CASE REPORT

Severe inflammatory upper airway stenosis in ulcerative colitis

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Severe inflammatory upper airway stenosis in ulcerative colitis. H. Rickli, C. Fretz, M. Hoffman, A. Walser, A. Knoblauch. ©ERS Journals Ltd 1994.

ABSTRACT: Severe upper airway stenosis was diagnosed in a 23 year old woman who presented with hoarseness, cough and dyspnoea 8 yrs after initial diagnosis of ulcerative colitis. The respiratory symptoms worsened over the next few months, the patient eventually developing dysphagia and ultimately severe upper airway obstruction. The narrowest site was the glottis, which was severely stenosed by inflammatory swellings. Systemic corticosteroids led to rapid clinical improvement and restoration of normal airway patency within a few months.

Ulcerative colitis is frequently associated with extraintestinal inflammatory manifestations. In the respiratory tract these usually take the form of chronic bronchitis, which occasionally develops into bronchiectasis. This case confirms that the inflammation can also involve the larynx and large airways.

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Ulcerative colitis is associated in up to 45% of cases with inflammation of extraintestinal organs. In a study of 202 patients, the joints (26%), skin (19%) and eyes (4%) were the most commonly affected sites [1]. A growing body of literature has recently described inflammatory pulmonary disease accompanying inflammatory bowel disease (IBD) with involvement of the lung parenchyma, airways, pleurae and pulmonary vasculature [2–9]. We report a case of severe but reversible upper airway obstruction due to pseudotumorous inflammation of the larynx, trachea and main bronchi.

Case report

History

The patient was a 23 year old nonsmoking female music student, in whom a histologically confirmed diagnosis of ulcerative colitis had been made at 15 yrs of age, following a 6 month history of persistent mucous diarrhoea. On long-term sulphasalazine, she experienced 2–3 colonic attacks each year, which required intermittent oral corticosteroid therapy. A switch from sulphasalazine to mesalazine at the beginning of 1991 produced no change in gastrointestinal symptoms.

During autumn 1991, the patient complained that her voice became easily tired and that she was occasionally hoarse. Laryngoscopy in December 1991 showed mild laryngeal swelling. In July 1992, two months after an attack of colitis was treated with oral corticosteroids, an apparent upper respiratory tract infection led to

increasing hoarseness, exertional dyspnoea, nonproductive cough and dysphagia, over a period of 5 weeks. The patient's weight decreased by 7 kg. Despite administration of various antibiotics (roxithromycin, ciprofloxacin, amoxycillin-clavulanic acid/tobramycin, doxycycline), the symptoms worsened and the erythrocyte sedimentation rate and C-reactive protein increased.

Clinical findings

On September 1 1992, the patient was afebrile and appeared generally unwell. Her voice was hoarse and there was a predominantly inspiratory stridor. The throat was reddened. A few dry crackles were audible on chest auscultation. There was no cyanosis, clubbing, skin changes or joint swelling. The abdomen was normal on palpation, with normal bowel sounds. Erythrocyte sedimentation rate was 78 mm·h⁻¹, C-reactive protein 140 mg·dl⁻¹, and white blood cell count 11.6×10⁹ cells·l⁻¹ with a normal differential. Haemoglobin was 104 g·l⁻¹. All other routine laboratory parameters were normal. Serological tests for adenovirus, influenza A and B, respiratory syncytial virus, Mycoplasma, Legionella, Chlamydia and Coxiella were negative. Antineutrophil cytoplasmic antibodies (ANCA) were negative. Chest X-ray showed marked narrowing of the tracheal air column. Magnetic resonance imaging of the upper chest and neck showed swelling of the tracheal wall (fig. 1). Lung function findings were as follows: forced vital capacity (FVC) 3.11 l (71% predicted), forced expiratory volume in one second (FEV₁) 1.57 l (41% pred), FEV₁/vital capacity (VC) 48% and total lung capacity (TLC) 5.33 l (89%

Initially, the most conspicuous endoscopic findings were marked increase in mucosal volume causing severe stenosis in the region of the vocal cords.

Varied patterns of respiratory involvement in inflammatory bowel disease have been reported, including airway disease (upper airway stenosis, bronchitis, occasionally associated with bronchiectasis, and chronic bronchiolitis), interstitial lung disease, necrotic parenchymal nodules and serositis [2]. CAMUS *et al.* [2] published the largest series with data in 33 cases, followed by HIGENBOTTAM *et al.* [4], BUTLAND *et al.* [6] and KRAFT *et al.* [8] with 10, 7 and 6 cases, respectively. More than 80% of all reported cases had ulcerative colitis, whilst the remainder suffered from Crohn's disease [2]. Respiratory involvement may be different in patients with ulcerative colitis and Crohn's disease [2]. Granulomatous involvement and oedema of the larynx, trachea and bronchi have been reported only in Crohn's disease. Conversely, chronic bronchiolitis and necrobiotic nodules have, so far, been reported only in ulcerative colitis [2]. Abnormal lung function was found in 38% of patients with ulcerative colitis and in 54% with Crohn's disease despite normal chest X-rays and lack of pulmonary symptoms [10, 11]. The pattern of ventilatory impairment was predominantly obstructive in ulcerative colitis and restrictive in Crohn's disease [10–12].

Severe upper airway inflammation has previously been mentioned in six cases, the subglottic area being mainly involved [2, 5, 13, 14]. In contrast, we found the most important site of involvement at the vocal cord level, with decreasing pseudotumoral narrowing in the lower trachea and mainstem bronchi.

In the cases so far published, including the case reported here, cough and hoarseness appeared early and were often misinterpreted. In contrast to chronic bronchial suppuration or bronchiectasis, which usually developed in patients with quiescent IBD, the majority of patients with upper airway obstruction had active IBD [2, 13, 14].

No other cause was found for the laryngotracheobronchitis in our patient. Differential diagnosis included infectious and noninfectious inflammatory processes, neoplasia and exogenous factors. Viral serology was negative, and repeated sputum tests and blood cultures revealed no evidence of bacterial infection. Various antibiotics had no effect on clinical or laboratory parameters of inflammation. The temporal association with the switch from sulphasalazine to mesalazine raised the possibility of an adverse drug effect. Acute alveolitis has recently been described in patients taking mesalazine [15], but there have been no reports of large airway involvement. Moreover, the obstruction resolved despite continued treatment with mesalazine. Isolated subglottic stenosis has been associated with several other disorders, including prior endotracheal intubation, sarcoidosis, amyloidosis, Wegener's granulomatosis, tuberculosis, midline granuloma, cutaneous pemphigus, relapsing polychondritis, lupus erythematosus, and tracheo-bronchopathia osteoplastica [16]. A series of six patients with subglottic stenosis has recently been reported in systemic, ANCA positive vasculitis [16]. ANCA was measured in 12 cases of IBD with respiratory involvement

and was found to be positive in four [2]. There was no correlation between ANCA and the pattern of respiratory involvement.

Histological examination of the tracheal mucosa showed chronic inflammation. The findings at concurrently performed colonic mucosal biopsy were consistent with moderately active ulcerative colitis. The similar pattern of inflammation suggests that the alterations of the upper airways and the colon might have been triggered by the same disease process. As HIGENBOTTAM *et al.* [4] suggested, the common embryological roots could explain the involvement of the respiratory system in some patients with IBD.

Our patient showed a good immediate and medium-term response to corticosteroids, with rapid clinical improvement and resolution of serological parameters of inflammation. The obstructive ventilatory impairment proved fully reversible. The response to corticosteroids has been mentioned by several authors [1–7, 17], with a dramatic effect following methylprednisolone in some cases. Laser beam ablation has been proposed for patients with life-threatening airway obstruction, and those with disease refractory to corticosteroids [2].

In our case, the medium-term outcome was good, the patient remaining in remission 8 months after diagnosis, including 1 month without oral corticosteroids. In four of the six previously reported cases of subglottic stenosis, the outcome was also favourable, the patients reporting few or no symptoms [2, 15]. One patient died after an attempt at tracheal dilatation [5], whilst another died of endotoxin shock related to colitis [16].

Our report supports the contention that severe inflammatory upper airway stenosis is part of the spectrum of IBD-related respiratory manifestations.

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