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

Taufiq Zaman[1], Joseph Watson[2] and Mohammad Zaman[1]

[1]Department of Medicine, Brookdale University Hospital Medical Center, Brooklyn, NY, USA.
[2]Ross University School of Medicine, Miramar, FL, USA.

Clinical Medicine Insights: Case Reports
Volume 10: 1–2

DOI: 10.1177/1179547617710672
SAGE

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

RECEIVED: August 1, 2016. **ACCEPTED:** April 15, 2017.

PEER REVIEW: Four peer reviewers contributed to the peer review report. Reviewers' reports totaled 817 words, excluding any confidential comments to the academic editor.

TYPE: Case Report

FUNDING: The author(s) received no financial support for the research, authorship, and/or publication of this article.

DECLARATION OF CONFLICTING INTERESTS: The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

CORRESPONDING AUTHOR: Taufiq Zaman, Department of Medicine, Brookdale University Hospital Medical Center, 1 Brookdale Plaza, Brooklyn, NY 11212, USA.
Email: tzaman09@gmail.com

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 antimyeloperoxidase antibody, high normal antiproteinase antibody, normal angiotensin-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, FEV_1 at 84%, and D_{LCO} at 58%. Arterial blood gas done at room air showed pH of 7.45, Pco_2 of 25, Po_2 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.

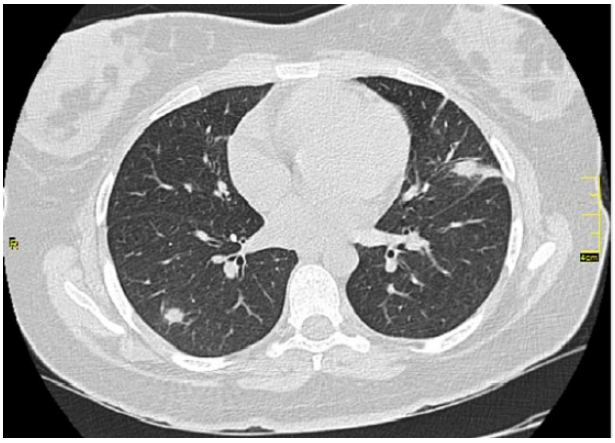

Figure 1. Computed tomography of chest showing lung nodules.

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.[5] 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.[6] 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.[6,7] Cultured specimens typically are void of microorganisms. Radiographic imaging typically demonstrates a generalized organizing pneumonia which may include a ground-glass appearance.[8] Pulmonary function tests may demonstrate a restrictive pattern.[5,7] Patients are often treated with one to multiple antimicrobial agents including but not limited to macrolides with no clinical improvement.[9]

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.[5,10] 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.[6] Extraintestinal pulmonary involvement has been noted in inflammatory bowel disease (IBD) with case reports demonstrating more cases in Crohn disease than ulcerative colitis.[11,12] 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.[13] These inflammatory responses can lead to airway disease, parenchymal disease, serositis, and in some cases even pulmonary embolism.[1,10,14,15]

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.[15-17] Of note, there are many patterns of respiratory parenchymal disease, but COP and interstitial lung disease are most common.[1,10] Accounts of sarcoidosis, necrobiotic nodules, and infiltrates with eosinophilia have been reported, as well as serositis with the potential to cause development of pleural effusions, pericarditis, and myocarditis.[2,10,15] 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.

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