palpable pathology to direct the extent of any wedge resection. The pleural effusion may have been prevented with oversewing of the cavities. However, this lesion was secretory and could have resulted in bronchorrhea instead. Nevertheless, we would recommend oversewing any such lesions in this setting. We speculate that the fluid was coming from the parenchymal lymphatics, and that the vacuum effect probably potentiated the flow. A literature review at the time we were considering radiation showed that others had reported limited success in non-BAC-pleural effusions using steroids, indomethicin, atropine, and adrenocorticotropic hormone, which were discussed and dismissed [2, 6, 7]. We present these for future consideration only.

In conclusion, should a thoracic surgeon encounter this rare entity either after surgery or in consultation for pleural effusion after percutaneous lung biopsy by radiology or pulmonary medicine, early and aggressive measures must be taken to avoid the loss of lung function and the cachexia associated with persistent drainage. This may be especially important if the incidence of BAC is increasing.

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Dieulafoy's Disease: A Cause of Massive Hemoptysis That is Probably Underdiagnosed

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Dieulafoy's disease is a vascular anomaly characterized by the presence of a tortuous dysplastic artery in the submucosa. The condition was first described as a cause of gastrointestinal bleeding in the stomach. Recently, there have been a few reports of Dieulafoy's disease involving the respiratory tract. Herein, we report 2 patients with massive hemoptysis who were treated with surgical resection and later diagnosed with bronchial Dieulafoy's disease.

(Ann Thorac Surg 2005;80:1126–8)

Dieulafoy's disease is a vascular anomaly characterized by the presence of a tortuous dysplastic artery in the submucosa; most cases involve the gastrointestinal tract [1, 2]. The French surgeon Georges Dieulafoy first described it in 1898 [3]. This condition features massive and recurrent hemorrhage. As a result, it is considered extremely serious and is a potentially life-threatening cause of gastrointestinal bleeding [1]. Dieulafoy's disease was initially described as a stomach lesion but has since been identified in other parts of the body, including the esophagus, small intestine, colon, and rectum [2]. Recently, there have been a few reports of occurrence in the respiratory tract [3–7]. In this article, we describe two cases in which massive hemoptysis was managed surgically, and the patients were diagnosed with Dieulafoy's disease postoperatively.

Case Reports

Case 1

A 28-year-old man was admitted to the hospital with sudden onset of massive hemoptysis. He had had another less severe episode of hemoptysis 18 years earlier but was otherwise healthy. He mentioned that he had completed a course of antituberculosis therapy after the initial episode. He was a smoker with a 15 pack per year habit. When he was hospitalized at our center with the second episode, massive hemoptysis was the only abnormal finding on initial clinical examination. Blood biochemistry parameters were within normal limits, and a posteroanterior chest roentgenogram was unremarkable. On thoracic computed tomography, the only abnormality detected was a nodular lesion in the left lower lung lobe. A diagnostic workup for tuberculosis was negative. During bronchoscopy, a fibrin clot was observed protruding from normal-looking mucosa in the left lower lobe. The appearance suggested a possible endobronchial tumor. An attempt to obtain a biopsy sample from the site induced significant arterial bleeding. The bleeding stopped spontaneously when attempts to perform a biopsy were stopped, and the bronchoscopy procedure ended without any tissue collected. The tentative diagnosis for the lesion was carcinoid tumor. The patient underwent left thoracotomy and left lower lobectomy. The patient's recovery was uneventful, and he remains well at 12 months of follow-up.

Accepted for publication Feb 17, 2004.

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0003-4975/05/$30.00
doi:10.1016/j.athoracsur.2004.02.118

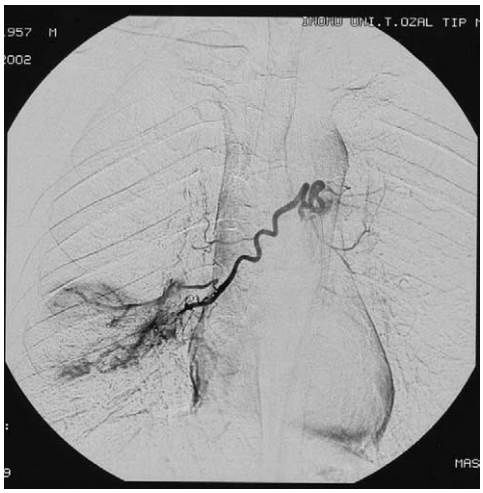

Fig 1. Selective bronchial arteriogram shows a dilated, tortuous right bronchial artery and profusely hypervascularized right middle lobe.

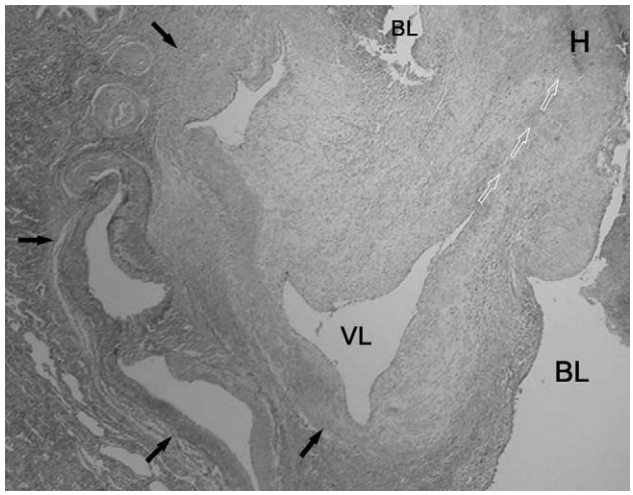

*Fig 2. Case 1: Abnormal, large vessel subepithelial area (*black arrows*). A thick-walled vessel communicating with the hematoma in the bronchial lumen (*open arrows*) (elastic van Gieson, ×40). (*BL = *bronchial lumen;* H = *hematoma;* VL = *vascular lumen.)*

Case 2

A 45-year-old man presented with massive hemoptysis. He was a smoker with a 35 pack per year habit and had experienced numerous similar episodes of severe hemoptysis in the 4 years before presentation. The only abnormalities detected on a posteroanterior chest roentgenogram and thoracic computed tomography were peribronchial lymphadenopathy in right middle lobe bronchus and consolidation in the same lobe. A middle lobe syndrome was considered, but bronchoscopy revealed no obvious narrowing of the middle lobe bronchus. Blood clots were noted in the middle lobe bronchus, but there was no active bleeding. Biopsy specimens of the bronchial mucosa were nondiagnostic. Skin tuberculin testing with purified protein derivative resulted in 17-mm diameter induration, so antituberculosis therapy was prescribed. He completed the full course of antituberculosis treatment. Recurrence of hemoptysis developed in the patient during this treatment, but he refused surgical intervention.

Further workup with bronchial arteriography revealed a tortuous, dilated right bronchial artery originating from the aorta at the level of the sixth thoracic vertebra and profuse bronchial hypervascularization of the right middle lobe (Fig 1). These lesions were interpreted as vascular changes due to recurrent inflammation in the middle lobe, and the abnormal bronchial artery was successfully embolized with polyvinyl alcohol particles.

One year after this intervention, the patient had another episode of massive hemoptysis. Roentgenographic investigation showed persistent consolidation in the middle lobe. As was observed during previous bronchoscopy procedures, bronchoscopy at this stage indicated that the bleeding source was in the middle lobe bronchus. Middle lobectomy was performed. His surgical recovery was uneventful, and he has been well through 5 months of follow-up.

Microscopic Findings

Microscopic examination of the surgical specimens revealed large thick-walled vessels in the wall of the bronchus and in the lung parenchyma (Figs 2, 3). One of these abnormal vessels opened into the bronchial lumen (Fig 2). In sections prepared with elastic van Gieson stain, the external and internal elastic laminae were only partially visible in the walls of some of the abnormal vessels. Masson trichrome staining demonstrated increased amounts of collagen in the intimal and medial layers of the walls of these vessels. The respiratory epithelium showed focal squamous metaplasia and diffuse thickening of the basal membrane. The pathologic diagnosis was Dieulafoy's disease of the bronchus.

On macroscopic inspection, we observed two firm,

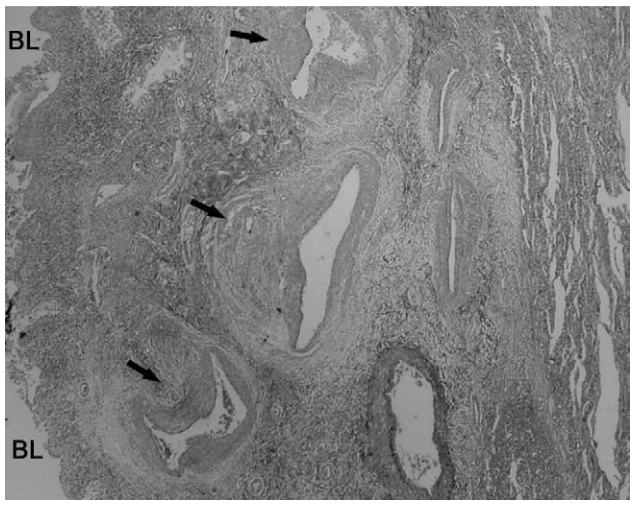

*Fig 3. Case 2: Abnormal dilated vessels underlying the bronchial mucosa (*arrows*) (elastic van Gieson, ×40). (*BL = *bronchial lumen.)*

yellow-white nodular masses (0.2 and 0.1 cm in diameter) in the resected lung parenchyma in the patient in case 1. Sections of these lesions prepared with hematoxylin-eosin showed central necrosis surrounded by hyalinized tissue and calcification, with foreign body–type giant cells at the periphery. No acid-fast bacilli were detected on Ziehl-Neelsen staining.

Sections of parenchyma from the patient in case 2 showed plugs of fibroblastic tissue blocking the alveoli and alveolar ducts, but no signs of chronic inflammation. Follicular hyperplasia was noted in the lymph nodes. These features were consistent with bronchiolitis obliterans organizing pneumonia and reactive lymphadenopathy, respectively.

Comment

Bronchial Dieulafoy's disease is extremely rare, and the etiology of this condition is still under debate. In two cases reported by Sweerts and colleagues [4], there was no obvious cause of vascular abnormality and the lesions were assumed to be congenital. In contrast, the history of pulmonary tuberculosis documented in two other published cases of Dieulafoy's disease suggests that chronic inflammation might lead to an acquired vascular pathology [5, 6]. Chronic inflammatory reactions were observed in both cases, although the direct role of chronic inflammation in the evolution of the vascular pathology remains ambiguous. This disorder may be congenital, and association with chronic inflammation may contribute to its clinical outcome as hemoptysis.

Recurrent hemoptysis is common in bronchial Dieulafoy's disease, and patients often have several episodes before the definitive diagnosis is made. Clinical and radiographic workup in these cases typically reveals no specific cause of the bleeding. The bronchoscopic appearance in patients with bronchial Dieulafoy's disease is not diagnostic because the site where the abnormal vessel opens into the bronchus is usually a pinpoint mucosal defect surrounded by normal-appearing mucosa and this small defect is often not seen on bronchoscopy due to pooling of blood or filling of the bronchial lumen with clots. This is what occurred in both of our cases. Bronchial Dieulafoy's disease can only be definitively diagnosed by histopathologic examination [7]; however, bronchial biopsy in such cases can result in severe hemorrhage [5]. In our first patient, pulsating bleeding occurred when we tried to sample a fibrin clot that we initially suspected was an endobronchial tumor. In the second case, there was no massive bleeding after bronchoscopic mucosal biopsy was performed; however, the samples collected were superficial and nondiagnostic, and we believe that this explains the lack of bleeding. It was recommended that bronchial arteriography was the most appropriate initial investigation in these cases [6]. There are no specific angiographic criteria for diagnosing Dieulafoy's disease, but the finding of a tortuous and ectatic artery, as detected in the patient in case 2, is suggestive of this condition.

Recent advances in endoscopic and angiographic therapy been widely accepted as modes for managing Dieulafoy's disease of the gastrointestinal tract. Surgery is reserved for patients who do not respond to angiographic and endoscopic forms of treatment [1]. In bronchial Dieulafoy's disease, selective embolization has been proposed as a method for stopping the bleeding [3, 6]. Bhatia and colleagues [3] reported that their patient had several hospital admissions due to repeated episodes of hemoptysis and that he was managed by embolotherapy on each occasion. However, bronchial artery embolization failed in one case in the literature [4], and the patient in our case 2 did not benefit from this intervention in the long term. In the case described by Stoopen and colleagues [7], the hemoptysis was initially controlled by bronchoscopic cauterization, but it recurred and the patient required surgical resection. At present, it appears that surgical treatment is still the most effective management option for bronchial Dieulafoy's disease because the bleeding in these cases is massive, often recurrent, and potentially fatal.

Dieulafoy's disease should always be included in the differential diagnosis for massive hemoptysis. Cases in which no definitive bleeding source is found on routine investigation are particularly suspicious. It is difficult to diagnose Dieulafoy's disease preoperatively, and this condition is often overlooked. We believe that this entity may be more common than the reports in the literature suggest. When bronchial Dieulafoy's disease is suspected based on bronchial arteriography, bronchial artery embolization should be considered as the first-line approach, and hazardous and unnecessary biopsy procedures should be avoided altogether. In uncontrolled cases, surgical treatment is a lifesaving approach that eliminates the possibility of recurrence and allows accurate histopathologic diagnosis of the disease.

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