Cryptogenic Organizing Pneumonia With Lung Nodules Secondary to Pulmonary Manifestation of Crohn Disease

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ABSTRACT: Crohn disease is an immune-mediated inflammatory condition with gastrointestinal and extraintestinal manifestations in patients. Pulmonary involvement of Crohn disease is one manifestation. There have been case reports which have shown Crohn disease and lung nodules which were noted to be histopathological as cryptogenic organizing pneumonia (COP). In our case, a 22-year-old woman with Crohn disease was seen with complaints of chest pain and cough. Computed tomographic scan of chest showed multiple bilateral lung nodules, for which biopsy was done, which showed COP. The case study is followed by a deeper discussion of COP and the extraintestinal manifestation seen in inflammatory bowel disease.

KEYWORDS: COP, Crohn disease, lung nodules

Introduction

Crohn disease is an immune-mediated inflammatory condition with known gastrointestinal and extraintestinal manifestations including pulmonary involvement. Previous case reports have shown Crohn disease with lung nodules which were noted to be histopathological as cryptogenic organizing pneumonia (COP).¹,²

Case Report

A 22-year-old woman with a known past medical history of Crohn disease and primary sclerosing cholangitis originally was referred for pulmonary evaluation when she presented to her primary care physician with chest pain and cough. Patient denied having any weight loss, fever, chills, diarrhea, constipation, and abdominal pain. At this time, chest x-ray was done, which showed several lung nodules. Further workup was done which included a computed tomographic (CT) scan of chest showed multiple bilateral lung nodules. Her Crohn disease and sclerosing cholangitis have remained in remission and was not on therapy for Crohn disease for several years. Blood work from pulmonary office showed normal complete blood count, comprehensive metabolic panel, elevated erythrocyte sedimentation rate, positive atypical Anti-neutrophil cytoplasmic antibody (ANCA), negative antineutrophil cytoplasmic antibody, high normal antiproteinase antibody, normal angiogenin-converting enzyme level, and negative Gold Quantiferon for tuberculosis. C-reactive protein was markedly elevated. Pulmonary function test revealed a restrictive defect with decreased diffusion capacity with a total lung capacity of 69% of predicted, FEV₁ at 84%, and DLCO at 58%. Arterial blood gas done at room air showed pH of 7.45, Pco₂ of 25, Po₂ of 123.

Chest x-ray and CT scan showed multiple lung nodules in the upper lung lobes bilaterally (Figure 1). CT-guided lung biopsy on 1-cm pleural-based left lung nodule showed COP on pathology. Patient did not follow-up with pulmonary regarding medical treatment as scheduled and returned to hospital approximately 2 months later with complaints of chest pain, back pain, cough, fever, and chills for approximately 3 weeks and was at this time 7 to 8 weeks pregnant. Patient was started on treatment with prednisone for organizing pneumonia with lung nodules secondary to pulmonary manifestation of Crohn disease. Patient was to be monitored closely by obstetrics-gynecology and pulmonary throughout treatment during pregnancy, but patient went to another institution for follow-up.

Discussion

Cryptogenic organizing pneumonia is a condition that has been described in medical literature.³,⁴ Typical patient presentation involves a gradual onset of generalized symptoms including...
fever, night sweats, cough, and malaise. Physical examination of patients with suspected bronchiolitis obliterans organizing pneumonia may include crackles and rales, but it is estimated that one-fourth of patients have a normal physical examination. Lab investigations can demonstrate leukocytosis, elevated acute phase reactants including erythrocyte sediment rate, and C-reactive protein, although these findings are not always found in all cases. Cultured specimens typically are void of microorganisms. Radiographic imaging typically demonstrates a generalized organizing pneumonia which may include a ground-glass appearance. Pulmonary function tests may demonstrate a restrictive pattern. Patients are often treated with one to multiple antimicrobial agents including but not limited to macrolides with no clinical improvement.

The clinical picture of COP then warrants the introduction of corticosteroids to prevent the continued development of granulation tissue and destruction of healthy tissue leading to fibrosis. Although prevalence and incidence of COP are unknown, it is believed that up to two-thirds of patients treated with steroids will have resolved symptoms; however, slow convalescence over weeks to months is notably more common. Optimal dosing of steroids is not known at this time.

Although limited information is available regarding direct causative links of COP, many case series and reports have identified that inflammatory conditions have been linked to the disease including systemic lupus, rheumatoid arthritis, scleroderma, and in the case of this patient, Crohn disease. Extraintestinal pulmonary involvement has been noted in inflammatory bowel disease (IBD) with case reports demonstrating more cases in Crohn disease than ulcerative colitis. Inflamm Bowel Dis. 2006;9:104–115.

Although the direct cause of pulmonary inflammation in Crohn disease is unknown, it is suggested that inflammatory cells in the lung accumulate and occlude the small airways, much like the inflammatory response seen in the gastrointestinal tract in Crohn disease. These inflammatory responses can lead to airway disease, parenchymal disease, serositis, and in some cases even pulmonary embolism.

In the setting of IBD, airway involvement has included bronchiectasis, acute and chronic tracheobronchitis, bronchiolitis, subglottic stenosis, and fistula formation. Although large and small airways can be affected by the inflammation, it is more common in the smaller airways. Of note, there are many patterns of respiratory parenchymal disease, but COP and interstitial lung disease are most common. Accounts of sarcoidosis, necrotic nodules, and infiltrates with eosinophilia have been reported, as well as serositis with the potential to cause development of pleural effusions, pericarditis, and myocarditis. Both clinical and diagnostic workups involve exclusion of other pathologies to rule in the diagnosis of pulmonary manifestations of Crohn disease. Examination of case reports and current medical literature provide insight into the possible extraintestinal manifestations of the patient with Crohn disease while still citing the need for further clinical and laboratory workup, as well as treatment response and analysis.

REFERENCES

1. Black H, Mendoza M, Murin S. Thoracic manifestations of inflammatory bowel disease. Chest. 2007;131:524–532.
2. Nelson BA, Kaplan JL, El Saleby CM, Lo MT, Mark EJ. Case records of the Massachusetts General Hospital. Case 39-2014. A 9-year-old girl with Crohn’s disease and pulmonary nodules. N Engl J Med. 2014;371:2418–2427.
3. Gudmundsson G, Sveinsson O, Isaksson HJ, Jonsson S, Frodotdottir H, Aspelund T. Epidemiology of organising pneumonia in Iceland. Thorax. 2006;61:805–808.
4. Kraft SC, Earle RH, Roessler M, Esterly JR. Unexplained bronchopulmonary disease with inflammatory bowel disease. Arch Intern Med. 1976;136:454–459.
5. Corder JF. Cryptogenic organizing pneumonia. Eur Respir J. 2006;28:422–446.
6. King TE Jr. Organizing pneumonia. In: Schwarz MI, ed. Interstitial Lung Disease. 5th ed. Shelton, CT: People’s Medical Publishing House; 2011:981.
7. Corder JF. Cryptogenic organizing pneumonia. Bronchiolitis obliterans organizing pneumonia. Clin Chest Med. 1993;14:677.
8. Corder JF, Loire R, Brune J. Idiopathic bronchiolitis obliterans organizing pneumonia. Definition of characteristic clinical profiles in a series of 16 patients. Chest. 1980;96:999–1004.
9. Stover DE, Mangino D. Macrolides: a treatment alternative for bronchiolitis obliterans organizing pneumonia. Chest. 2005;128:3611–3617.
10. Camus P, Paillat P, Ashcroft T, Gal AA, Colby TV. The lung in inflammatory bowel disease. Medicine (Baltimore). 1993;72:151–183.
11. Sostegni R, Daperno M, Pera A. Pulmonary manifestations of inflammatory bowel disease. Dig Liver Dis. 2007;39:239.
12. Tunc B, Filik L, Bilgic F, Arda K, Ulker A. Pulmonary function tests, high-resolution computed tomography findings and inflammatory bowel disease. Acta Gastroenterol Belg. 2006;69:255–260.
13. Higenbottam T, Cochrane GM, Clark TJ, Turner D, Millis R, Seymour W. Bronchial disease in ulcerative colitis. Thorax. 1980;35:381–385.
14. Mahadeva R, Walsh G, Flower CD, Shneerson JM. Clinical and radiological characteristics of lung disease in inflammatory bowel disease. Eur Respir J. 2000;15:43–48.
15. Storch I, Sachar D, Katz S. Pulmonary manifestations of inflammatory bowel disease. Inflamm Bowel Dis. 2003;9:104–115.
16. Kuzmiak T, Sleiman C, Brugière O, et al. Severe tracheobronchial stenosis in a patient with Crohn’s disease. Eur Respir J. 2010;35:209–212.
17. Spira A, Grossman R, Balter M. Large airway disease associated with inflammatory bowel disease. Chest. 1998;113:1723–1726.