artery fistula draining into the coronary sinus

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ARTICLE INFO

Article history:

Received 8 December 2016

Received in revised form 4 August 2017

Accepted 7 August 2017

Available online 8 August 2017

Keywords:

Coronary artery fistula

CAF

Left circumflex artery

Coronary sinus

Intracardiac shunting

ABSTRACT

Coronary artery fistula (CAF) is a rare anomalous connection between a coronary artery and a cardiac chamber or a major vessel. The majority arise from the right coronary artery and drain into the right heart structures. In this report, we present a rare case of a left circumflex coronary artery fistula draining into the coronary sinus managed successfully with surgical intervention.

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Introduction

Coronary artery fistula (CAF) is defined as an abnormal connection between a coronary artery and a cardiac chamber or any of the great vessels.¹ The majority of CAFs are congenital and not gender specific. In adults, coronary artery fistula (CAF) with pathologic intra-cardiac shunting is very uncommon (<0.002% of the general population), as most are detected and repaired during childhood.^{1–4} The left circumflex coronary artery (LCX) is rarely involved (up to 17% of all cases), and drainage of CAF into the coronary sinus (CS) is found in 3–7% of all cases (see Table 1).

Case report

A 41-year-old woman with no significant past medical history presents with shortness of breath for the last 3 months. She had no constitutional symptoms except dyspnea and chest discomfort with moderate exertion which had affected her ability to carry out normal daily activities. She was hemodynamically stable and cardiovascular exam was negative for abnormal findings. Chest x-ray and 12 lead ECG were normal. Transesophageal echocardiography showed disturbed flow signal at the proximal CS, which appeared to be dilated (Fig. 1A). There was no regional wall dilation, motion abnormality, or valvular dysfunction. Cardiac catheterization confirmed a large CAF with origin at the distal

LCX converging with the proximal CS (Fig. 1B, white arrow). Due to the size and location of the fistula the patient was not considered for percutaneous approach by the interventional cardiology service, and an open approach on cardiopulmonary bypass was offered. Intraoperatively, pulmonary artery catheterization confirmed a physiologic left to right shunt with significant step up in mixed venous oxygenation (SvO₂) to 92%. Via median sternotomy and cardioplegic arrest, the fistula was identified (Fig. 1C) and ligated. The SvO₂ returned to 70% immediately upon ligation of the fistula. Post op day 1 she reported complete

Table 1
Incidence of coronary artery fistulas.^{1–4}

Origin	Incidence
Right coronary artery	50%–60%
Left anterior descending artery	25%–42%
Circumflex artery	17–18.3%
Diagonal branch	1.9%
Left main coronary artery or circumflex-marginal branch	0.7%
Drainage	
Pulmonary artery	15%–43%
Right ventricle	14%–40%
Right atrium	19%–26%
Left ventricle	2%–19%
Coronary sinus	3–7%
Left atrium	5%–6%
Superior vena cava	1%

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Peer review under responsibility of The Society of Cardiovascular Academy.

Fig. 1. A) Color Doppler echocardiogram showing disturbed flow signal at the coronary sinus. B) Distal LCX CAF converging on the coronary sinus. C) Intraoperative view of the LCX CAF.

resolution of symptoms and was discharged 4 days later. At 6 month follow up, she was free of symptoms and back to her daily routine.

Discussion

In the setting of a symptomatic adult, urgent CAF exclusion (via open or endovascular approaches) constitutes optimal management. If left untreated the patient is at risk to develop highly morbid fistula related complications which include steal syndrome, dysrhythmias, endocarditis, and myocardial infarction.⁵ Moreover, CAFs that drain to the coronary sinus are particularly associated with progressive CHF, as we found in our patient. An open approach is indicated when the anatomy is not suitable (i.e. large fistula and/or aneurismal coronary artery) to undergo percutaneous closure and when there is additional pathology that can be corrected surgically. In our patient, surgical ligation was feasible because there was no transmural involvement and the large fistula was located at the distal end of the LCX. Surgical techniques for closure are associated with 16–22% risk of recurrence when compare to 10–50% with endovascular techniques.⁶ Another interesting finding in our patient was the usefulness of SvO₂ to identify the intracardiac shunt. Normalization of the SvO₂ upon ligation confirmed that the intervention had been successful from a physiologic stand point. In closing, large CAFs with high flow rates are often symptomatic and can lead to life-

threatening complications. Prompt intervention is indicated and surgical approaches still offer excellent short and long term outcomes.

Conflict of interest

The authors declare no conflict of interest.

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