Case report

Steal and strain: A case of coronary artery fistula presenting with coronary steal syndrome and underlying bronchiectasis

Joshua Tetteh Narh [a,*], Erum Zahid [b], Kiran Shivaraj [a], Sonu Sahni [a], Pramod Theetha Kariyanna [c], Abdullah Khan [d]

[a] Dept. of Internal Medicine, Brookdale Hospital Medical Center, Brooklyn, NY, USA
[b] Dept. of Pulmonary and Critical Care Medicine, Brookdale Hospital Medical Center, Brooklyn, NY, USA
[c] Dept. of Cardiology, State University of New York, Downstate Medical Center, Brooklyn, NY, USA
[d] Dept. of Cardiology, Brookdale Hospital Medical Center, Brooklyn, NY, USA

ABSTRACT

Background: Coronary artery fistula is a relatively rare disorder with an incidence rate of 0.05–0.9%, and the majority of fistulae are detected incidentally. Most coronary artery fistulae are congenital, and the acquired variant is very rare. Herein, we present a possible acquired coronary artery to pulmonary artery fistula, most likely secondary to bronchiectasis in the adjacent lung. We will analyze the hemodynamic significance of the fistula in this case and also seek to understand the outcomes of various treatment modalities.
Case presentation: A 56-year-old male patient presented with hypoxemia secondary to acute pulmonary edema during a hypertensive emergency. He developed myocardial ischemia after treatment with diuretics and nitroglycerin, due to shunting of blood from the right coronary artery to the right lower lobe branch of the right pulmonary artery, via the fistula. This resulted in coronary steal syndrome. Coronary angiogram confirmed the fistula connecting the right coronary artery to the right lower lobe branch of the right pulmonary artery. An attempt at coil embolization was unsuccessful due to the inability to advance the microcatheter beyond the fistula.
Discussion: The majority of coronary artery fistulae are asymptomatic as they are hemodynamically not significant and are incidentally detected by coronary angiography, CT angiogram, echocardiogram or multi-detector row computed tomography (MDCT) with 3D reconstruction. The development of congenital fistula can be explained by the Hackensellner involution-persistence hypothesis, but the anatomy in this case and the bronchiectasis in the part of the lung adjacent to the fistula makes an acquired cause very likely due to local inflammation and the age of patient at initial diagnosis. An initial diagnosis of bronchiectasis was made at age 51, which was 5 years prior to the detection of the coronary artery fistula in this patient. Symptoms have been described mostly in the elderly and include chest pain, dyspnea, fatigue, syncope, and palpitations. Such symptomatic fistula should be treated either by percutaneous transluminal embolization or surgical ligation.
Conclusion: This is a unique case of acquired coronary to pulmonary artery fistula in the setting of bronchiectasis in a patient in which PTE was attempted and failed. More research is required to understand the pathophysiology of acquired fistula. The decision regarding the method of closure should be individualized and decided on a case by case basis.

1. Introduction

Coronary artery fistula (CAF) is a rare congenital or acquired cardiopulmonary condition, that is defined as an abnormal vascular connection between one or more coronary arteries, either the pulmonary or systemic vascular network or a cardiac chamber without an interposing myocardial capillary bed [1]. CAF has been reported to have an incidence rate of 0.05–0.9% with the majority of cases being diagnosed during coronary angiography [2]. Furthermore, coronary-to-pulmonary artery fistula (CPAF) has been reported to make up 15–30% of all CAF cases, which are mostly detected by multi-detector row computed tomography (MDCT) with 3D reconstruction [3].

* Corresponding author. Department of Internal Medicine, Brookdale University Hospital Medical Center, Address: 1 Brookdale Plaza, Brooklyn, NY, 11212
E-mail address: jnarh@bhmcny.org (J.T. Narh).

Typically, CAF is asymptomatic; however, symptomatic patients present with chronic dyspnea and angina caused by myocardial ischemia or ventricular volume overload from a left-to-right or left-to-left shunt, thus resulting in either coronary steal phenomenon or heart failure, respectively. Therapeutic strategies are largely based on expert opinion due to the paucity of retrospective studies and randomized trials [4]. There exists only a few reported cases of CPAF with underlying bronchiectasis with the pathophysiological relationship remaining unclear. Herein, we present a CAF patient with coronary steal syndrome and acute on chronic combined systolic and diastolic heart failure in the setting of bronchiectasis. We also aim to analyze the hemodynamics and also attempt to explain the anatomical changes associated with the CAF.

2. Case presentation

A 56-year-old Asian male with a past medical history of essential hypertension, chronic bronchiectasis, thoracic aortic aneurysm and remote history of smoking of 10 pack years presented to the emergency room with progressively worsening shortness of breath. At baseline patient experienced dyspnea only on exertion but eventually developed shortness of breath at rest which was associated with orthopnea. Initial vital signs on arrival were a blood pressure of 255/138 mmHg, heart rate was 153 beats/min, respiratory rate of 44 breaths/min, oxygen saturation 74% on room air. Physical examination findings revealed a middle-aged male with a BMI of 24 kg/m^2, in respiratory distress, lung auscultation revealed decreased breath sounds at the bases with bibasilar crackles. Cardiac auscultation revealed continuous cardiac murmur, loudest at the left second intercostal space. Examination of extremities revealed prominent digital clubbing of the upper extremities.

Laboratory analysis revealed troponin (0.079 ng/mL) and pro-B-type natriuretic peptide (pBNP; 417 pg/mL) levels. Arterial blood gas analysis showed hypoxemia and respiratory acidosis. The rest of laboratory findings as stated in Table 1. Initial EKG showed sinus tachycardia and left ventricular hypertrophy with repolarization changes in the anterior chest leads (Fig. 1).

A portable anterior-posterior Chest x-ray showed possible left lower lung consolidation and cardiomegaly with a cardiothoracic ratio of 0.63. The patient was initiated on non-invasive positive pressure ventilation with bilevel positive airway pressure (BiPAP) support. However due to persistent hypoxia associated with worsening mental status patient was intubated and started on mechanical ventilation. Given elevated blood pressure, pBNP levels, and chest x-ray findings suggestive of pulmonary edema, nitroglycerin infusion was initiated for hypertensive emergency in conjunction with diuretics with an appropriate response. Repeat EKG revealed normal sinus rhythm with deep T-wave inversions in the inferior lateral leads (II, III, aVF, I, aVL, V4–V6) suggesting possible

Table 1
Laboratory findings on arrival in the Emergency room.

Hematology		Biochemistry			
WCC x 10^9/L	13.5	Na$^+$	136 mmol/L	LDH	1118 U/L
Neutrophils %	57.8	K$^+$	4.2 mmol/L	CRP	1.6 U/L
Lymphocyte %	33.0	Cl$^-$	95 mmol/L	Lactate	5.8 mmol/L
Hgb g/dl	15.8	BUN	27 mg/dl	CPK	199 U/L
Platelet x 10^9/L	280	Cr	1.44 mg/dl	TSH	1.86 mIU/L
ESR mm	9	Phosphorus	4.7 mg/dl	ALP	108 U/L
		Magnesium	2.6 mg/dl	T-Bil	0.6 mg/dl
		ALT	152 U/L	D-Bil	0.2 mg/dl
		AST	232 U/L		

ALP: Alkaline Phosphatase; ALT: Alanine aminotransferase; AST: Aspartate aminotransferase; BUN: Blood urea nitrogen; Cl: Chloride; CPK: Creatine kinase; Cr: Creatinine; CRP: *C*-reactive protein; D-Bil: Direct bilirubin; ESR: Erythrocyte sedimentation rate; Hgb: Hemoglobin; K: Potassium; LDH: Lactate dehydrogenase; Na: Sodium; T-Bil: Total Bilirubin; TSH: Thyroid stimulating hormone; WCC: White cell count.

coronary ischemia (Fig. 2). Repeat troponin level was still elevated (1.32 ng/mL). Patient was initiated on treatment for non-ST-elevation myocardial infarction with dual anti-platelets, statin, and anticoagulant. The patient initially received empiric antibiotic therapy due to concerns for underlying pneumonia, Covid-19 testing by a nasopharyngeal swab test was found to be negative. Repeat chest x-ray showed a marked improvement in the pulmonary congestion after diuresis (Fig. 3).

Of note the patient was known to the cardiology, pulmonary and cardiothoracic surgery service from prior admission in 2015 where he was worked up for chronic dyspnea. At that time CT angiogram of the chest revealed 4.8-cm aneurysmal dilatation of the ascending thoracic aorta and extensive bronchiectasis, primarily involving the right middle, upper, and left lower lobes (Fig. 4). Transthoracic echocardiogram (TTE) showed an aortic root diameter of 4.2 cm, an aortic valve opening of 1.65 cm, a left atrium diameter of 2.72 cm, an ejection fraction of 40–45%, left ventricular eccentric hypertrophy, an estimated peak pulmonary artery pressure of 50 mmHg, and normal right ventricular systolic function.

Coronary angiography that was performed 8 months prior to this admission showed a coronary-to-pulmonary artery fistula (CPAF) between the right coronary artery and right lower lobe branch of the right pulmonary artery, which corresponded to the area of pulmonary bronchiectasis seen in computer tomography. At that time a multidisciplinary team planned for the repair of the fistula on an outpatient basis but however, he was lost to follow-up.

During the patient's current admission, repeat TTE showed left ventricular dilatation, and visual assessment displayed reduced systolic function and an ejection fraction in the range of 40–45% with diffuse hypokinesis. These changes were consistent with eccentric hypertrophy. Doppler parameters were consistent with abnormal left ventricular relaxation (grade 1 diastolic dysfunction) and a normal peak pulmonary artery pressure with normal right cardiac systolic function. Repeat coronary angiography performed during this admission also showed right coronary artery patency along with the previously detected fistula (Fig. 5). An attempt at coil embolization was unsuccessful due to the inability to advance the microcatheter beyond the fistula. After failed coil embolization, patient was transferred to the CCU in stable condition however 5 days thereafter he suffered a cardiac arrest and eventually passed away.

3. Discussion

Herein we report a unique case of CPAF with coronary steal syndrome and acute on chronic combined systolic and diastolic heart failure in the setting of bronchiectasis. This case helps us understand the unusual hemodynamics involved with CPAF, and provide a platform to further understand etiology and pathogenesis in this particular patient. To better understand CPAFs in general it is important to review the embryology. During embryogenesis, coronary arteries are formed from an island of cells around the heart which later develops into the primary capillary plexus. These cells, particularly the ones found around the aorta, penetrate the aorta and form the coronary ostia [5]. This occurs a few weeks after the division of the truncus arteriosus into the aorta and pulmonary artery; therefore, it is likely that penetration of the primary capillary plexus into the pulmonary artery may lead to anomalous origin of the left coronary artery from the pulmonary trunk. However, in our patient, the coronary ostia were in the normal aortic position but there was an abnormal connection between the coronary artery and one of the branches of the right pulmonary artery. The exact mechanism of this fistula formation remains unknown. However, a popular theory was proposed in 1955 by Hackensellner, in which the coronary fistula was suggested to be due to persistence of more than 2 out of 6 coronary analgens, 3 of which arise from the aorta and 3 from the pulmonary artery [6].

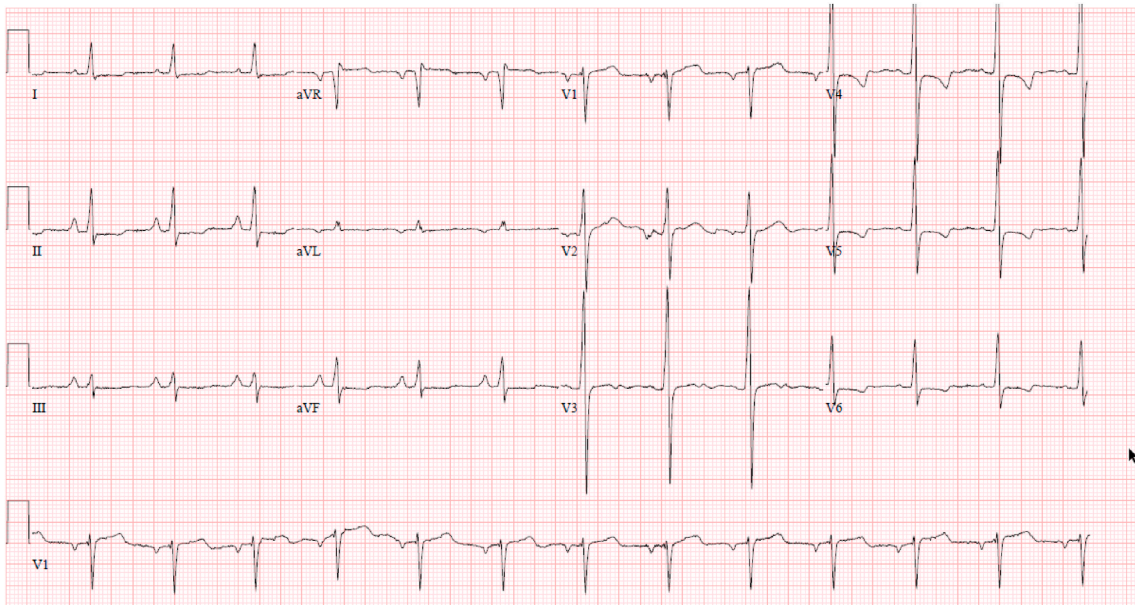CPAF may be explained anatomically by the proximity of the right

Fig. 1. EKG on arrival in the ED, showing ischemic changes (T-wave inversion) in the inferior leads (II, III, aVF).
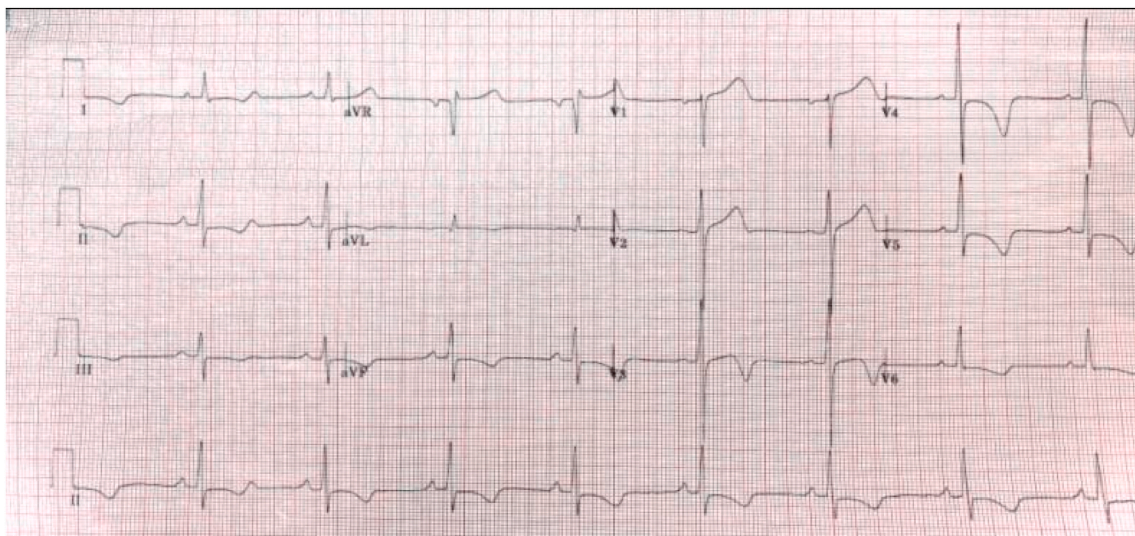

Fig. 2. EKG after normalization of blood pressure, showing ischemic changes (T-wave inversion) in the inferior leads.
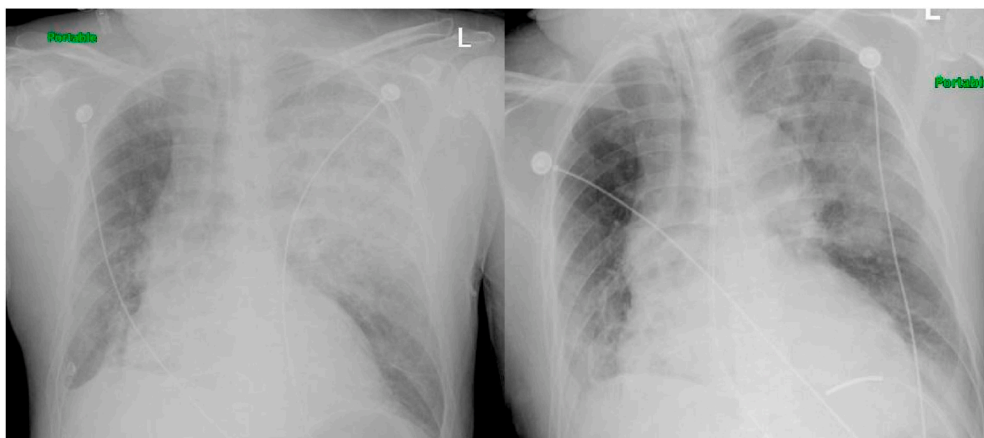

Fig. 3. Chest x-ray showing improvement in pulmonary congestion after diuresis.

Fig. 4. CT angiogram of the chest showing the dilated pulmonary artery.
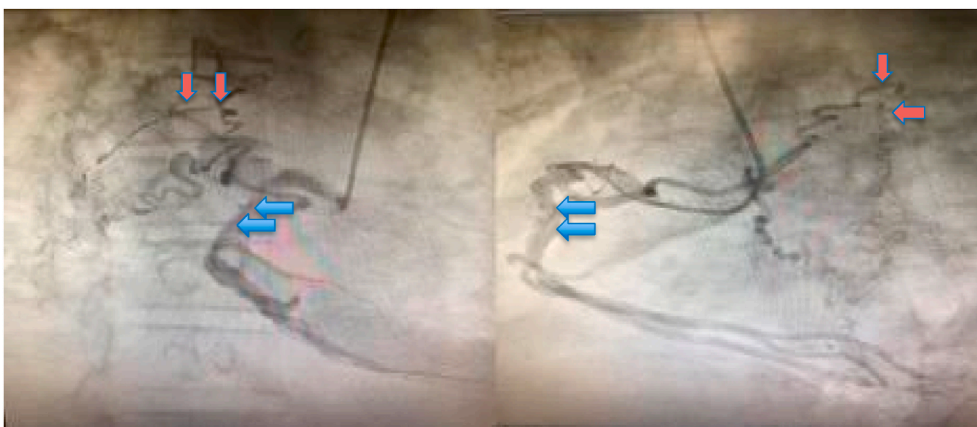

Fig. 5. Coronary angiogram of the right coronary system in the right anterior oblique cranial view (left) and the left anterior oblique cranial view (right) showing the normal right coronary artery (blue arrows) and the right coronary-to-pulmonary artery fistula (red arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

coronary artery to the pulmonary trunk as well as to the right and left pulmonary arteries. Earlier studies have found that fistula termination was found in the pulmonary artery in up to 15–20% of cases, with the most common form being coronary-cameral fistula [7], where it is terminated either in the right atrium or ventricle. Among CPAF cases, 92% terminate in the pulmonary trunk, whereas the rest are divided between the right and left pulmonary arteries. When coronary artery origin is considered, fistulas arising from the left coronary artery are found to be more common (89%) when compared with the right coronary artery [8,9]. This may be because the left coronary artery has a longer course that runs behind the pulmonary artery, thus increasing the chances of developing a fistula.

However, as revealed in this case by coronary angiography, it is difficult to explain the formation of the congenital fistula between the right lower lobe branch of the right pulmonary artery and the proximal right coronary which makes it unique. Acquired coronary fistulas are extremely rare and are typically iatrogenic, but their development has also been described after myocardial infarction or penetrating trauma [10]. Our case is interesting because the fistula corresponded to the area of bronchiectasis in the adjacent lung (Fig. 6). A few reports of fistulas between the coronary artery and both the bronchial and pulmonary vasculatures have been reported but it is unclear if this is due to the formation of new connections via angiogenesis that are secondary to the intense recurrent inflammation associated with bronchiectasis or due to
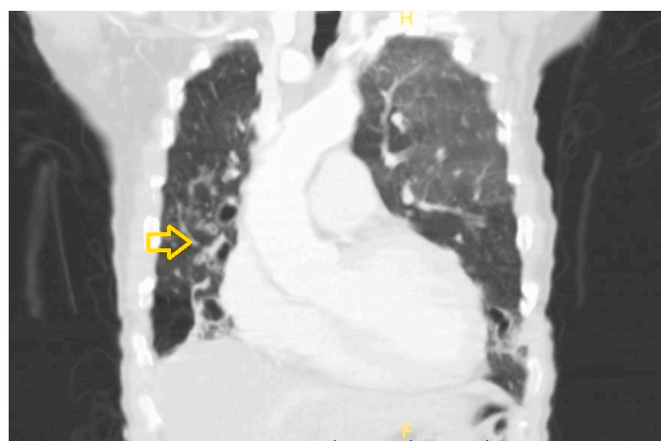

Fig. 6. CT scan of the chest showing bronchiectasis of the lung adjacent to the right side of the heart where the fistula is located.

the opening of a pre-existing dormant fistula as a result of the vasodilation caused by the local inflammation [11–13].

Another important aspect that this case demonstrates is the significance of hemodynamic abnormalities caused by CAFs. Cardiopulmonary

hemodynamics in the setting of CAFs mainly depends on the size of the fistula and also the pressure gradient between its origin and termination. The difference in the high-pressure coronary artery and the low-pressure pulmonary artery eventually results in a left-to-right shunt and leads to pulmonary hypertension. However, in this case, we believe that the pulmonary hypertension was secondary to bronchiectasis, which is common. Uncontrolled hypertension was an added complication, which led to left ventricular hypertrophy, as shown by EKG and eccentric hypertrophy on TTE. When the patient received diuretics and nitroglycerin for the pulmonary edema that developed secondary to the hypertensive crisis, the venous return to the right side of the heart was compromised, which led to low pressure in the pulmonary artery and created a high gradient between the coronary and pulmonary arteries. This gradient diverted the blood from the coronary artery to the pulmonary artery, leading to coronary steal phenomenon. This was demonstrated by the T-wave inversion shown in the EKG, which over a period of time led to systolic heart failure due to the dominant right circulation.

Angelini et al. [14] reported that coronary fistulas are considered significant if: 1. Significant flow is demonstrated in the fistula; 2. There is angiographic evidence of the receiving structure; 3. There are signs of volume overload in the cardiac chambers; 4. There is evidence of coronary steal phenomenon. The ratio of blood flow between the nutrient vessel and fistula will determine the significance of the fistula [15]. The presence of hypertension is well tolerated, but when there is an episode of relative hypotension, as seen in our patient, the flow is diverted into the fistulous tract, resulting in low flow to the distal nutrient vessel and leading to distal myocardial ischemia. In our patient, there was evidence of coronary steal, as shown by the EKG changes, as well as evidence of ongoing ischemia, as indicated by the elevated troponin levels. The absence of significant occlusion in the distal nutrient artery also suggested that the fistula was the cause of the ischemia.

The majority of coronary artery fistulas are asymptomatic and hemodynamically insignificant and are incidentally detected by CT angiogram studies, Doppler echocardiograms, MDCT, or coronary angiography [16]. Symptoms have been described mostly in the elderly and include chest pain, dyspnea, fatigue, syncope, and palpitations [16, 17]. The presence of symptoms depends on the size of the fistula, with larger fistulas resulting in pulmonary hypertension and coronary steal [18]. Our patient had symptoms of acute on chronic congestive heart failure and developed a myocardial infarction during admission, which we believe was caused by the coronary steal phenomenon because it is a known complication of coronary fistulas. Other complications that have been previously described include aneurysm of the fistula, fistula rupture, thrombosis, and endocarditis.

Treatment of CAF is centered on closure which can be achieved via percutaneous transcatheter embolization (PTE) or surgical ligation. The fistula repair method depends on the complexity of the fistula anatomy as well as the presence of any underlying comorbidities that require intervention [19]. There is no consensus guidelines on a preferred repair method, but it is agreed that symptomatic and larger fistulas must be repaired. A Dutch study reported that cases with proximal fistulas, fistulas with termination away from the normal coronary artery, and older individuals are ideal candidates for PTE [20]. In contrast, those with larger fistulas, multiple fistulas, associated cardiac disease requiring surgical management, failure of PTE, tortuous fistulas, and large aneurysms are ideal candidates for surgical ligation [21,22]. In our case, we attempted PTE for treatment of the fistula, but unfortunately the procedure was unsuccessful due to the inability to enter the distal end of the fistula, which is known to happen in up to 6% of patients [21].

4. Conclusion

Coronary artery fistulas are a relatively rare disorder and are typically discovered incidentally. Most of the cases are of congenital etiology, but acquired etiology has been described. This is a unique case of what we believe is an acquired fistula due to the adjacent lung inflammation secondary to bronchiectasis. Treatment is required when the fistula becomes hemodynamically significant, and the treatment method must be individualized and determined on a case-by-case basis.

Declaration of competing interest

All authors have no conflict of financial interest to disclose.

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The authors have no conflicts of interest to disclose regarding this case report and review.

Acknowledgements

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