

Dieulafoy's disease of the bronchus: a rare cause of massive hemoptysis

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Keywords

Dieulafoy's disease, hemoptysis.

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Received: 03 November 2013; Accepted: 12 November 2013

Respirology Case Reports 2014; 2(2): 55–56

doi: 10.1002/rccr.2.47

Abstract

We present the case of a 30-year-old non-smoker who presented with unexplained, massive hemoptysis and was diagnosed with a rare vascular malformation.

Introduction

Dieulafoy's disease is a vascular anomaly characterized by the presence of dilated, tortuous arteries that project into the submucosa and have an increased susceptibility to bleeding. Although most frequently seen in the gastrointestinal tract, it has been reported in patients presenting with large-volume hemoptysis [1].

Although the etiology is not well understood, Dieulafoy's disease has been postulated as a congenital abnormality. Advancing age and heavy smoking are believed to increase the risk of bleeding-related complications; however, cases have been reported in young, otherwise well individuals [2]. Dieulafoy's disease of the bronchus is a rarely reported, and possibly underdiagnosed, cause of life-threatening hemoptysis.

Case Report

A 30-year-old male, nonsmoker, with no significant medical history was admitted to a regional hospital following an episode of massive hemoptysis. He was otherwise asymptomatic and had no infectious prodrome. There was no history of precipitating events such as trauma or illicit drug use. His coagulation studies and inflammatory markers were normal

and computed tomography (CT) pulmonary angiography was negative for pulmonary emboli. He had a further five episodes of hemoptysis – each involving greater than 100 mL of fresh blood – before being transferred to our care.

The patient was commenced on empirical antibiotic therapy, tranexamic acid, and steroids but continued to have significant hemoptysis. Ear, nose, and throat (ENT) examination could find no cause for his bleeding.

Vasculitic screen was negative. CT chest showed nonspecific, bibasal alveolar changes. Echocardiogram found normal pulmonary pressures.

At bronchoscopy, the patient was found to have old blood forming almost a complete cast of the right lung. Although no endobronchial lesion could be seen, once the airway was cleared, he was noted to have fresh bleeding from the basal segments of the right lower lobe. Attempts were made to control the bleeding with adrenaline and cold saline at this time.

The patient proceeded to CT thoracic angiogram and pulmonary arteriogram, which were unable to identify the site of bleeding or any vascular malformation. Given the findings at bronchoscopy, particle embolization of the distal right bronchial artery was performed, but this failed to prevent a recurrence of his bleeding and ultimately the patient required a lobectomy.

On formal pathological examination of the right lower lobe, there was evidence of hemorrhage throughout the airways and a large torturous artery extending into the bronchial mucosa.

The postoperative course was uneventful, and at follow-up 22 months later, the patient remained well with no further episodes of hemoptysis.

Discussion

Massive hemoptysis is a serious, life-threatening condition. In 7–25% of cases, an underlying cause is not identified despite extensive investigation.

Since the first reports of this condition in 1995, Dieulafoy's disease of the bronchus has been increasingly recognized as a cause of pulmonary hemorrhage. However, it remains a difficult condition to detect preoperatively and is therefore probably still underdiagnosed.

Identification of the likely source of bleeding (and exclusion of ENT and gastrointestinal tract causes) is key to the management of massive hemoptysis, as this will guide intervention and measures to stabilize the patient. As in the case presented here, gravitational pooling and spillage of blood from one lung to another can lead to misleading physical examination and radiological findings. Therefore, bronchoscopy remains the key diagnostic, and potentially therapeutic, intervention.

Rigid bronchoscopy is usually preferred due to its greater capacity for suction and wider range of interventional capability. Again, blood may have tracked throughout the lungs, and therefore, visualization of active bleeding is necessary to be confident about the territory involved.

If the site of bleeding can be identified or localized to a particular lung segment, attempts to control the hemorrhage can be made with a range of approaches including topical adrenaline, iced saline lavage, or balloon tamponade. Single lung ventilation can also be performed by advancing an endotracheal tube into the contralateral lung. The goal is for the inflated cuff of the endotracheal tube to protect the nonbleeding lung from further spillage of blood and ensure adequate gas exchange.

Mucosal abnormalities – typically small, nonpulsating, benign-looking protuberances – are occasionally seen during bronchoscopy. Despite the need for tissue to make the diagnosis, biopsy should be avoided if Dieulafoy's disease is strongly suspected because of the risk of precipitating hemorrhage [3]. There may be some role for endobronchial ultrasound during bronchoscopy to further evaluate suspicious lesions [4].

Although there are no defined diagnostic criteria, arteriography may occasionally identify a vascular abnormality, which, if clinically correlated, can help to make the diagnosis. Selective arterial embolization is a reasonable first-line treatment, particularly in these cases, but it seems to have modest long-term success and many patients ultimately proceed to surgery.

Diagnosis is confirmed on histological examination of resected lung tissue, when a dysplastic artery is identified in the bleeding territory without evidence of other underlying lung disease, vasculitic changes or neoplasm.

Dieulafoy's disease is an important consideration in cases of massive hemoptysis in which more common causes have been excluded. Much remains to be learnt about this condition and the underlying cause remains an area of speculation. The case reported here in a young man with no past medical history or exposures supports the argument for a congenital aetiology. In the series described by Parrot *et al.*, all seven patients were current, heavy smokers [5]. However, smoking need not be a prerequisite as our, and other, cases demonstrate [1]. While most reported cases involved bronchial arteries, van Der Werf *et al.* [3] reported a similar anomaly involving the pulmonary tree. This may explain the variable efficacy of arterial embolization.

Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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