

Dieulafoy disease of the trachea with recurrent episodes of massive hemoptysis

A case report

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Abstract

Rationale: Dieulafoy disease is characterized by the presence of dilated, tortuous arteries that project into the submucosa of the gastrointestinal tract and less frequently the bronchus.

Patient concerns: A 60-year-old woman with recurrent episodes of massive hemoptysis.

Diagnoses: Dieulafoy disease of the trachea.

Interventions: Selective arterial embolization was undertaken.

Outcomes: The intervention was successful and no fresh episode of acute hemoptysis was observed.

Lessons: Apart from the bronchus, vascular anomaly may also be present in the trachea in Dieulafoy disease.

Abbreviation: CT = computed tomography.

Keywords: arterial embolization, case report, computed tomography angiography, Dieulafoy disease, trachea

1. Introduction

Dieulafoy disease is noted for a vascular anomaly characterized by the presence of dilated, tortuous arteries that project into the submucosa of the gastrointestinal tract.^[1] Dieulafoy disease of the bronchus was subsequently reported in patients with unexplained massive hemoptysis and hitherto a total of 47 cases of Dieulafoy disease of the bronchus have been described.^[2] Dieulafoy disease of the bronchus is considered a congenital anomaly and it is speculated that the disease arises from a failure of caliber-persistent artery running within the submucosa to differentiate into capillaries.^[3] The dilated tortuous arteries in the submucosa originate from the bronchial artery and pulmonary artery and are vulnerable to bleeding.

Dieulafoy disease of the trachea has not been previously described. Herein, we report the first case of Dieulafoy disease of the trachea in a 60-year-old woman with recurrent episodes of massive hemoptysis. The condition was diagnosed by bronchoscopy and computed tomography (CT) angiography and successfully managed by selective arterial embolization.

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The authors report no conflicts of interest.

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2. Case report

A 60-year-old woman patient was admitted to our hospital on Feb. 18, 2016 because of frequent episodes of hemoptysis for 2 weeks. In the previous decade, the patient had frequent but less severe episodes of hemoptysis, which typically ensued following an upper respiratory tract infection. Ear, nose, and throat examination and laryngoscope failed to identify an apparent source of bleeding. CT chest scan revealed no abnormality. The most recent episode occurred after an upper respiratory tract infection 2 weeks ago. The patient coughed up as much as about 500 mL of fresh blood in an episode of hemoptysis, which was alleviated by anti-tussive therapy. The patient denied a history of smoking and illicit drug use. Diagnostic workup revealed no evidence of coagulopathy. Upon admission, physical examination revealed signs of rhonchi and reduced breath sounds. Bronchoscopy showed a 1-cm lesion at the membranous trachea 2 cm to the carina. Tortuous blood vessels were observed running in the submucosa of the trachea (Fig. 1). CT angiography was performed, demonstrating an artery extending into the submucosa from the descending aorta (Fig. 2). A diagnosis of Dieulafoy disease of the trachea was entertained. Since the blood vessel was considered to be the culprit of hemoptysis, selective arterial embolization was performed 1 week later. No fresh episode of acute hemoptysis was observed and the patient was still being followed up at the time of writing this report.

This study was approved by Ethics Committee of Shanghai Jiao Tong University Affiliated Sixth People's Hospital, and also got an informed written consent from the patient.

3. Discussion

Dieulafoy disease of the bronchus was first reported by Sweerts et al^[2] in 1995 in 2 patients with unexplained massive hemoptysis, approximately 100 years after the first description of the condition in the stomach by the French surgeon Georges Dieulafoy in 1898 following his study of fatal gastric

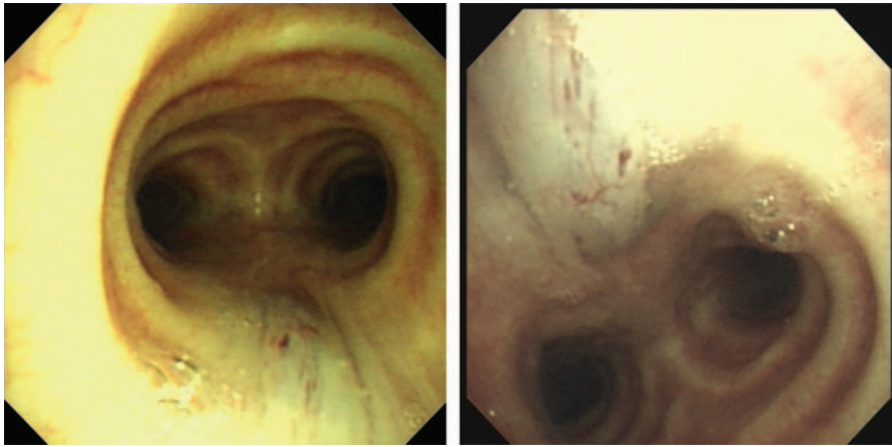


Figure 1. Bronchoscopy in a 60-year-old woman patient with Dieulafoy disease shows a 1-cm lesion at the membranous trachea 2 cm to the carina. Tortuous blood vessels are observed running in the submucosa of the trachea.

haemorrhage in 3 asymptomatic young men.^[4] Dieulafoy disease of the bronchus, though considered rare, may have higher occurrences because the condition is underappreciated and underdiagnosed.^[5] Over the past decade, Dieulafoy disease of the bronchus has been increasingly recognized as a cause of pulmonary hemorrhage. Parrot et al^[6] reported a series of 7 patients with Dieulafoy disease of the bronchus. These patients were all current heavy smokers and experienced massive and unexplained episodes of recurrent hemoptysis. Yang et al^[7] reported 3 cases of bronchial Dieulafoy disease and reviewed 19 additional cases from the literature. These patients typically developed sudden onset of massive hemoptysis.

Dieulafoy disease of the bronchus remains a difficult disease to detect to the small size of the nodular lesion within the lumen of the bronchus, which may be covered with coagulum. Our patient had a 10-year history of recurrent massive hemoptysis without a

definite diagnosis. Bronchoscopy demonstrated tortuous blood vessels running in the submucosa of the trachea. Dieulafoy disease of the trachea was further established by vascular imaging with CT angiography. We did not perform bronchoscopic biopsy as it entails the risk of triggering sudden massive hemoptysis. van der Werf et al^[5] reported that bronchoscopic biopsy triggered fatal massive bleeding in a 70-year-old woman with Dieulafoy disease of the bronchus. Literature reviewed by Yang et al^[7] also showed that biopsy injury often caused massive or lethal hemoptysis.

The anomalous arteries in Dieulafoy disease of the bronchus may originate from the bronchial artery or less commonly from the pulmonary artery. Yang et al^[7] reported that in those patients with a known source of the anomalous artery, 82% (9/11) were from the bronchial artery and 18% (1/11) were from the pulmonary artery while in half of the cases (11/22) the originating artery could not be determined. In another review of 17 cases of Dieulafoy disease of the bronchus, the authors also found that in those patients with a known source of the anomalous artery, 75% (9/12) were from the bronchial artery and 25% (3/12) were from the pulmonary artery. Our CT angiography demonstrated that the anomalous artery originated from the descending aorta. Selective bronchial artery embolization is recommended of the culprit vessel in Dieulafoy disease of the bronchus. We also carried out successful selective arterial embolization and to date no recurrent episode of hemoptysis has been observed in the patient.

In conclusion, our current case suggests that Dieulafoy disease may also occur in the trachea. Dieulafoy disease of the respiratory tree should be considered in patients with recurrent massive hemoptysis. Vascular imaging by CT angiography may be used to diagnose the condition and selective arterial embolization may be promptly instituted to avoid lethal massive hemoptysis.



Figure 2. Computed tomography (CT) angiography demonstrates an artery extending into the submucosa from the descending aorta. Arrowhead indicates the submucosa vessel.

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