Dieulafoy's disease of the bronchus: rare but potentially fatal

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Case Report

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Abstract

Background: Dieulafoy's disease of the bronchus can cause massive and even fatal hemoptysis. Even though it is rare, it should be considered by physicians all over the world, as several cases are reported by literature.

Methods and results: We report the case of a 41-year-old man, so far in good health, presenting with massive hemoptysis. Bronchoscopy showed blood clots and a protruding lesion covered by mucosa with a white pointed cap at the entrance of the right upper lobe. Biopsies were not attempted. Embolization of bronchial artery was first realized and was not successful, with post procedure complications. Surgical intervention stopped the bleeding and pathological examination of the resected specimen confirmed Dieulafoy's disease of the bronchus.

Conclusion: To our knowledge, this is the first case of bronchial Dieulafoy's disease to be reported in Tunisia and North Africa. When the diagnosis is suspected, bronchoscopy biopsy should be avoided as it might lead to fatal hemorrhage. Selective bronchial artery embolization can stop the bleeding, but surgery can be required.

Background:

Massive hemoptysis is a medical emergency which is still feared by most physicians. It presents several diagnostic and therapeutic challenges. Determining the origin of bleeding and underlying etiology is a cornerstone of the treatment plan. However, it may not be immediately apparent and a thorough investigation must be lead. We present the case of a young patient suffering from massive hemoptysis due to bronchial Dieulafoy's disease.

Case Presentation:

A 41-year-old man was admitted in February 2022 to the pneumology department with sudden onset of massive hemoptysis.

He had had a less severe episode of hemoptysis one year ago, concomitant to dental extraction, but was otherwise healthy. He worked as a university professor and was an occasional smoker. Initial clinical examination was normal, aside from sinus tachycardia. Blood biochemistry parameters showed a decrease in hemoglobin level from 14 g/dl to 9 g/dl, indicating blood transfusion.

Bronchoscopy showed bleeding stigma in the right main bronchi and a protruding lesion at the entrance to the right upper lobe. The surface was covered by mucosa and had a white pointed cap (Fig. 1). Blood clots were also noted in the right inferior lobe bronchus. Computed tomography (CT) scans revealed ground glass opacities at the upper, middle and inferior lobes of the right lung (Fig. 2). The right bronchial artery had an ectopic origin from the aortic arche. Further workup with bronchial arteriography revealed no tortuous arteries nor any vascular blush. However, the patient was still coughing up important

amounts of fresh blood, approximately 300 ml in an episode, despite prescribing systemic hemostatic treatment.

All of the investigations performed found no evidence of any etiology and vasculitic screen was negative.

Given the findings of the bronchoscopy, we performed an arterial embolization of the right bronchial artery and the 5th right intercostal artery, but this failed to prevent the recurrence of his bleeding. The patient developed complications after the procedure. He suffered from splenic infarction, bilateral renal infraction and a posterior inferior cerebellar artery stroke. The patient also developed acute respiratory failure due to bilateral pulmonary embolism, proximal on the left side. Ultimately, he required hemostasis surgery.

He had a right pneumonectomy on veno-venous extracorporeal membrane oxygenation with simple surgical follow up. Curative heparin was prescribed to treat his pulmonary embolism.

The pathological examination of the resected specimen had found abnormally dilated, sinuous and anastomotic vessels extending into the bronchial mucosa consistent with the diagnosis of bronchial Dieulafoy's syndrome (Fig. 3).

The postoperative course was uneventful aside from a post traumatic chest wall hematoma. A follow-up 4 months later, the patient was well with no further episodes of hemoptysis.

Discussion:

Dieulafoy's disease has been first reported in 1898 by Georges Dieulafoy usually affecting the digestive tract (1). The bronchial location of this disease has been first reported by Sweerts et al. in 1995 (2). It's an extremely rare affection which may manifest by massive hemoptysis. Over the past decade, cases of the disease have been increasingly outlined. A recent systematic review of the literature published by Qian et al. collected 73 cases from 1995 to 2019 (3). To our knowledge, this case would be the first one published in Tunisia and North Africa.

The cause of the disease is still unknown. Theories vary from congenital vascular malformations to bronchial injury secondary to previous infections (2). Parrot et al. (4) suggested a possible association with inflammatory lesions in tuberculosis or stretching and dilation of the bronchial artery. Advanced age and tobacco smoking have been implicated in the increase of bleeding-related complications (5). However, the disorder may affect people at every age especially middle-aged adults (3), and also non-smokers.

Clinical manifestations are non-specific, but the most common one is recurrent hemoptysis. Massive and even fatal hemoptysis may occur especially while performing bronchoscopy guided biopsy (6). Other symptoms such as a cough, chest pain, infection or respiratory failure can be reported by patients (7).

Establishing the diagnosis of Dieulafoy's disease can be quite difficult. An exhaustive evaluation was lead with chest X-rays, computed tomography (CT) scans, bronchoscopies, biopsies and bronchial angiographies in historical cases.

In Dieulafoy's disease of the bronchus, chest X-rays and computed tomography (CT) scans are rarely contributive to the diagnosis. They show mostly manifestations of an intrapulmonary hemorrhage with ground glass opacities (3). The relevance of this exam is its contribution to excluding other lung diseases causing the bleeding. Endobronchial nodes have been identified by chest CT in some cases (8). Multi-slice CT angiography can show a tortuous and dilated bronchial artery (9). Due to lack of specificity and sensitivity, most authors agree that chest X-rays and CT scans are not the best modality by which to diagnose Dieulafoy's disease.

Bronchoscopy mainly show massive endobronchial hemorrhage, which may be accompanied by blood clot formation. A mucosal protrusion is frequently observed in the site of the bleeding bronchus, which diameter can be only a few millimeters (3, 7). The mucosa covering the protruding surface looks like a "white cap", without a pulsating sensation. The surrounding mucosa can be normal or congested. In most cases, the abnormal lesions were located in the right bronchus (3).

These bronchoscopic findings in Dieulafoy's disease of the bronchus are not diagnostic since the abnormal vessel is usually pinpoint mucosal defect surrounded by normal-looking mucosa. Moreover, a small lesion can be undetected due to pooling of blood or clots within bronchial lumen. Among 74 cases reported by Qian et al. (3), biopsies were attempted for 19 patients whom presented a nodular lesion without a typical vascular lesion. It was primarily suspected to be an endobronchial mass or carcinoid tumor. Seventeen patients had bleeding after the biopsy, and six deaths occurred due to massive hemorrhage. Bronchial biopsies in such diseases entail the risk of triggering fatal hemoptysis. Since 2014, with a better understanding of Dieulafoy's disease, biopsies have been avoided for nodules suspected to be caused by Dieulafoy's disease (10), which has reduced the risks of massive hemorrhage. In 2010, Guiroli et al. (11) have demonstrated the clinical utility of endobronchial ultrasound EBUS) in the evaluation of bronchial alteration suspicious of Dieulafoy's lesion. This technique can be helpful to clarify the nature of the nodular lesion and contributes to the diagnosis, avoiding potentially disastrous interventions (12). The major manifestation is a fluid echo-free zone in the submucosal lesion. The Doppler mode can be used to detect blood flow. However, convex probe EBUS cannot reach the upper lobe bronchus nor segmental bronchus. Radial probe EBUS can be used instead but it has no doppler mode and cannot determine blood flow within the lesion.

Bronchial angiography has a twofold benefit. It has a role in the diagnosis as it can show a rich blood supply to the corresponding site of the lesion; a deformed, tortuous and dilated artery with signs of bleeding (3). When detected, these abnormalities can indicate a selective bronchial embolization, which has an important therapeutic value. However, lesions of the arteries may not be visualized, as it was the case for 2 patients in the systematic review of Qian et al. (3) and for our patient as well. The bronchial arteries usually originate from the proximal descending thoracic aorta. Arteries that originate elsewhere in

the aorta or from other vasculature are termed ectopic (13). Right bronchial arteries occasionally originate from the aorta but more commonly share their origin with another artery, usually an intercostal artery. Choi et al. (14) evaluated in their study the spectrum of variations in bronchial artery and among ectopic origins, concavity of the aortic arch was the most common.

To make a definite diagnosis, many researchers consider that pathological examination of biopsies, surgical or autopsy specimens is required. However, there are no uniform diagnostic criteria and due to risks involved, the need for pathological diagnosis remains controversial. The pathological exam usually shows an arterial malformation in the bronchial submucosa. The tortuous, dilated and deformed artery forms small nodules coated with bronchial mucosa and protruding from the bronchial lumen (4, 15). Diagnosis is confirmed when a dysplastic artery is identified in the bleeding territory without evidence of other underlying lung disease, vasculitic changes or neoplasm.

Treatment options include conservative internal medication, surgical lung resection, selective bronchial artery embolization (SBAE) and bronchoscopic ablation. Conservative treatment and the use of hemostatic agents is rarely efficient in stopping the hemorrhage. Niu al. (16) have reported that pituitrin and thrombin may occasionally have good therapeutic effect in some cases of infantile bronchial Dieulafoy’s disease. Bronchoscopic ablation has been tried in a minority of cases (3) and is not without perils. Mediastinitis, esophageal injuries, and broncho esophageal fistulas are all potential complications (17). Selective bronchial artery embolization is often performed as a first-line treatment and is efficient in most patients (7). But hemoptysis may reoccur after the procedure. The study of Qian et al. (3) revealed that 52.6% patients who underwent SBAE required secondarily lobectomy. One patient had undergone SBAE seven times due to recurrent hemorrhage (18). Up to date, surgery has been the main definitive treatment with a success rate of nearly 100% in all reports (3). Recurrence of hemoptysis in unlikely after resection of the diseased lung lobe.

Conclusions:

Dieulafoy's disease of the bronchus is a rarely reported, and possibly underdiagnosed, cause of life-threatening hemoptysis. It should be included in the differential diagnosis of patients with massive hemoptysis with no other evident etiology. Bronchial angiography and EBUS may be highly suggestive of this disease. While SBAE is a less invasive procedure, surgical treatment remains a lifesaving approach that reduces the probability of recurrence. Therefore, it is the best choice which also allows an accurate histopathological diagnosis.

List Of Abbreviations:

CT: Computed tomography

EBUS: endobronchial ultrasound

SBAE: selective bronchial artery embolization

Declarations:

I confirm that the patient consented to participate and publish their clinical data and images.

Ethical Approval:

We obtained the Hospital Ethical Committee approval

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No competing interests

Authors' contributions:

Daboussi Salsabil: conceived the work, reviewing and finalization of the manuscript

Kacem Marwa: collected clinical details, analysis and interpretation of data,

contriubuted to writing.

Boubaker Nouha: approval of the final version.

Chaabene Mariem: approval of the final version.

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Figures

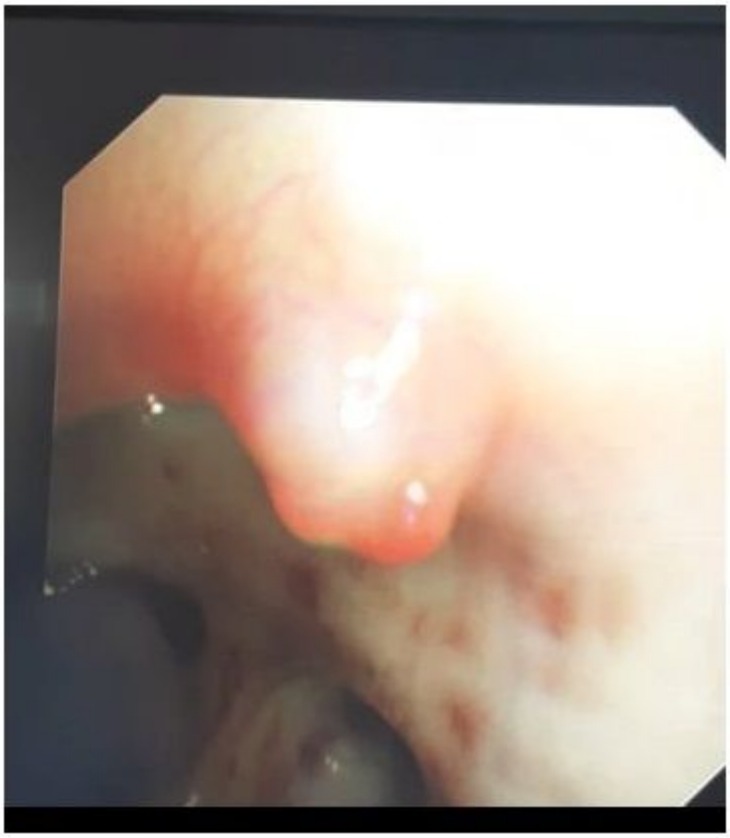

Figure 1

Nodular protrusion from the mucosa at the entrance of the right upper lobe

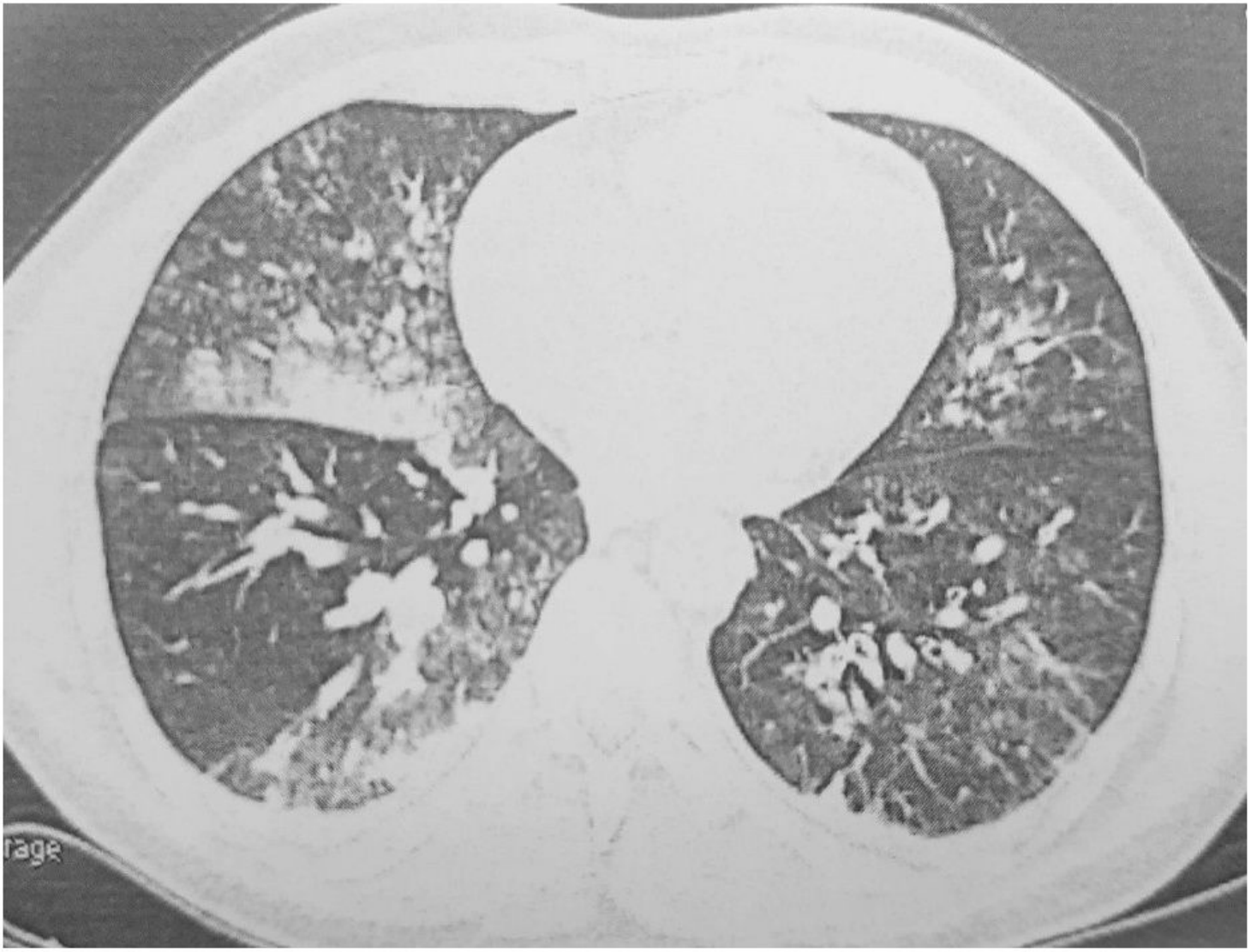

Figure 2

Ground glass opacities of both lungs in computed tomography scans

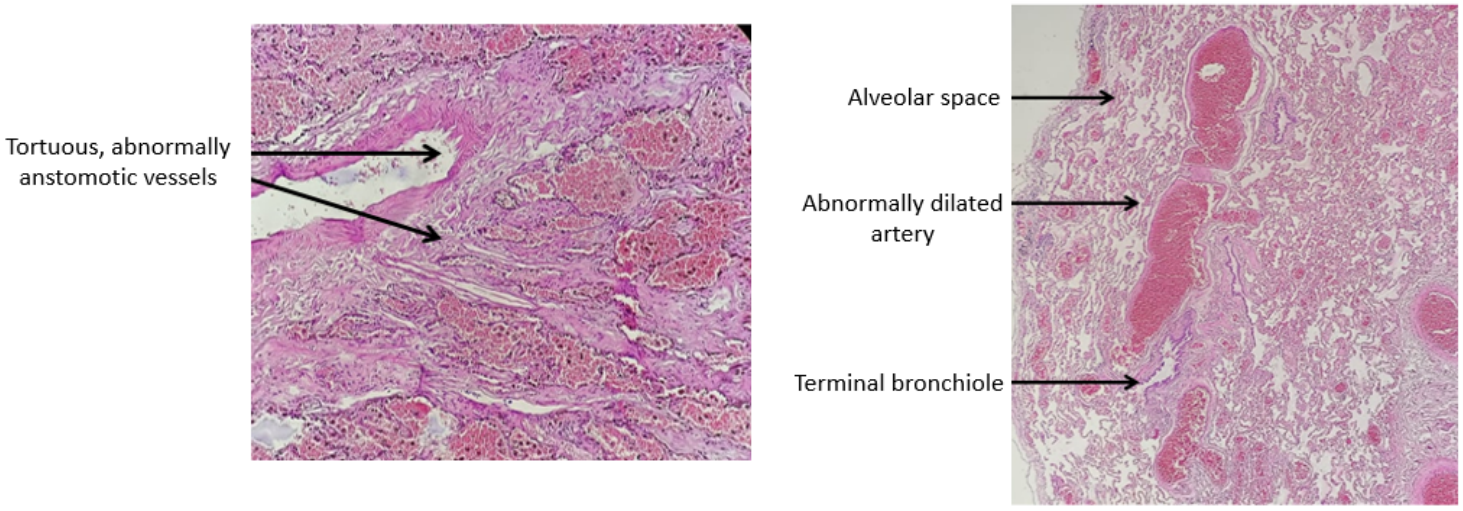

Figure 3

Pathological findings: Abnormally dilated, sinuous and anastomotic vessels extending into the bronchial mucosa