Most coronary artery fistulas (CAFs) have a congenital origin. Acquired CAF is a rare entity that may occur without an identifiable causative link. We present here the case of an acquired CAF in which optical coherence tomography was instrumental in outlining that its possible cause was a spontaneously ruptured, communicating, subadventitial coronary hematoma.

Case Presentation

A man aged 67 years was admitted for oppressive chest pain and sudden-onset dyspnea. He was a former smoker, had dyslipidemia and diabetes mellitus, and was in permanent atrial fibrillation. He also had had 2 mechanical valves implanted in the mitral and aortic positions in 1993 and 2003, respectively. No intervention in the tricuspid valve was performed. Before the second surgery, a coronary angiography revealed normal anatomy and the absence of obstructive atherosclerosis (Figure 1). At admission, he was normotensive and had mild signs of systemic congestion. A continuous soft murmur on the left sternal border was also heard. The ECG showed atrial fibrillation and a previously known complete left bundle-branch block with secondary ST-T changes. Transthoracic echocardiography showed normal function of both valves and of the left ventricle. Finally, the international normalized ratio was 4.0, and serial cardiac marker assays uncovered a rise in troponin I (peak, 5.29 U). Thus, myocardial infarction in the presence of a left bundle-branch block was diagnosed, and coronary angiography was scheduled. This study revealed minimal luminal irregularities in the left anterior descending and circumflex arteries and an acquired CAF from the right coronary artery to the right chambers (Figure 2 and Movie I in the online-only Data Supplement). Optical coherence tomography (Ilumien, St. Jude Medical, St. Paul, MN) revealed a relatively healthy vessel with a normal 3-layer appearance of the coronary wall in almost all the segment studied. However, distally to the site of the CAF, a vascular structure compatible with vasa vasorum was found. Subsequently, toward the CAF, the media layer became gradually expanded, accompanied by a low-signal region outside the external elastic membrane, compatible with an intramural hematoma extending to the subadventitial space (Figure 2). At the fistula origin, optical coherence tomography demonstrated a discontinuity in the vessel wall and irregular structures with a dorsal shadow compatible with red thrombus. Thus, a spontaneously ruptured, communicating, subadventitial hematoma was proposed as the cause of the acquired CAF. His case was presented to our Heart Team, and the patient was admitted for percutaneous treatment. Finally, a polytetrafluoroethylene-covered stent 3.0×16 mm (Jostent Coronary Stent Graft, Jomed, Germany) was deployed percutaneously and completely sealed off the acquired CAF (Movie II in the online-only Data Supplement). His recovery was uneventful.

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Discussion

Intramural coronary hematomas are defined as an accumulation of blood within the media that displaces the internal elastic membrane inward and the external elastic membrane outward, with or without identifiable entry and exit points. This phenomenon has been found after percutaneous coronary intervention and has been suggested as a cause of acute coronary syndromes and spontaneous coronary artery dissections. In the latter, it has been proposed that a primary disruption of the vasa vasorum within the media can lead to an intramedial hemorrhage, which ultimately produces an intimal tear. Importantly, in the absence of atherosclerosis (as in our case), it has also been suggested that the dissection plane more probably lies in the outer third of the media or at its junction with the adventitia, leading to a subadventitial noncommunicating hematoma. Finally, after the rupture of the internal elastic membrane, the subadventitial hematoma may access the true lumen of the vessel, producing spontaneous coronary artery dissections. Our case is extraordinary because, in an artery without evidence of atherosclerosis, a subadventitial hematoma was probably able to gain access not only to the vessel lumen but also to the inside of the right cardiac chambers, therefore being responsible for the acquired CAF.

CAFs are rare vascular anomalies characterized by an abnormal communication between a large epicardial artery and a cardiac chamber or thoracic vessel. The vast majority of these anomalies are congenital in origin. However, acquired CAFs have been rarely reported and attributed mainly to coronary trauma, including pacemaker placement, endomyocardial biopsy, blunt trauma, therapeutic chest radiation, barotrauma after percutaneous coronary angioplasty, myocardial infarction, and cardiac surgery. In our case, the absence of flow-mediated dilatation of the proximal artery and the clinical picture strongly suggest an acute establishment of the acquired CAF at the present event. However, even when a 9-year period separates the last surgery from the clinical presentation, it is unknown whether the development of the subadventitial hematoma was related to the surgical procedures, chronic anticoagulation, or other factors. To the best of our knowledge, ours is the first case in which an acquired CAF was studied further with optical coherence tomography. This technique provided additional insights into this rare anomaly and allows us to propose the spontaneous rupture of a subadventitial hematoma.

Figure 2. A, Coronary angiogram of the right coronary artery revealing the acquired coronary artery fistula (CAF) between the vessel lumen and right cardiac chambers. B, Optical coherence tomography (OCT) of the distal vessel revealing a normal 3-layer structure, with both the internal and external elastic membranes clearly visible (*). C, A few millimeters distal to the CAF, a well-limited poor-signal structure was found, compatible with vasa vasorum (arrow). D, At the site of the acquired CAF, OCT showed a discontinuity in the vessel wall and irregular structures with a dorsal shadow compatible with red thrombus (+). In the border of the rupture, the space between the internal and external elastic membranes was expanded (arrows). This finding is compatible with an intramural hematoma. E, Just proximal to the acquired CAF, the 3-layer structure of the vessel is recovered. However, the space between the internal and external elastic membranes (*) remains expanded, and a poor-signal region is visible behind the external elastic membrane (arrows). These findings are compatible with a subadventitial hematoma. Please note that residual lining thrombus is present (+). F, Finally, the normal structure of the vessel is fully recovered. G, The long axes of the lumen and vessel wall proximal to, distal to, and at the site of the acquired CAF.
into a vessel lumen and a cardiac chamber as one of the possible mechanisms behind an acquired CAF.

Disclosures
Drs Escaned and Gonzalo have served as speakers at educational events organized by St. Jude Medical. The other authors report no conflicts.

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Optical Coherence Tomography Findings in an Acquired Coronary Fistula
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