

Bronchiectasis Following Colectomy in a Patient With Ulcerative Colitis and Factor V Leiden Mutation

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For some years it has been known that lung disease may be present in patients with inflammatory bowel disease (IBD). Diseases of the central airway, bronchi, and lung parenchyma are among the most common forms of lung involvement in IBD patients. Bronchiectasis is frequent and almost always appears after the onset of disease and in close association with inflammatory activity. However, reports of the appearance of bronchiectasis following colectomy for an exacerbation of IBD have been rare. We present the case of a 36-year-old man with severe ulcerative colitis who, following a total colectomy, developed bilateral bronchiectasis 12 months after surgery and for whom both preoperative and postoperative computed tomography scans were available.

Key words: *Bronchiectasis. Ulcerative colitis. Inflammatory bowel disease.*

Bronquiectasias tras colectomía en paciente con colitis ulcerosa y mutación del factor V Leiden

Desde hace años se sabe que los pacientes con enfermedad inflamatoria intestinal (EII) pueden presentar afectación pulmonar. De entre las formas de aparición, las más frecuentes incluyen la afectación de la vía aérea central, la afectación bronquial y la afectación del parénquima pulmonar. Las bronquiectasias son frecuentes en los pacientes con EII, casi siempre después del inicio de ésta y en muy íntima relación con la actividad inflamatoria de la EII. Sin embargo, los casos publicados sobre la aparición de bronquiectasias tras una colectomía por un brote de EII son excepcionales. Nosotros presentamos el caso de un varón de 36 años con una colitis ulcerosa grave que, tras una colectomía total, desarrolló un cuadro de bronquiectasias bilaterales a los 12 meses de la intervención, y en el que se dispone de tomografía axial computarizada antes y después de éstas.

Palabras clave: *Bronquiectasias. Colitis ulcerosa. Enfermedad inflamatoria intestinal.*

Introduction

Ulcerative colitis and Crohn's disease are traditionally grouped together under the term inflammatory bowel disease (IBD). It has been known for some years that lung disease can exist in patients with IBD.¹ While prevalence in asymptomatic patients is unknown, it may be around 50%^{2,3} in certain subgroups of patients. Diseases of the central airway, bronchi, and lung interstitium are among the most common forms of respiratory involvement. Bronchiectasis is a frequent form of bronchial involvement in patients with IBD, and it almost always appears after the onset of the disease and in close association with inflammatory activity. However, reports of the appearance of bronchiectasis following colectomy due to an exacerbation of IBD have been

rare.^{4,5} We report what is to our knowledge the only case of a patient with ulcerative colitis who developed bronchiectasis following colectomy for whom both preoperative and postoperative high-resolution computed tomography (HRCT) scans were available.

Case Description

The patient was a 36-year old man with no known history of alcoholism, smoking, or use of street drugs and with a history of frequent nephrolithiasis and ulcerative colitis diagnosed in 1995. He was referred to us from the gastroenterology clinic of our hospital, where he was being followed. He had severe corticosteroid-dependent ulcerative colitis that caused multiple episodes of diarrhea and associated complications. For the previous 3 years, the patient had been suffering from uveitis treated with topical corticosteroids and from inflammatory arthritis. During that time he had also had an episode of cold antibody hemolytic anemia. Attempts had been made on several occasions to control the patient's disease with 5-aminosalicylic acid, but in view of his poor response to the treatment, he was kept on 10 mg of prednisone a day. In September 2002 he underwent

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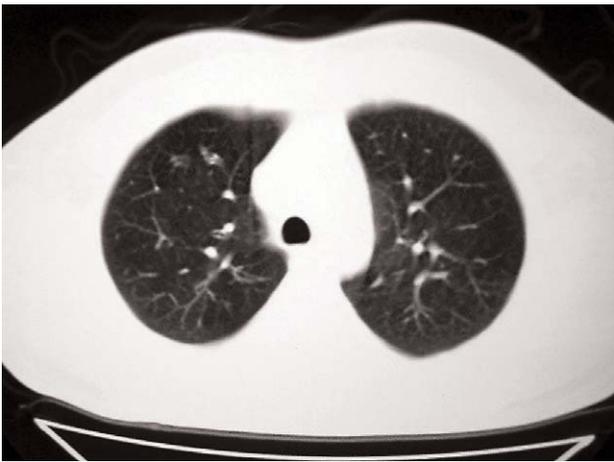


Figure 1. High-resolution computed tomography scan taken in 2002, before colectomy, showing normal lung parenchyma.



Figure 2. High-resolution computed tomography taken in 2003, after colectomy, showing bilateral bronchiectasis.

total colectomy with total ileostomy due to an exacerbation of ulcerative colitis with toxic megacolon. He was also diagnosed with hereditary thrombophilia (factor V Leiden mutation) after suffering a pulmonary thromboembolism in 1998 and deep venous thrombosis in 1999 following withdrawal of an oral anticoagulant; a brother of the patient was also subsequently diagnosed with the genetic defect. In March 2002 a HRCT scan was taken in the emergency department of our hospital to confirm or rule out suspected pulmonary thromboembolism, but no signs of pulmonary thromboembolism or alterations of the lung parenchyma were observed (Figure 1).

In September 2003 he was admitted to our department with right-sided subscapular pleuritic pain and frank hemoptysis of 15 mL that had begun 24 hours earlier, with no clinical picture of infection or fever. The patient had had no previous episodes of hemoptysis, although he presented a clinical picture of abundant mucopurulent morning expectoration, with no infection, dating from January 2003.

On examination the patient was found to be well perfused and without fever or dyspnea at rest. Arterial blood pressure was 120/70 mm Hg and arterial oxygen saturation 92%. Heart and lung auscultation revealed only rhonchi in the apical and medial regions of the right hemithorax and isolated crackles in the right-mid axillary line. Examination of the abdomen revealed only a laparotomy scar and the ileostomy bag. The lower limbs showed no edema or signs of deep venous thrombosis. Biochemistry was normal and the hemogram showed no evidence of anemia, leukocytosis, or neutrophilia. Coagulation testing showed an international normalized ratio of 2.4 with prothrombin activity of 34%. D-dimer assay results were negative. Arterial blood gases measured at an inspiratory oxygen fraction of 21% showed a PaO₂ of 62 mm Hg, PaCO₂ of 42 mm Hg, pH of 7.41, and arterial oxygen saturation of 92%. The chest x-ray taken upon admission revealed an alveolar infiltrate with an air-bronchogram sign in the right upper lobe. Sputum smear examinations and cultures were negative. A HRCT scan of the chest revealed signs of bilateral bronchiectasis with upper-lobe predominance, although the lower lobes were also involved. Bleeding bronchiectasis was found at the apex of the right lung (Figure 2). Bronchoscopy yielded no significant findings, and the microbiological samples obtained were negative for common

pathogens, acid-fast alcohol resistant bacilli, opportunistic pathogens, and fungi. Bronchoalveolar lavage showed an elevated neutrophil concentration. Lung function tests were performed with the following results: forced expiratory volume in 1 second (FEV₁), 70% (2.93 L); forced vital capacity (FVC), 88.2% (4.41 L); ratio of FEV₁ to FVC, 66%, and maximal midexpiratory flow rate, 30%. Carbon monoxide diffusing capacity was normal, and a methacholine challenge test was negative. Clinical signs had disappeared 24 hours after admission. The patient was discharged and prescribed a 6-month course of systemic corticosteroids at doses higher than those he had been using, together with inhaled corticosteroids and bronchodilators. The HRCT findings persisted at the 6-month follow-up visit, as did the signs of obstructed breathing observed in spirometry, although the daily sputum production had decreased.

Discussion

While it has been known for years that lung disease may be present in patients with IBD, the true incidence of lung involvement is unknown. True IBD-associated lung disease must be distinguished from lung disease produced as a consequence of the immune suppressant effects of treatment (mainly with sulfasalazine,⁶ in which lung involvement usually takes the form of eosinophilic pneumonia). True IBD-associated lung disease includes diseases of the airway, of lung interstitium, or other rarer conditions, such as serositis, necrobiotic nodules, and pulmonary vasculitis.⁷ In 85% of cases, pulmonary manifestations appear after the onset of IBD, and in 28% of cases symptoms begin following colectomy. The Table shows the wide spectrum of IBD-associated lung diseases

Camus et al⁸ say that the appearance of IBD-associated bronchiectasis has been described in several articles that generally show it to be more frequent in patients with ulcerative colitis than in those with Crohn's disease and to appear in a period of time ranging from days to years following the onset of IBD

TABLE
Types of Lung Disease Associated With Inflammatory Bowel Disease

Site of Involvement	Disease
Glottis, larynx	Glottic, subglottic stenoses
Trachea	Tracheal inflammation and stenoses
Bronchi	Simple chronic bronchitis Isolated mucoid impaction Scarred bronchial granulomas Chronic bronchial suppuration Bronchiectasis
Peripheral airway	Granulomatous bronchiolitis Necrotizing bronchiolitis Diffuse panbronchiolitis Bronchiolitis obliterans
Lung parenchyma	Respiratory bronchiolitis associated with diffuse interstitial lung disease Nonspecific interstitial pneumonia Granulomatous interstitial lung disease Desquamative interstitial pneumonia Pulmonary infiltrates with eosinophilia Necrobiotic nodules

symptoms. Frequency is not greater in any particular age group. The most common symptom is bronchial hypersecretion, and onset is usually insidious, but benign. The course of the disease usually parallels that of IBD, with episodes of respiratory symptoms occurring together with episodes of intestinal disease. This has led to the hypothesis that the disease might arise in the lung, as both the digestive tract and the lung have the same embryonic origin. Cases of bronchiectasis following colectomy lend support to this hypothesis; in such cases, when the organ that is the target of the inflammatory process is removed, the respiratory system becomes the next target of inflammatory damage.⁹ Foci of bronchiectasis are usually sterile and only show elevated levels of neutrophils in bronchoalveolar lavage.^{10,11}

A MEDLINE search located only 2 reports of bronchiectasis following a colectomy in a patient with IBD, and in neither case was a preoperative tomography scan available. In the case we report, given that a HRCT scan had been taken before the colectomy, we were able to compare the regions of the lung parenchyma before and after surgery.

IBD-associated bronchiectasis generally responds well to corticosteroid therapy,^{4,8} usually given in conjunction with treatment for the underlying disease. High-dose inhaled corticosteroids are the first treatment option, followed by oral prednisone (1-2 mg/kg/day). No evidence is available to support an optimal duration of treatment. Cases have been reported in which the development of side effects and poor control of the respiratory disease led to the use of serial bronchial lavages with a corticosteroid solution. Response to therapy in those cases was good.⁸ In the case we report, corticosteroid treatment led to a clinical but not to a radiologic or functional response.

The case we describe contributes to a better understanding of the pathogenesis of IBD-associated lung disease, especially in relation to colectomy. It also underscores the need for patients with IBD who develop respiratory symptoms to be referred to a pneumologist as soon as possible, as delay in defining the clinical picture and establishing a diagnosis can make repair of the lung parenchyma impossible even with proper treatment.

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