To the Editor: Dieulafoy’s disease is characterized by the tortuous arteries of abnormally large caliber running within the submucosa, which is susceptible to ulceration and bleeding. This vascular abnormality usually occurs in the gastrointestinal tract. The involvement in the respiratory system is rare. Massive hemoptysis caused by Dieulafoy’s disease of the bronchus is a life-threatening condition. An immediate and effective treatment is required in emergency to improve survival. Here, we report a patient who had such a vascular anomaly in bronchus and presented with recurrent massive hemoptysis. The patient signed information consent form.

A 21-year-old nonsmoking female was transferred to our clinical center from local hospital with massive hemoptysis. There was no medical history in this patient. Before admission, she had two severe episodes without any premonitory symptoms, approximately 300–400 ml blood for each time. General physical examinations and laboratory tests were unremarkable except for moderate anemia. Chest computed tomography (CT) scan showed ground-glass opacity due to aspiration of the blood in the right lower lobe, and CT angiography (CTA) revealed convoluted vessels originated from the thoracic aortic [Figure 1a and 1b]. Under the bronchoscope, there were some bleeding from the right lower lobe bronchus and two apophyses of about 3–5 mm in diameter were found at the entrance to the right lower lobe bronchus. It arose from the surface with apparently normal mucosa [Figure 1c]. Because of suspicion of vascular malformation of the bronchus, the bronchoscopist was refrained from performing any biopsy. Selective bronchial arteriography was consequently performed and it showed tortuous, dilated, and elongated branches of the bronchial artery in the region of the right lower lobe bronchus [Figure 1d]. Following the bronchial arteriography, embolization of the feeding artery of the angiomma-like vascular malformation of the right bronchial artery was performed using three 3-mm coils to control the hemoptysis [Figure 1e]. We rechecked bronchoscopy in 3 days later after the embolization. We observed two mucosal apophysis lesions at the entrance to the right lower lobe bronchus had almost disappeared, and there were only less obsolete bloodstains in the bronchial lumen [Figure 1f]. Hemoptysis did not recur in the follow-up period of 13 months after discharge.

Georges Dieulafoy first depicted a tortuous and dilated submucosal artery as a gastrointestinal bleeding source in 1898. Since then, this kind of vascular malformation was described as Dieulafoy’s disease. The bronchial variant of Dieulafoy’s disease was initially reported in 1995, and it has been considered to be a rare disease.[1] The causes of Dieulafoy’s disease of the bronchus remained uncertain. Some scholars believed that it might be a congenital disorder.[2] Culprit vessel of Dieulafoy’s disease usually locates on the right lobe or segment bronchus, and its origin could be heterogeneous. Most clinicians considered the vessel as originating from the systemic vasculature, while some others identified the vessel belonged to the pulmonary vasculature because of the failure of bronchial artery embolization (BAE).[3,4] Dieulafoy’s disease can be asymptomatic or occasionally present with spontaneous massive hemoptysis, just like this patient. The culprit vascular often surrounds the bronchus and runs within the bronchial submucosa. This convoluted and dilated vascular leads to a pressure corrosion of the bronchial mucosa with consequent rupture of the vascular wall, and it may eventually cause severe hemorrhage.[3] The bronchoscopists usually described it as small mucosa-colored tumor-like lesions with a few millimeters in diameter or length, which have a benign appearance under bronchoscope. In this case, we found two apophysis lesions of about 3–5 mm in diameter at the beginning of the right lower lobe bronchus during bronchoscopy. It arose from the surface with normal mucosa. This appearance of the lesion was consistent with prior reports. We found that these two mucosal apophysis lesions had almost disappeared after BAE, and we thought that this interesting finding further supports the diagnosis of Dieulafoy’s disease. Nevertheless, conventional bronchoscopy is actually of limited value in the diagnosis of Dieulafoy’s disease because the bronchoscopic findings are nonspecific. Some studies showed that radial endobronchial ultrasound may be conducive to exploring the vascular nature of the lesion, but utility was also limited by the location of the lesion and the size of ultrasound probe.[5] It should

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be noted that the endoscopic recognition is critical to prevent iatrogenic hemorrhage. The iatrogenic life-threatening bleeding often occurs immediately after the biopsy, and it could also happen after an interval of several days. A bronchoscopist must pay extra attention in case of hemoptysis and avoid biopsy when Dieulafoy’s lesion is suspected.

Selective bronchial arteriography and CTA can reveal dilated bronchial arteries, abnormal arteries, vascular shunts, fistulae, and extravasation of contrast into the lungs, which can be helpful in making the diagnosis and ruling out other causes of hemoptysis. If lesions of the bronchial arteries fail to show up, pulmonary arteriography would be essential.

At present, there is no consensus on treatment of bronchial Dieulafoy’s disease due to its rarity. BAE is probably the initial treatment choice although it is not always successful. Failure of BAE may be owing to a deformed vessel originating from pulmonary circulation, and the bronchopulmonary shunts may also impair success rate of BAE. In case BAE is unavailable or unsuccessful, surgery can be a reasonable treatment option. It was reported that surgery alone or after failure of BAE had a success rate of nearly 100%. Besides conventional management options, some reports described therapeutic bronchoscopic management including argon plasma coagulation (APC) and Nd:YAP laser ablation for this condition. The advantages of endobronchial APC and laser ablation are rapid, effective coagulation, and short operating time.

Some clinicians believe that Dieulafoy’s disease of the bronchus is more frequent than thought. Savale et al. described that bronchus Dieulafoy’s disease accounted for at least 6% of the patients undergoing surgery for hemoptysis overall and up to 55% of the patients undergoing surgery for hemoptysis presumed to be cryptogenic. Therefore, this diagnostic option should be considered when the patient has recurrent massive hemoptysis which cannot be explained otherwise. Selective embolization of the feeding bronchial artery or resection of the affected pulmonary segment should be performed without delay to avoid fatal outcomes.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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