

Dieulafoy disease of the bronchus involving bilateral arteries

A case report and literature review

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Abstract

Rationale: Dieulafoy disease of the bronchus is a rare vascular deformity. To the best of our knowledge, reports of these involving both lung vasculature are hitherto absent.

Patient concerns: A 67-year-old male was admitted to our department due to agnogenic hemoptysis.

Diagnoses: Bronchoscopy was performed and some smooth, pulsatile nodular lesions were found in the middle and lower lobes. Computed tomography angiography of the bronchial artery confirmed a left bronchial artery arising from the aortic arch at T4 level, and both bronchial arteries were dilated and tortuous.

Interventions: Bronchial artery embolization was performed successfully.

Outcomes: The patient was discharged with no hemoptysis. In addition, patient is under follow-up until today without any further incidents.

Lessons: This case reminds us that Dieulafoy disease of the bronchus could be a potential etiology for unexplained hemoptysis. The clinician should be aware of this disease when bronchoscopy revealed multiple some smooth, pulsatile nodular lesions, thereafter, bronchoscope biopsy should be avoided, as it could lead to fatal hemoptysis.

Abbreviations: CT = computed tomography, INR = international normalized ratio, NBI = narrow band imaging, PT = partial thromboplastin time.

Keywords: bronchoscopy, computed tomography angiography of bronchial artery, Dieulafoy disease of the bronchus, Hemoptysis, vascular deformity

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This study was approved by The Affiliated Hospital of Medical School of Ningbo University Ethics Committee. Furthermore, the patient signed a written informed consent form.

Informed written consent was obtained from the patient for publication of this case report and accompanying images.

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1. Introduction

Dieulafoy disease is a rare vascular anomaly characterized by dilated, tortuous arteries in the submucosa. It was observed to occur commonly in the gastrointestinal tract and was described for the first time by Dieulafoy in 1898. Dieulafoy disease of the bronchus is rarely seen, which could be presented with recurrent and unexplained massive hemoptysis or with asymptomatic. To our knowledge, 29 cases have been reported since it was first reported by Sweets in 1995,^[1] but involving bilateral vascular lesions is hitherto absent. Here, we present the case of a 67-year-old male who was diagnosed to have Dieulafoy disease of the bronchus, computed tomography angiography of the bronchial artery confirmed a left bronchial artery arising from the aortic arch at T4 level, and both bronchial arteries were dilated and tortuous.

2. Case report

A 67-year-old nonsmoking man was admitted with productive cough and hemoptysis. He had been hospitalized 8 months earlier because of the same symptom and coughed up approximately 10 ml of fresh blood. After cessation of bleeding, there was no hemoptysis during the period before this hospitalization. One day before the current admission to our hospital, he developed recurrent hemoptysis and coughed up fresh blood (about 100 ml) without any clots.

Physical examination of the head, neck, chest, and abdomen was within normal limits. Laboratory tests including white blood cell count, hemoglobin level, platelet count, hematocrit, prothrombin time, partial thromboplastin time (PT), and international normalized ratio (INR) did not reveal any obvious abnormal findings.

Computed tomography angiography showed a left bronchial artery arising from the aortic arch at T4 level,

from which a branch supplied the pulmonary circulation, and both bronchial arteries were dilated and tortuous (Fig. 1A, B).

Bronchoscopy was performed and some smooth, pulsatile nodular lesions were found in the middle and lower lobes; 2 cone-shaped endobronchial protrusions covered by a smooth mucosa were noticed in the right upper lobe (Fig. 1C, D), and fluorescent staining revealed green fluorescence (Fig. 1E). We did not



Figure 1. (A, B) CT angiography shows a left bronchial artery arising from the aortic arch at T4 level, from which a branch supplied the pulmonary circulation and the bronchial arteries on both sides were dilated and tortuous; (C, D) Bronchoscopy of the right lung shows 2 smooth and pulsatile nodular lesions located at the opening of the apical and posterior segments of the superior lobe, and fluorescent staining revealed green fluorescence (E); (F, G) Angiography of the bronchial arteries shows that both arteries were dilated, tortuous, and deformed.

perform a biopsy because we suspected that vascular lesions were present in the bronchial tree.

Then, bronchial angiography was performed and it showed bilateral bronchial arteries were dilated and tortuous (Fig. 1F, G). Then, bronchial artery embolization was performed successfully. The patient was discharged with no hemoptysis. In addition, patient is under follow-up until today without any further incidents. Informed consent was obtained from the patient for publication of this case report and accompanying images.

3. Discussion

Massive hemoptysis is a common critical disease in respiratory medicine. There are many causes of hemoptysis, such as bronchiectasis, tuberculosis, or neoplasm. Dieulafoy disease of the bronchus is a rare clinical entity.

Dieulafoy disease is a rare vascular anomaly characterized by dilated, tortuous arteries in the submucosa.^[2] It appears to occur throughout the gastrointestinal tract. Since Dieulafoy disease of the bronchus was first reported by Sweerts et al in 1995,^[1] multiple similar cases have been reported in the literature.^[1–23] However, the pathogenesis of this vascular anomaly is unclear.^[3] Herein, we present a case of Dieulafoy disease of the bronchus and then we identified 23 published reports with a total of 29 cases and summarized the clinical features of all the cases in Table 1.

Studies show that this condition is associated with bronchial pulmonary congenital dysplasia, chronic airway inflammation, or injury.^[4] Most patients had no history of smoking (21/30) or existing respiratory diseases (25/30). Dieulafoy disease of the bronchus should be suspected if a patient is suffering from recurrent and unexplained episodes of hemoptysis. However, the clinical manifestations are non-typical, comprising productive cough, chest pain, recurrent pneumonia, or exacerbation of bronchiectasis.

In most cases, only inflammatory change was observed, with atelectasis or a non-significant abnormality on routine imaging examination, either on a chest X-ray image or chest CT scan. Only 3 cases presented with an intraluminal nodular lesion and 2 cases showed consolidation on chest CT scan. However, CT angiography is a useful diagnostic tool to detect and locate the origin of abnormal arteries and bleeding.^[5] In our study, we found that CT angiography have been performed on only 6 cases of 30 patients which revealed bronchial artery malformation. The examination revealed the presence of a dilated, tortuous bronchial artery and bronchial-pulmonary artery fistula. According to the literature, the dysplastic artery appears in the right (23/30) lung more than it does in the left side (5/30), and bilateral vascular lesions are rarer (1/30).^[1] The incidence of lesions seems higher on the right side, but in our case, the lesions were bilateral vascular involvement. According to the observations from the pathological examination of dissection specimens, these dilated or deformed arteries pass the bronchial wall and travel through the submucosa.^[4] Besides, abnormal vessels arise more from the systemic than the pulmonary circulation. At present, bronchial arteriography is a useful method for making a definite diagnosis.

Through bronchoscopy, mucosal abnormalities are expressed as small (about 2–7 mm in diameter), non-pulsatile, and smooth nodular lesions with a white cap (18/30), with active hemorrhage (8/30) and blood clot in the bronchus (5/30). Nevertheless, the diagnosis depends on pathological examination; however, biopsy should be considered carefully because of the risk of fatal

Table 1

Clinical manifestations of Dieulafoy disease of the bronchus^{*[1–23]}

Characteristics	Number	Total (n, %) 30
Gender		
Male		19 (63.3)
Female		11 (36.7)
Age (yr)		13–78
Smoking		
Yes		8 (26.7)
No		21 (70)
Unknown		1 (3.3)
Previous respiratory disease		
Yes		5 (16.7)
No		25 (83.3)
Symptoms (Hemoptysis)		
Yes		27 (90)
Massive hemoptysis		17 (56.7)
No		3 (10)
Chest radiograph		
Inflammatory change		12 (40)
Atelectasis		4 (13.3)
Consolidation		2 (6.7)
Bronchiectasis		2 (6.7)
Intraluminal nodular lesion		3 (10)
Negative		7 (23.3)
Unknown		5 (16.7)
Bronchoscopy		
Nodular lesion		18 (60)
Tortuous and nonpulsatile vessel		1 (3.3)
Active hemorrhage		8 (26.7)
Blood clot		5 (16.7)
Negative		1 (3.3)
Origin of vessels		
BA		18 (60)
PA		3 (10)
Mediastinal vessel		1 (3.3)
Unknow		8 (26.7)
Biopsy		
Total		8 (26.7)
Massive hemorrhage		6 (20)
Death		4 (13.3)
Location		
L		5 (16.7)
R		23 (76.7)
Trachea carina		1 (3.3)
L and R		1 (3.3)
Treatment and outcomes		
Embolization success		10 (33.3)
Embolization failure		8 (26.7)
Surgery success		15 (50)
Surgery failure		1 (3.3)

* BA=bronchial artery, L and R=left lung and right lung, L=left lung, PA=pulmonary artery, R=right lung.

hemorrhage.^[6] Among the 30 cases in this study, 8 patients underwent a biopsy, 6 cases presented massive hemorrhage and 4 patients died. Therefore, we do not recommend bronchoscopic biopsy once the disease is suspected.

When bronchoscopy is performed in a patient presenting with hemoptysis, it is important to identify the characteristics of the intraluminal nodular lesions, thus reducing the risk of fatal hemorrhage. With the use of a conventional white light bronchoscope alone, up to 29% of the vascular anomalies and dysplastic mucosal and dysplastic mucosal lesions may not be detected.^[24,26] Narrow band imaging (NBI) can aid in visualizing

the superficial mucosa to detect vascular lesions by using 2 different ranges of wavelengths: 390 to 445 nm and 530 to 550 nm.^[26] Vascular lesions in the submucosa appear blue-green under NBI; the color blue indicates that the vessel is at a shallow location, while green indicates that the location is at a deeper location. Endobronchial ultrasound scan is also a useful tool to detect vascular abnormalities which are usually hypoechoic or anechoic with hyperechoic or echogenic walls.^[24,25] Combining these 2 technologies can help to define the property of intraluminal nodular lesions.

Up till now, selective bronchial artery embolization has been an effective and safe procedure for controlling hemoptysis. This procedure offers an alternative method for patients who cannot undergo lobectomy owing to compromised general health status or simply do not want to undergo surgery and demonstrates a good curative effect (10/18). Despite this, 8 cases presented with recurrent hemoptysis, which may have been associated with malformed vessels originating from the pulmonary circulation rather than systemic circulation. In addition, revascularization, neovascularization, and drop of the embolus dropping could also have caused failed embolization.^[5]

If the malformed vessels are originating from the pulmonary circulation, embolization is often ineffective, and lobectomy can be an alternative option for cure. Lobectomy should be considered when the patient develops massive hemoptysis or embolization is unsatisfactory.

To the best of our knowledge, reports of these involving both lung vascular are hitherto absent. We present the first case report of recurrent hemoptysis resulting from dilated and tortuous bilateral bronchial arteries causing Dieulafoy disease. Our patient had a short-term history of slight hemoptysis. Moreover, the time of his first episode compared to others was relatively late. The bronchial angiography and CT angiography revealed the same features, that is, dilated and tortuous bilateral bronchial arteries; however, this is an infrequent situation within clinical practice. Then, the patient showed satisfactory curative effect through bronchial artery embolization.

In summary, this case we reported and those reviewed support that Dieulafoy disease of the bronchus should be considered if a patient presents with recurrent and unexplained hemoptysis. Combining narrow band imaging and endobronchial ultrasound scan can help to identify the characteristics of an intraluminal nodular lesions before biopsy, thus reducing the risk of fatal hemorrhage. Bronchial arteriography and selective bronchial artery embolization should be performed in time to reduce the risk of life threatening hemoptysis. Lobectomy could be considered as a radical intervention.

Author contributions

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