Upper Lobe Fibrosis in Ulcerative Colitis

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Abstract
We report a case of ulcerative colitis where chest radiograph and high-resolution computed tomography (HRCT) of the lungs revealed bilateral upper lobe fibrosis, which was misdiagnosed and treated as pulmonary tuberculosis. The diagnosis of ulcerative colitis was confirmed by sigmoidoscopy and rectal biopsy.

INTRODUCTION
The pulmonary associations of inflammatory bowel disease (IBD) are poorly characterized despite the known systemic manifestations of IBD. This association is often overlooked as an extra-intestinal manifestation of either ulcerative colitis (UC) or Crohn’s disease (CD). Pulmonary abnormalities in UC can present years after the onset of the bowel disease and can affect any part of the lungs.1 The most prevalent and distinctive pattern of respiratory involvement in IBD is airway inflammation causing airway narrowing, although interstitial lung disease and serositis may also occur but are less common.2 The predominant clinical picture appears more commonly to be a bronchial disease rather than alveolitis.3 Early recognition of the pulmonary abnormalities in UC is important, as they can be strikingly steroid-responsive.

CASE REPORT
A 64 year old post-menopausal female housewife presented to us with an 8 years history of cough with mucoid expectoration and exertional dyspnoea. Patient had received multiple courses of empirical antituberculous treatment on the basis of symptoms and chest radiograph with no improvement in either. There was history of passing frequent stools since childhood, which had increased over the last 5 years to about 4-5 episodes per day for which she was on symptomatic treatment from her general practitioner.

On physical examination, there was pallor but no clubbing. Chest examination revealed bronchial breath sounds in the right upper lobe and bilateral coarse crackles. Hematological investigations showed anaemia but other biochemical parameters were within normal limits. Her chest radiograph (1.84 litres) and FEV1/FEV ratio of 54%. High resolution computed tomography (HRCT) of the lungs (Fig. 2a, b) revealed extensive areas of fibrosis with fibrocystic lesions in both upper lobes with scattered lesions in the superior segments of both lower lobes. Two-dimensional echocardiography (2-D Echo) revealed severe pulmonary hypertension with right atrial and right ventricular dilatation. Sigmoidoscopy revealed friable colonic mucosa with loss of vascularity and presence of granulations. Rectal biopsy (Fig. 3) showed superficial erosion of the rectal mucosa with mixed inflammation in the lamina propria consisting of polymorphs, lymphocytes, plasma cells and few eosinophils. Lymphoid aggregates were prominent, crypts showed crypt abscesses, goblet cells were well preserved, suggestive of acute phase of ulcerative colitis. Thus, based on the findings of chest radiograph, HRCT and rectal biopsy, a diagnosis of bilateral upper lobe lung fibrosis in ulcerative colitis was made. The patient was started on inhaled bronchodilators and high dose inhaled steroids along with regular chest physiotherapy for her respiratory illness and on 5-aminosalicylic acid for her ulcerative colitis.

DISCUSSION
Pulmonary involvement is a relatively uncommon extra-intestinal manifestation of ulcerative colitis (UC), it is unusual for it to be symptomatically apparent.3 It is to be recognized and treated early, as it leads to destructive and irreversible changes in the airway wall or to the “end-stage lung”.4 Pulmonary abnormality in UC can present years after the onset of the bowel disease, the shift of the inflammatory process from the bowel to the lung is because of the common ancestry from the primitive gut.3 The non-specific inflammatory changes beneath the bronchial epithelium are similar to those beneath the colonic epithelium in UC. A systemic factor is responsible for the common response at both epithelial sites in patients with UC. The lung manifestations in UC are as given in Table 1.4

The common symptoms include cough and expectoration along with breathlessness due to airway inflammation.6 Symptoms such as stridor and hoarseness occur with upper
Fig. 2a,b: High resolution computed tomography (HRCT) of the lungs showing extensive areas of fibrosis with fibrocystic lesions in both upper lobes.

Table 1: Lung manifestations in ulcerative colitis

<table>
<thead>
<tr>
<th>Site of involvement</th>
<th>Pattern</th>
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<tbody>
<tr>
<td>• Upper airway</td>
<td>Glottic/subglottic stenosis</td>
</tr>
<tr>
<td>Larynx/glottis</td>
<td></td>
</tr>
<tr>
<td>Trachea</td>
<td>Tracheal inflammation and stenosis</td>
</tr>
<tr>
<td>• Bronchi</td>
<td>Simple chronic bronchitis, Chronic bronchial suppuration, Bronchiectasis</td>
</tr>
<tr>
<td>• Small airways</td>
<td>Necrotising bronchiolitis, Diffuse pan- bronchiolitis</td>
</tr>
<tr>
<td>• Lung parenchyma</td>
<td>Bronchiolitis obliterans organizing pneumonia (BOOP), Non-specific Interstitial pneumonitis (NSIP), cellular, Pulmonary infiltrate and eosinophilia (PIE), Sterile necrobiotic Nodules</td>
</tr>
</tbody>
</table>

Table 2: Differential diagnosis of bilateral upper lobe fibrosis

- Old healed tuberculosis
- Subacute silicosis
- Chronic sarcoidosis
- Coal worker’s pneumoconiosis
- Chronic extrinsic allergic alveolitis
- Ankylosing spondylitis
- Rheumatoid disorders
- Berylliosis
- End stage histiocytosis X
- Late manifestation of radiation therapy for carcinoma breast
- Idiopathic upper lobe pulmonary fibrosis

Airway involvement, pneumothorax, pneumomediastinum or severe obstruction to airflow in cases with involvement of the smaller airways. Patients with UC, may develop various forms of interstitial lung disease following...
A high degree of suspicion is necessary to detect the disease early as it may present years after the bowel disease and the patient with airway disease may lack the classical symptoms and yield a normal chest radiograph.

**REFERENCES**