Rare disease

A case presentation of a pulmonary complication of ulcerative colitis

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Summary

We present the case of a 25-year-old Afro-Caribbean man with a longstanding history of ulcerative colitis and primary sclerosing cholangitis. The patient presented to clinic and reported pleuritic-type chest pain. A routine chest radiograph requested from the clinic revealed an incidental right middle zone opacity in the right lung. A subsequent high-resolution CT showed multiple lung nodules. The patient also had a positive cytoplasmic anti-neutrophil cytoplasmic antibody (cANCA) and proteinase 3 antibodies. Bronchoscopy was inconclusive. A video-assisted thoracoscopic surgery biopsy was then taken. The histology revealed changes suggestive of bronchiolitis obliterans organising pneumonia. The pulmonary manifestations of inflammatory bowel disease are poorly characterised. Our literature search has revealed cases hypothesising that immune system dysregulation could display pulmonary complications of ulcerative colitis. The aetiology is thought to be related to the treatment with mesalazine. However, our patient also had a positive vasculitic screen. Previous cases have resolved with supportive management or steroid therapy.

BACKGROUND

Pulmonary manifestations of inflammatory bowel disease (IBD) are rare and most general clinicians may not have even heard of them. Hence, it is an important learning point and diagnosis to consider in patients with IBD presenting with respiratory symptoms.

CASE PRESENTATION

Introduction

Up to 25% of patients with IBD will display a combination of extraintestinal manifestations.¹ The organs most commonly affected include the skin, joints, biliary tract and eyes. However, the pulmonary manifestations of IBD are less commonly reported. A link between pulmonary disease and IBD was suggested nearly 40 years ago.² Case reports and series document a spectrum of respiratory pathology in association with IBD that spans from the larynx to the pleura. These can be briefly classified into pathologies affecting the airways (bronchiectasis, bronchiolitis and subglottic stenosis): the lung parenchyma, the interstitium, pulmonary infiltrates with eosinophilia, serositis and drug-induced pulmonary disease mainly associated with 5-aminosalicylic acid (5-ASA) compounds. However, bronchiectasis is the single most common disorder.³

Case report

A 25-year-old gentleman with a 4-year history of ulcerative colitis and primary sclerosing cholangitis presented to clinic with a 4-week history of intermittent pleuritic chest pains. He also complained of a productive cough (but no haemoptysis), night sweats and some weight loss during this time. He denied any significant dyspnoea on exertion and his exercise tolerance was not affected.

His last flare of ulcerative colitis had been over a month prior to the onset of the respiratory symptoms. The patient had no other significant medical history and had been treated with azathioprine and occasional courses of mesalazine since the diagnosis.

The patient is of Jamaican origin and was born in the UK. He had last travelled to Jamaica 10 years ago and had no personal or contact history of tuberculosis. He also had no contact with birds or other domestic animals.

Respiratory and cardiovascular examination was unremarkable and there was no lymphadenopathy or organomegaly.

Routine bloods tests revealed raised inflammatory markers (C reactive protein 77, white cell count 17 and neutrophils 12.2) and deteriorating liver function (alanine transaminase 186, alkaline phosphatase 1592 and bilirubins 21).

A chest radiograph in October 2010 (figure 1) was reported as normal but another one in May 2011 showed right middle zone opacity (figure 2). A CT scan was then performed, which was reported as showing multiple bilateral lung nodules with minor ground glass changes around them. The lung nodules were thought to be due to an infective process, possibly as a result of immunosuppression (see figure 3A,B).

In the interim, tuberculin skin testing and sputum tests for acid-fast bacilli excluded a diagnosis of tuberculosis.

Pulmonary function tests did not reveal any significant abnormality.

INVESTIGATIONS

It was thought that the nodules were most likely due to vasculitis or mesalazine-induced variant of Wegener's granulomatosis which has been reported in the literature.

Figure 1 Chest radiograph from October 2010.

This was supported by the patient having positive cANCA and proteinase 3 antibodies.

A subsequent bronchoscopy revealed copious watery secretions, but no other abnormality in the bronchial tree. A video-assisted thoracoscopic surgery biopsy was then taken. The histology revealed a well-circumscribed area of consolidation with many of the alveolar spaces with granulation tissue accompanied by foamy-type macrophages suggestive of organising pneumonia, formally known as bronchiolitis obliterans organising pneumonia; see figure 4. There was no evidence of a vasculitic process.

TREATMENT

The patient was started on a trial of prednisolone (40 mg for 2 months) and his symptoms greatly improved.

DISCUSSION

The susceptibility to immune-driven processes affecting both the gastrointestinal tract and respiratory system

Figure 2 Chest radiograph May 2011: rounded soft tissue opacity overlying the anterior end of the right third rib—not present in figure 1.

Figure 3 (A and B) Multiple nodules within the lung parenchyma with the largest nodule in the right apex (measuring 2.8 cm x 2.3 cm) on the CT scan May 2011.

Figure 4 Photomicrograph of the lung biopsy showing a bud of loose granulation tissue with an alveolar space (arrow) indicating an organising pneumonia.

could stem from the fact that they both share the same embryonic origin.

Our literature search has revealed 21 reported cases of OP as an extraintestinal manifestation of IBD.³

In terms of the association between IBD and vasculitis, seven cases of pulmonary complications resembling the appearance of Wegener's granulomatosis in UC patients have been reported.¹ The majority of these patients had a positive ANCA. The authors in this instance hypothesised that the histological changes were related to treatment with sulphasalazine or mesalazine. However, there is little information regarding the pathophysiology of this process.

From our current knowledge, ulcerative colitis has an immunological aetiology as evidenced by the fact that 50–80% of UC patients have a positive pANCA.⁴

However, the exact association between respiratory pathology, IBD, positive vasculitic screens and the role of 5-ASA compounds as a possible contributing factor is yet to be explained.

Learning points

- ▶ Always inquire about respiratory symptoms in patients with inflammatory bowel disease (IBD), though in the initial stages they may be asymptomatic.
- ▶ The spectrum of respiratory disorders occurring among patients with IBD is very broad, with bronchiectasis being the most frequently reported association.
- ▶ Pulmonary abnormalities in association with IBD can present years after the onset of bowel disease.

Competing interests None.

Patient consent Obtained.

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