Case Report

A Rare Case of Ulcerative Colitis with Diffuse Parenchymal Lung Disease, Spontaneous Pneumomediastinum and Subcutaneous Emphysema

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Abstract

A 50-year-old male, a tobacco smoker, who was known to have ulcerative colitis presented with dry cough, chest pain, dyspnea and frequent passage of blood and mucous mixed stools. Physical examination revealed clubbing, subcutaneous emphysema of upper chest and auscultatory findings of crunching sound over pre-cordial area and basal crepitations. Spirometry was suggestive of restrictive pattern. High resolution computed tomography (HRCT) of thorax revealed pneumomediastinum, subcutaneous emphysema, bilateral diffuse centrilobular nodules and ground-glass haziness with mosaic pattern along with posterior basal fibrotic changes. The present case documents the uncommon pulmonary involvement of spontaneous pneumomediastinum and subcutaneous emphysema diffuse parenchymal lung disease, in a patient with ulcerative colitis. [Indian J Chest Dis Allied Sci 2014;56:109-111]

Key words: Ulcerative colitis, Diffuse parenchymal lung disease, Spontaneous pneumomediastinum, Subcutaneous emphysema.

Introduction

Inflammatory bowel disease (IBD) is a chronic inflammatory disease that commonly involves the gastrointestinal tract of unknown aetiology. Crohn’s disease and ulcerative colitis are the two main forms of IBD. In IBD, extra-intestinal manifestations are frequently known to occur. These include dermatological manifestations (erythema nodosum and pyoderma gangrenosum); ocular manifestations (uveitis and episcleritis); hepatobiliary manifestations (primary sclerosing cholangitis and autoimmune hepatitis); musculo-skeletal manifestations, peripheral arthritis and axial arthropathy. In contrast, pulmonary involvement is rare. The patterns of involvement in IBD are glottic/sub-glottic stenosis, chronic bronchitis, bronchiectasis, bronchiolitis obliterans, granulomatous interstitial lung disease, desquamative interstitial pneumonia, pulmonary infiltrates and eosinophilia, and sterile necrobicotic nodules, sarcoidosis, alpha-1-antitrypsin deficiency, pulmonary vasculitis, venous thromboembolism, pleural and pericardial manifestations.

We report a case of ulcerative colitis with spontaneous pneumomediastinum, subcutaneous emphysema with diffuse parenchymal lung disease, that is rare.

Case Report

A 50-year-old male presented in July 2012 with breathlessness at rest, acute substernal chest pain, dry cough, and change in character of voice of two days duration. He also complained of loose stools for the preceding two years. The patient was a tobacco smoker, alcoholic till recently and a chef by profession. In October 2010 he consulted the department of gastroenterology with complaints of bleeding per rectum, diarrhoea, and abdominal distension. Laboratory testing revealed haemoglobin 12.3 g/dL, white cell count 6000/mm³, platelet count 2.8 lakhs/mm³. Erythrocyte sedimentation rate (ESR) was 54 mm at the end of first hour; C-reactive protein (CRP) was 6.35 mg/dL. Sigmoidoscopy (Figure 1) revealed marked ulceration and loss of vascular pattern suggestive of grade IV ulcerative colitis. Histopathological examination of the biopsy specimen confirmed ulcerative colitis in active phase. He was treated with mesalamine, azathioprine, systemic corticosteroids with enemas (mesalamine enema).

Figure 1. Sigmoidoscopy showing marked ulceration and loss of vascular pattern suggestive of grade IV ulcerative colitis.

[Received: February 4, 2013; accepted: July 29, 2013]

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Heart rate was 112 beats/min, blood pressure 114/76 mmHg, oxygen saturation 78% on room air by pulse oximetry. Physical examination revealed digital clubbing, subcutaneous emphysema over at the anterior chest wall extending to the supra-scapular areas and neck. Auscultation revealed a crunching sound over the pre-cordium, synchronous with the heart beat and crepitations over the posterior basal regions. Chest radiograph (postero-anterior view; Figure 2) demonstrated multiple black luencies in soft tissue of supra-clavicular and cervical region, thin radiolucent strip along the heart borders, bilateral haziness mid and lower zones, and elevated right diaphragm. Arterial blood gas analysis revealed PH 7.35, partial pressure of oxygen in arterial blood (PaO₂) 45 mm Hg, partial pressure of arterial carbon dioxide (PaCO₂) 35 mmHg and bicarbonate (HCO₃⁻) 22 mEq/L suggestive of type I respiratory failure. Biochemical investigations including liver function tests were normal.

Serum total immunoglobulin E (IgE) levels were greater than 2000 IU/mL. Stool examination, skin prick test for common allergens and autoantibody screen detected no abnormality. Ultrasonography of the abdomen revealed hepatomegaly. Spirometry was suggestive of restrictive pattern. High resolution computed tomography (HRCT) (Figure 3) of thorax demonstrated pneumomediastinum, subcutaneous emphysema, bilateral diffuse centrilobular nodules, ground-glass haziness with mosaic pattern along with posterior basal fibrotic changes.

The patient was managed with high flow oxygen therapy, nebulised bronchodilators, antibiotics, oral prednisolone (40 mg daily), azathioprine (50 mg daily) and mesalamine (800 mg thrice-daily). There was symptomatic and radiological improvement with resolution of the pneumomediastinum and subcutaneous emphysema and bilateral mid zone shadowing after three weeks treatment (Figure 4). Stool frequency decreased, serum immunoglobulin (Ig)-E levels reduced to 450 IU/mL, ESR came down to 15 mm at the end of the first hour, CRP levels dropped down to 1.26 mg/dL. The patient was discharged on short-acting beta-agonists (SABA), mesalamine, azathioprine with tapering dose of prednisolone.

Discussion

Pulmonary involvement of inflammatory bowel disease is rare. Both the colonic and respiratory epithelia share embryonic origin from the primitive foregut and both types of epithelial cells include goblet cells and submucosal glands. The lungs and the gastrointestinal
tract contain sub-mucosal lymphoid tissue that plays crucial role in host mucosal defense. Due to similarity in the mucosal immune system, same pathogenetic changes are evident in respiratory involvement in IBD.

A variety of immunologic changes have been documented in ulcerative colitis. Cytotoxic T-cells accumulate in the lamina propria of the diseased colonic segment. This change is accompanied by an increase in the population of B cells and plasma cells, with increased production of IgG and IgE.

A small proportion of patients with ulcerative colitis have anti-smooth muscle and anti-cytoskeletal antibodies. Serum and mucosal auto-antibodies against intestinal epithelial cells may be involved. The presence of anti-neutrophil cytoplasmic antibodies (ANCA) and anti-

Saccharomyces cerevisiae antibodies (ASCA) is a well-known feature of IBD. Subcutaneous emphysema and pneumomediastinum occur frequently in critically ill patients in association with blunt or penetrating trauma, soft-tissue infections, or any condition that creates a gradient between intraalveolar and peri-vascular interstitial pressures. A continuum of fascial planes connects cervical soft tissues with the mediastinum and retroperitoneum, permitting aberrant air arising in any one of these areas to spread elsewhere. Subcutaneous emphysema and pneumomediastinum has also been described in a patient with ulcerative colitis.

In another report, occurrence of mediastinal and subcutaneous emphysema following retroperitoneal perforation in a patient with severe ulcerative colitis without toxic dilatation has been described. In another report, occurrence of pneumorrhachis, pneumomediastinum, pneumopericardium, and subcutaneous emphysema has been described after proctocolectomy for ulcerative colitis.

Two cases of ulcerative colitis in which toxic megacolon was complicated by the unusual occurrence of air tracking retroperitoneally through the diaphragm and the mediastinum, without signs of free intraperitoneal air, ultimately presenting as subcutaneous emphysema in the neck has also been reported. In a young adolescent boy with ulcerative colitis development of pneumomediastinum and subcutaneous emphysema in the neck and surrounding cervical soft tissues secondary to a probable retroperitoneal perforation of the colon has been described. Histopathological diagnosis could not be established in the present case as the patient did not consent for the procedure. Thus, the present case documents the rare occurrence of spontaneous development of pneumomediastinum and subcutaneous emphysema with diffuse parenchymal lung disease in a patient with ulcerative colitis.

References