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CASE REPORT | INFLAMMATORY BOWEL DISEASE

Necrobiotic Pulmonary Nodules in Ulcerative Colitis: Not Just a "Crohnic" Phenomenon

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ABSTRACT

Necrobiotic pulmonary nodules are an exceptionally rare extraintestinal manifestation of inflammatory bowel disease. Recognition is imperative because it may mimic other autoimmune pathologies such as granulomatosis with polyangiitis or sarcoidosis. We describe a 19-year-old man with a known history of ulcerative colitis who was found to possess bilateral pulmonary nodules on computed tomography imaging. Investigations that included an extensive autoimmune and infectious workup were inconclusive. Biopsy of the nodules revealed fibrinous exudate and palisading histiocytes that confirmed the diagnosis. He was started on prednisone therapy. A follow-up computed tomography a month later revealed near complete resolution.

INTRODUCTION

Extraintestinal manifestations in inflammatory bowel disease (IBD) affect up to 50% of all those affected.[1] Pulmonary complications of IBD have been less commonly reported, most of which involve the upper airways, resulting in bronchitis and bronchiectasis.[2,3] Pulmonary nodules, particularly those of necrotizing pathology, are exceedingly rare.[4] Its recognition is important because it may mimic infectious, autoimmune, or vasculitic pathologies. The findings in our case coincide with the findings classically described in IBD-associated lung nodules. They are often bilateral, possess a necrotic granulomatous infiltrate on histopathology, and finally respond well to steroid therapy.[5] The following case seeks to aid in the formulation of a stepwise diagnostic approach and highlights common pitfalls.

CASE REPORT

Our patient is a 19-year-old man with a known history of ulcerative colitis (UC) presented with complaints of bloody stools, fatigue, and unintentional weight loss for 2 weeks. Before this, he had been diagnosed with UC, 2 months ago, when he had presented with similar symptoms. Colonoscopy from the time revealed diffuse involvement of the entire colon including the rectum. He had been placed on 1.2 g twice a day of mesalamine for the same, and adequate symptom control had been achieved (mild disease as per severity classification) until this emergency department visit. On admission, he had denied the presence of fever, chills, chest pain, shortness of breath, cough, abdominal pain, nausea, or vomiting.

On initial assessment, the patient was hemodynamically stable. His physical examination revealed the presence of conjunctival pallor, although the rest of his examination, including his abdominal examination was benign. His laboratory test results were significant for a hemoglobin level of 9.3 gm/dL that was at baseline and a white blood cells count of 10,600 cells/μL. His serum lipase, aspartate transaminase, alanine transaminase, alkaline phosphatase, and bilirubin levels were within the normal range. Stool studies were negative for the presence of common diarrheal pathogens. An abdominal computed tomography (CT) scan revealed the presence of diffuse thickening of the colon and incidentally identified multiple ill-defined nodules at the bases of the lungs that were presumed to be possibly infectious or embolic in origin. He had received intravenous hydration with normal saline and antibiotics for presumed sepsis.

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The following day, he underwent a flexible sigmoidoscopy that revealed pancolitis, suggesting a flare up of his UC. Abdominal and thoracic CTs were obtained to better delineate the nodules (Figure 1). This had demonstrated diffuse cavitating pulmonary nodules present bilaterally ranging from 0.2 to 1.6 cm. Infectious workup that included blood cultures, interferon-gamma release assay for tuberculosis, urine histoplasma antigen, serum cryptococcal antigen, blastomyces antibody, and coccidioides antibody was negative. Human immunodeficiency virus testing was negative as well. A rheumatologic workup was then obtained, which was significant for a positive antinuclear antibody of homogenous pattern at 250 dilutions (ref 1–49 dilutions). In addition, positive were Scl-70 antibodies, anticentromere antibodies, and antineutrophil cytoplasmic autoantibody (ANCA). However, he did not fulfill the systemic lupus erythematosus or scleroderma criteria, and it was inferred that the immune markers were elevated because of his IBD flare. An interventional radiology-guided biopsy of the nodules was performed. Histopathology of the biopsy revealed the presence of giant histiocytes and fibrinous material (Figure 2). A diagnosis of necrobiotic pulmonary nodules in the setting of IBD was made. The patient was discharged on 40 mg of prednisone daily in addition to mesalamine. A repeat thoracic CT during 1-month follow-up revealed almost complete resolution of the pulmonary nodules with better control of his UC (Figure 3).

DISCUSSION

Although extraintestinal manifestations of IBD have been described as early as in the 1960s, it was only until much later that the pulmonary manifestations came to light.[6,7] The patterns of pulmonary involvement include chronic bronchitis (21%), bronchiectasis (24.6%), interstitial lung disease (26%), and necrobiotic pulmonary nodules (5.3%).[8,9] The occurrence of pulmonary nodules is exceptionally rare and often a diagnosis of exclusion.[9] Although previous articles have suggested that the occurrence of pulmonary nodules is more common in UC than Crohn disease, more recent data appear to suggest otherwise.[9–12]

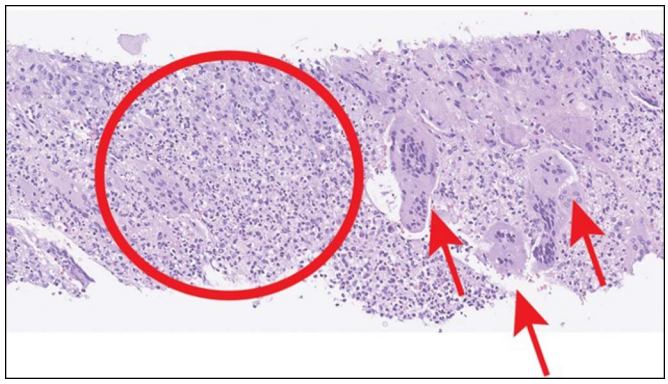

Figure 2. Chronic fibrinopurulent exudate (represented by the circle) and histiocytic giant cells (represented by the arrows).

There are similarities between cases published in literature and our own case, with a few notable differences. The classical patient developing necrobiotic pulmonary nodules is often a young individual (age <30 years) with a known diagnosis of IBD who presents with respiratory symptoms.[13–15] Most cases described in literature reveal the presence of active luminal disease.[10,11] Although our patient did fit the reported age bracket and presented with active colitis, he did not present with respiratory symptoms and was thus diagnosed incidentally. His asymptomatic presentation is of paramount importance because many of these nodules may serve as a nidus for infection.[16] A thorough respiratory evaluation may thus be warranted in a patient with IBD.

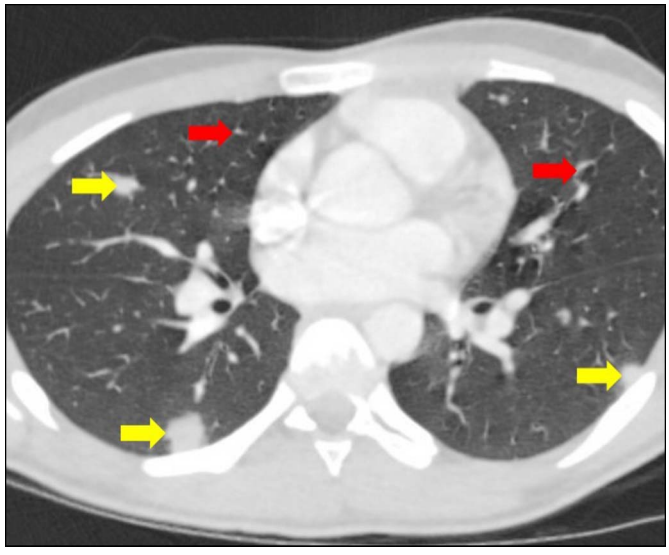

Figure 1. Thoracic computer tomography revealing multiple pulmonary nodules (represented by yellow arrow) and cavitating lesions (represented by red arrow).

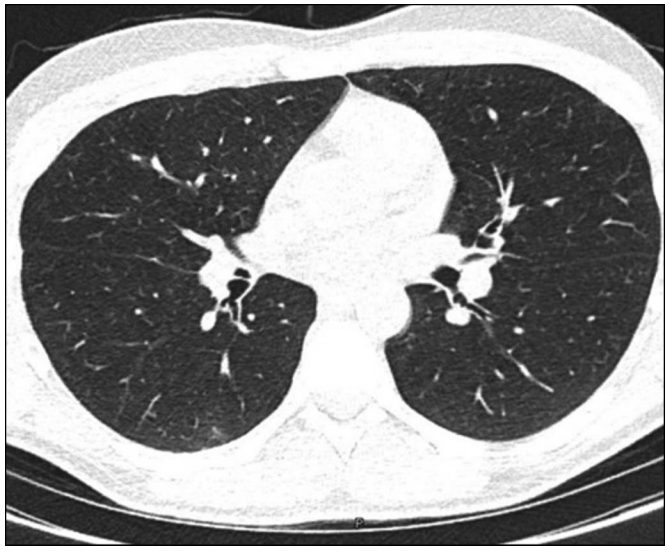

Figure 3. Follow-up thoracic computed tomography revealing near complete resolution of the lesions.

The classical radiological finding described is a diffuse interstitial lung disease with the presence of bilateral pulmonary nodules measuring between 8 and 30 mm, many are reported to be subpleural in nature as was the case in our patient.[11,13,]14,16 The diagnostic workup typically includes a comprehensive infectious and autoimmune evaluation that is usually inconclusive.[5,17,18] Histopathology of the nodules almost universally reveals the presence of a chronic infiltrate with the presence of necrotic material often with the presence of histiocytes.[5,14,17]

The importance of the case rests in its ability to masquerade as other autoimmune and infectious phenomenon. Differentials include granulomatosis with polyangitis, sarcoidosis, and infectious emboli.[9,11,19,20] Distinguishing one from the other is a challenging task because differences may be subtle and require thorough investigation. For instance, in granulomatosis with polyangiitis, the ANCA is predominantly that of the proteinase 3 (PR3-ANCA) subtype, whereas the ANCA in UC is of the peripheral type (p-ANCA).[11,20] Ruling out sarcoidosis may prove to be more challenging and may have to rely on bronchoalveolar lavage testing and Gallium 67 scanning.[18]

Although a few of the cases in literature did reveal spontaneous resolution, most of the symptomatic cases required the administration of immunomodulatory therapy such as prednisone and mesalazine.[5,11,17,18] The commonly used regimen was that of prednisone at 1 g/kg/d that resulted in significant reduction in the size of the lung nodules.[5,12–14,17] Owing to the scarcity of literature, the predilection of pulmonary nodules toward either 1 of the 2 types of IBD remains controversial. Suffice to say that more data are required for the same. Based on our literature review, this would be the seventh case in literature describing the occurrence of necrobiotic nodules in UC.

DISCLOSURES

Author contributions: S. Mukherjee wrote the manuscript, reviewed the literature, and is the article guarantor. P. Harne wrote the manuscript, reviewed the literature, and revised the manuscript for intellectual content. K. Mirchia wrote the manuscript and revised the manuscript for intellectual content. A. Sharma and D. Manocha revised the manuscript for intellectual content.

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