Multiple Coronary to Bronchial Artery Fistulas Detected During Workup of a Young Patient with Cystic Fibrosis and Bronchiectasis for Lung Transplant

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Abstract

We report a rare case of multiple coronary bronchial artery fistulas in a 34-year-old young male with cystic fibrosis and bronchiectasis while he was actively being worked up for a lung transplant. He had atypical chest pain with compromised left ventricular ejection fraction on echocardiogram. Patients with longstanding bronchiectasis can have acquired anomalous connections from coronary arteries and have to be excluded before lung transplant or any major surgery, especially if they have cardiac symptoms or preliminary investigations are abnormal.

Learning Objective: Our case report highlights that coronary artery bronchial artery fistulas are common in patients with longstanding bronchiectasis and need to be excluded before considering for any major surgical procedure either by MDCT or conventional coronary angiogram.

Keywords: Coronary Bronchial Artery Fistulas; Bronchiectasis; Cystic Fibrosis

Introduction

Coronary bronchial artery fistulas (CBAF) are a rare disorder and altogether less than 30 cases are described in published literature [1]. The etiology of these fistulas is unclear and both lung and cardiac conditions have been described, but by far the commonest cause is bronchiectasis [2]. We present a rare case of multiple CBAF in a young male with a diagnosis of cystic fibrosis while he was worked up for a lung transplant.

Case Report

He is a 34-year-old young male with an early childhood diagnosis of cystic fibrosis and having recurrent respiratory tract infections due to bilateral severe bronchiectasis. Of late his functional capacity has been deteriorating to an extent that he is being considered for a lung transplant. He also has pancreatic insufficiency and on long-term enzyme supplements. Surface ECG was unremarkable and an echocardiogram showed normal chamber sizes but for unknown reasons moderately impaired left ventricular function with calculated ejection fraction of 36% and normal right-sided study. He is a non-smoker, occasionally consumes alcohol and has a family history of coronary artery disease. Stress myocardial perfusion scan (MPS) showed fixed perfusion defect inferobasal area and an EF of 46%. The coronary angiogram (CAG) showed normal epicardial arteries, but the right coronary artery (RCA) and left circumflex artery had small branches with aberrant communication to pulmonary circulation. We were not sure of the distal course of these anomalous connections on CAG, hence organized for a contrast multi detector computer tomography (MDCT) study. The scan confirmed an aberrant tiny branch arising

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from the RCA just distal to the origin of the acute marginal branch and communicates with the dilated, tortuous bronchial arteries. Probably this is an explanation of occasional chest pain complaint, which he had mentioned before, and also the reason for positive stress result.

**Figure 1:** Coronary bronchial artery fistula from Left Circumflex artery and RCA by conventional CAG.

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**Discussion**

CBAF is a rare finding and its incidence varies from 0.08% to 0.61% [3]. Bronchiectasis is considered the most common cause of coronary-bronchial fistula [2]. Previously, a cystic fibrosis patient having multiple coronary bronchial artery fistulas in all three coronary arteries has been described [4].

The pulmonary vascular destruction associated with peripheral inflammation causes distal hypoxia and decreases the pulmonary arterial flow. Other causes include Takayasu arteritis; pulmonary thromboembolism; pulmonary artery tumor; chronic bronchitis; Tetralogy of Fallot with pulmonary atresia; hypoplasia of the pulmonary artery; supravalvular aortic stenosis; and pulmonary tuberculosis [1].

MDCT is considered the diagnostic procedure of choice to detect these anomalies and it offers a better delineation and resolution of the distal communications when compared to conventional angiograms [5]. Most cases of coronary-bronchial artery fistula have no clinical implications because they are asymptomatic, whereas some cases can present with chest pain, breathlessness, hemoptysis or a positive stress test [3]. Our case had some atypical chest pain and positive stress result on myocardial perfusion scan. The chest pain and positive stress test results were due to coronary steal phenomenon from shunting of myocardial blood supply to dilated and low resistance bronchial arteries. Multiple coronary bronchial artery fistulas are quite rare and usually seen in patients with bronchiectasis [2,6]. Cystic fibrosis with end stage bronchiectasis and multiple coronary bronchial artery fistulas before a lung transplant pre-operative evaluation in a very young patient is never been described before and highlights the vigilance needed during workup of such cases. Most of

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the patients are conservatively managed and in symptomatic patients they are treated by surgical ligation or percutaneous transcatheter embolization (PTE) using a detachable balloon, microspheres, coils, microcoils, polyvinyl alcohol, or a covered stent [1]. In our case as he is worked up for a lung transplant, he is planned for a surgical ligation during the transplant procedure.

**Conclusions**

Coronary bronchial artery fistulas are common in patients with certain cardiac and pulmonary diseases and have to be excluded if there is any suspicion on clinical evaluation.

**Disclosures**

The authors declare that there is no conflict of interest. We have not received any financial aid in preparation of this manuscript.

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**Bibliography**


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