Case Report

Left circumflex artery fistula to left atrium: a rare case report

Kaoutar Imrani*, Sanae Amalik, Nabil Moatassim Billah, Ittimade Nassar

Radiology department, Ibn Sina University Hospital, Mohammed V University, Rabat

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ABSTRACT

Coronary artery fistulas are rare congenital anomalies of coronary termination. There are 3 types: (1) cameral coronary fistulas, (2) pulmonary coronary fistulas, and (3) bronchial coronary fistulas. Left circumflex coronary artery to left atrial fistula are exceptional. Imaging, especially the CT angiogram, must establish an anatomical classification of the fistula for therapeutic purposes, by specifying its origin, its path, its size and its termination.

We report a rare case of the left circumflex coronary artery to left atrial fistula in a 31 year old man, which is an uncommon presentation of coronary termination anomalies. The CT coronary angiogram must describe the origin segment of the fistula and the drainage site which have therapeutic consequences.

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Introduction

Coronary artery fistulas (CAF) are rare and most often asymptomatic. CT coronary angiogram allows to characterize these fistulas by specifying their origin, their path and their drainage site, which has therapeutic consequences. There are 3 types: (1) cameral coronary fistulas, (2) pulmonary coronary fistulas, and (3) bronchial coronary fistulas. They rarely drain into the left atrium and left ventricle [1].

Case report

A 31 years old man, chronic smoking, consults for a mild shortness of breath during ordinary activities, stage 2 of NYHA (New York heart association), evolving for a few months. The clinical examination found a heart rate of 69 beats per minute and a blood pressure of 13/8 cmHg. Cardiovascular examination found no peripheral signs of heart failure. The rest of the somatic examination was normal. The electrocardiogram registers a regular sinus rhythm without repolarization disorders or parietal hypertrophy.

Transthoracic echocardiography and color Doppler showed dilated left coronary artery with a supernumerary vessel communicating with the left cardiac cavities without obvious shunt.

Coronary angiography demonstrated a fistulous connection between the left coronary artery and the left atrium (Fig. 1).

The CT coronary angiogram showed a fistula measuring 2 mm in diameter originating from the common trunk of the left
**Fig. 1** – Coronary angiography showing a fistulous connection between the left coronary artery and the left atrium (arrow).

**Fig. 2** – CT coronary angiogram showing a fistula originating from the common trunk of the left coronary artery, describing a tortuous path (Red arrow: common trunk, Blue arrow: the anterior interventricular artery, Green arrow: circumflex artery, Yellow arrow: fistula, white arrow: tortuous path with collateral circulation). (Color version of figure is available online.)

**Fig. 3** – CT coronary angiogram in volumetric construction showing the left coronary artery fistula (white arrow).

**Fig. 4** – CT coronary angiogram showing the drainage of the fistula (arrow) at the left atrium (star).

**Discussion**

Coronary artery fistulas are rare congenital anomalies of coronary termination affecting 0.003% of the general population, accounting for 0.3% of congenital heart diseases. It means abnormal connection between the coronary artery and a cardiac cavity or blood vessel without going through the capillary circulation. There are 3 types: (1) cameral coronary fistulas (coronary cavitary fistula), (2) pulmonary coronary fistulas, and (3) bronchial coronary fistulas [1,2].

They may be congenital or acquired. Congenital causes are the most common, they are due to the persistence of myocardial sinusoids, which is connected to the primitive tubular heart, this will result in communications between coronary arteries and cardiac cavities. As for coronary arteriovenous fistulas, they are due to the persistence of embryonic remains.
connecting the coronary arteries to a mediastinal vessel or to the superior vena cava [1,3,4].

Acquired causes can be post-traumatic, post-myocardial infarction, or iatrogenic (catheterization, surgery) [1,3,5].

The CT coronary angiogram, compared to other imaging techniques, provides a better characterization of coronary fistula and a high spatial resolution. Imaging must establish an anatomical classification of the fistula for therapeutic purposes, by specifying its origin, its path, its size and its termination [6,7].

Fistula can originate from the right or the left coronary artery (common trunk, anterior inter ventricular artery, circumflex artery).

The origin segment includes 2 types according to Sakakibara, proximal CAF (type A) which originates from a major epicardial coronary artery. The short proximal segment of the coronary artery upstream of the fistula’s departure is dilated and, downstream, the caliber and branches of the coronary artery are normal. There are no major coronary branches that feed the myocardium originating from the fistula, although there is a lower risk of infarction during surgical or endovascular treatment [1,8,9].

Distal CAF (type B) originates from the distal extremity of a major coronary artery. The coronary artery upstream of the distal fistula gives normal branches that vascularise the myocardium. The increase in flow in the coronary artery results in dilation and the development of tortuosities of the coronary artery proximal to fistula. In this type, the thrombotic risk post-intervention is high [1,9,10].

The drainage site of the coronary fistula can be cameral or vascular. For cameral drainage, it usually involves low pressure right cardiac cavities, commonly the right ventricle, followed by the right atrium. A drainage to low pressure cavities may cause aneurysm formation. CAF rarely drain into the left atrium and left ventricle (3%-5%), as in our patient [1,11].

Left circumflex coronary artery to left atrial fistula, as in our patient, has been rarely reported in the littérature [2,12].

On the other hand, Vascular drainage can be done in the superior vena cava, the innominate venous trunk, the pulmonary artery or in the bronchial arteries. Drainage sites may be single or multiple with multiple channels.

Finally, it is necessary to identify the last branches with myocardial destiny to avoid ischemic accident [13].

**Conclusion**

Cameral coronary artery fistulas are an uncommon presentation of coronary termination anomalies. They rarely drain into the left cardiac chambers. The CT coronary angiogram must describe the origin segment of the fistula and the drainage site which have therapeutic consequences.

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**Patient Consent**

Written informed consent for publication was obtained from the patient.

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