



## Case Report

# A Rare Coronary Anomaly Coincidentally Encountered During Primary Percutaneous Coronary Intervention: A Coronary-Pulmonary Artery Fistula

 Cagdas Kaynak,<sup>1</sup>  Sitki Kucukbuzcu,<sup>1</sup>  Izzet Okcesiz<sup>2</sup>

<sup>1</sup>Department of Cardiology, Siirt State Hospital, Siirt, Turkey

<sup>2</sup>Department of Radiology, Siirt State Hospital, Siirt, Turkey

### Abstract

Coronary-pulmonary artery fistula (CPAF) is one of the coronary anomalies rarely encountered in coronary angiography series. CPAFs are usually congenital but uncommonly remain asymptomatic in the advanced ages. Herein, we aimed to report our approach to a case of CPAF, which remained asymptomatic until advanced ages and was determined coincidentally in a patient undergoing percutaneous intervention for acute anterior myocardial infarction.

A 70-year-old male patient, who was admitted to the Emergency Department with extensive chest pain, hypotension, cold sweating, and impaired general status, underwent coronary angiographic intervention after being diagnosed with acute anterior myocardial infarction. During the intervention, a plexiform fistula originated from the conus branch of the right coronary artery and extended to the pulmonary artery was detected coincidentally. The 64-section computed tomography of the thorax confirmed that fistula drained into the left pulmonary artery. Taking the clinical and demographic characteristics of the patient into account, we decided medical follow-up as the first treatment option.

Nowadays, coronary fistulas are usually closed by coiling or surgery even they are asymptomatic. We are in the opinion that conservative approach would be more reasonable for asymptomatic fistulas in advanced age patients having additional cardiac pathologies.

**Keywords:** A coronary-pulmonary artery fistula

Uncommon types of coronary anomalies are encountered coincidentally owing to widespread use of interventional angiography recently. Coronary artery fistula is a type of coronary anomaly comprising an abnormal connection between one or more of the coronary arteries and other coronary vessels, abnormal vascular structures, cardiac chambers or great vessels arising from the heart.<sup>[1]</sup> The frequency of this uncommon situation firstly defined in 1865 by Krause is 0.002% in general population and it is coincidentally detected by 0.05%-0.2% during angiography procedures.<sup>[2, 3]</sup> Coronary-pulmonary artery fistulas (CPAFs) account for 20% of all coronary fistulas.<sup>[4]</sup> These

fistulas, which are generally asymptomatic, may become symptomatic particularly with aging and due to fistula enlargement. Herein, we present a single case of CPAF among nearly 1500 cases undergoing angiography in a state hospital, which was coincidentally detected in a patient undergoing primary percutaneous coronary intervention (PCI) for acute anterior myocardial infarction (MI).

### Case Report

A 70-year-old male patient with a history of documented asthma and heavy smoking (2 packet/day) was admitted to the Emergency Department of Siirt State Hospital with

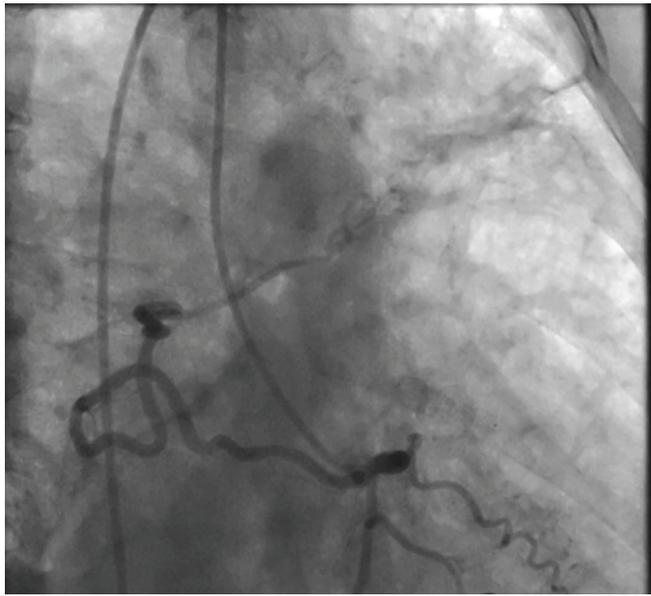
**Address for correspondence:** Cagdas Kaynak, MD. Siirt Devlet Hastanesi Kardiyoloji Anabilim Dalı, Siirt, Turkey

**Phone:** +90 484 224 81 61 **E-mail:** cagdaskaynak@hotmail.com

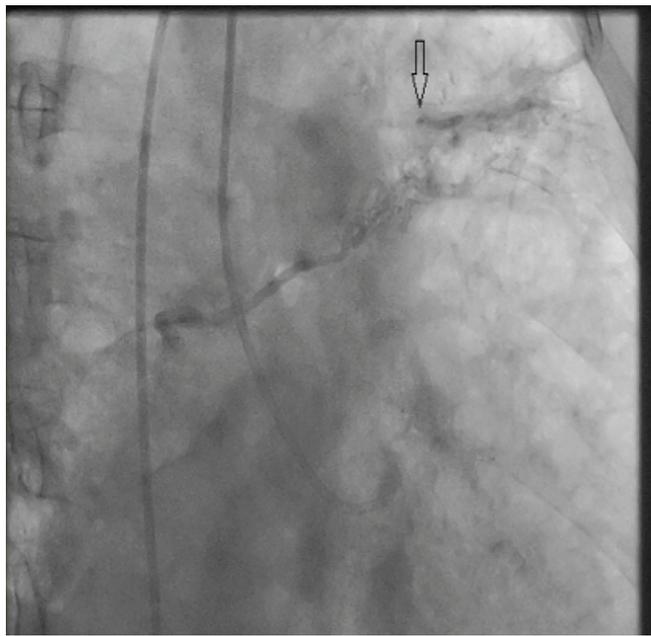
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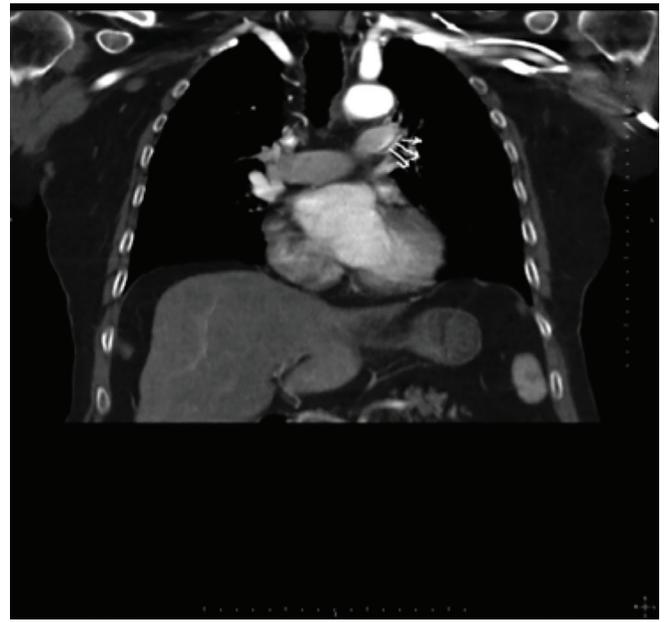


**Figure 1. (a)** A plexiform fistula originating from the developed conus branch of the right coronary artery and extending to the left pulmonary artery.



**Figure 1. (b)** Distal flow image of coronary-pulmonary artery fistula (about 2 mm diameter) originating from the developed conus branch of the right coronary artery and extending to the left pulmonary artery (arrow).

extensive chest pain, hypotension, cold sweating, and impaired general status. His blood pressure was 80/60 mmHg and pulse rate was 110 beats/min. His electrocardiography revealed normal sinus rhythm, a heart rate of 110 beats/min, and an ST-segment elevation of 4 mm in D1-aVL and V1-V6 derivations. The patient was commenced on an-



**Figure 2.** Sixty-four-section thoracic computed tomography coronal image of the coronary-pulmonary artery fistula (arrow).

ti-ischemic, antiaggregant, and anticoagulant treatments. Being diagnosed with acute extensive anterior ST elevation MI, the patient was referred to the hemodynamics Laboratory for primary PCI. His coronary angiography revealed thrombosis and total occlusion of the mid-segment of the left anterior descending (LAD) artery. A 6F guiding catheter was placed into the left coronary system and pre-dilatation was performed in the mid segment of the LAD artery at 12 atmospheric pressure using a 2.0x12 mm balloon. After pre-dilatation, a drug (sirolimus+organic acid) eluting stent (3.0x20 mm in size) was implanted. Thereafter, the stent was post-dilated at high atmospheric pressure using a 3.0x15 mm non-compliant balloon. Tirofiban perfusion was initiated. Subsequently performed right system diagnostic angiography revealed a developed conus branch of the right coronary artery (RCA) extending to the pulmonary artery trace where it was drained into; this was considered as CPAF (Fig. 1a). Distal flow diameter of fistula was about 2 mm (Fig. 1b). Elective examination was planned to assess the fistula; however, he developed extensive back pain 10 minutes later and his ST elevation remained on the control electrocardiography. His control angiography demonstrated acute thrombosis of the stent in the LAD artery. Sequential dilatations of the stent using non-compliant balloon were performed together with intracoronary tirofiban administration. Complete arterial flow was achieved. On his bedside transthoracic echocardiography, ejection fraction (EF) was 40%, the anterior lateral wall of the apex was hypokinetic, there were mild mitral and tricuspid insufficien-

cy, and pulmonary pressure was 25 mmHg. His 64-section computed tomographic angiography of the thorax performed after his clinical status became stable revealed a developed conus branch of the RCA draining into the left pulmonary artery; this finding confirmed CPAF diagnosis (Fig. 2). The fistula was considered suitable for coiling due to the facts that there were risk factors for surgery, the fistula originated from a single developed conus branch, and the fistula was suitable for cannulation by angiographic catheters. However, we planned to evaluate the fistula for coiling electively during follow-up visits due to the following reasons: the patient was extremely prone to thrombosis in the acute period due to acute MI and stent thrombosis, his EF reduced in the post-MI period, myocardium was in the stunning period and considered to be unable to tolerate a potential hemodynamic change during coiling, and although it was diagnosed in the advanced age, the fistula had a small diameter, caused no aneurysm, and was asymptomatic in the pre-MI period.

## Discussion

Coronary-pulmonary artery fistulas are usually congenital; however, acquired cases have also been reported after chest trauma, coronary angiography or bypass surgery.<sup>[5]</sup> Although some of the earlier studies have reported that coronary fistulas originate from the RCA, recent studies have stated that these fistulas more frequently originate from the LAD artery.<sup>[5-7]</sup> Coronary fistulas generally drain into the right cardiac structures by 90%.<sup>[6]</sup> In a systematic review, 63% of the CPAF cases were male and 89% of the fistulas drained into the main pulmonary artery.<sup>[7]</sup> Our case was an advanced age male patient having a CPAF originating from the conus branch of the RCA and drained into the left pulmonary artery.

Although CPAFs are rarely diagnosed during infancy, the diagnosis is most commonly established when they become symptomatic during adolescent period. The patients may develop angina pectoris, dyspnea, cardiac failure due to volume overload, cardiomyopathy, recurrent arrhythmias, pericardial effusion, and pulmonary hypertension. CPAFs may lead to coronary ischemia by causing vascular steal, to cardiac failure by causing volume changes, to infective endocarditis by causing tendency to infection, or to rupture by causing aneurysm. Thus, treatment decision depends on the origin and structure of fistula, the area where fistula drains into, as well as fistula's size, associated shunt, and resultant symptoms, and patient's age and additional cardiac pathologies.<sup>[4, 8, 9]</sup> Moreover, an asymptomatic fistula in early ages may become symptomatic with increasing age. CPAFs are encountered coincidentally in advanced age patients due to widespread use of angiography and

decreased cardiac-cause mortality in recent years. Treatment options for symptomatic cases include medical therapy, coiling via transcatheter route, stent grafting, and surgical ligation.<sup>[10]</sup> However, the literature has gaps about the approach to asymptomatic small fistulas in advanced ages and in the presence of additional cardiac pathologies. Our case was also an advanced age patient in whom the fistula originated from the developed conus branch of the RCA, the conus branch narrowed after a long course and formed a plexiform structure, and drained into the left pulmonary artery. The conus branch of the RCA appeared quite suitable for cannulation to perform coiling. However, considering the potential risks of coiling (embolus, fistula rupture, etc.), we thought that the procedure would be more risky during thrombosis-prone period if performed in a patient asymptomatic for fistula. On the other hand, particularly, evaluation of the termination site of the fistula revealed that the fistula formed a plexiform structure and drained into the pulmonary artery. Since the fistula opening is seen through the pulmonary artery in the fistulas ended as plexiform, surgical treatment is rather preferred to avoid residual fistulas.<sup>[5, 10]</sup> However, our case was obviously a high-risk patient also for surgery for the following reasons: advanced age, history of coronary artery disease, MI, asthma, and smoking, and reduced EF. Hence, we preferred conservative approach with medical follow-up as the initial treatment and we subsequently planned to evaluate the patient for coiling if the patient becomes symptomatic for fistula.

Earlier studies have mostly focused on approaching to fistulas in patients with normal coronary arteries. However, there is insufficient data on how to approach to coronary fistulas not responsible for clinical entity in patients presented with manifest coronary artery disease. We think that patients' characteristics as well as anatomical and hemodynamic structure of fistulas need to be considered in such cases while deciding therapeutic approach and type and time of intervention. Larger case-series are needed to gain experience on this subject.

## Disclosures

**Informed consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

**Peer-review:** Externally peer-reviewed.

**Conflict of Interest:** None declared.

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