Oesophago-bronchial fistula causing cough and recurrent lower respiratory tract infections in a patient with Crohn's disease

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INTRODUCTION

Crohn's disease of the oesophagus is a recognized entity with associated significant morbidity and mortality when complicated by fistula formation. The authors report a case of recurrent respiratory tract infections in a Crohn's disease patient with an oesophago-bronchial fistula demonstrated by barium swallow and endoscopy. The authors also review other cases in the literature and discuss the treatment options of this uncommon, but potentially serious complication of Crohn's disease.

DISCUSSION

Crohn's disease is a chronic granulomatous disease characterized by segmental transmural inflammation of the alimentary tract between the mouth and anus, with the majority of cases confined to the terminal ileum and colon. Involvement of the upper gastrointestinal (GI) tract and in particular the oesophagus is uncommon but, when it does occur, there is nearly always concomitant disease in the small bowel or colon (Dancygier and Frick, 1992). In a study by Lenaerts et al (1989) of 230 cases of Crohn's disease only 15 (6%) had oesophageal involvement demonstrated by endoscopy and barium studies.

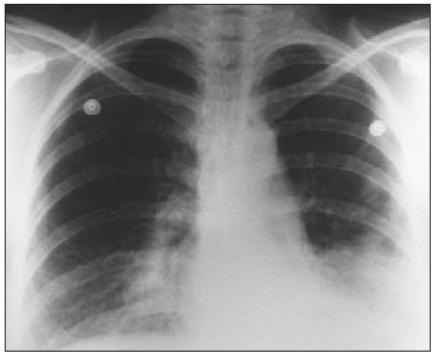

Figure 1. Postero-anterior chest radiograph demonstrating left lower lobe consolidation

In oesophageal Crohn's disease the presenting symptoms are dysphagia, odynophagia, or substernal chest pain (Haeremai et al 1982; Steel et al, 1988). The activity of inflammation usually parallels activity of the disease elsewhere in the GI tract, with isolated involvement of the oesophagus described in only a few cases (Gore and Ghahremani, 1986). The endoscopic hallmarks include hyperaemia, granularity, nodular thickening of the distal oesophageal folds and aphthous ulceration (Dancygier and Frick, 1992).

Geboes et al (1986) endoscoped and biopsied 107 patients with Crohn's disease with upper GI symptoms and diagnostic histological changes of the disease were seen in only 9 cases (1.8%). This is a result of superficial biopsies of a transmural disease and skip lesions (Dancygier and Frick,

CASE REPORT

A 40-year-old non-smoking woman was referred to the outpatient clinic because of an increasing dry cough and recurrent lower respiratory tract infections over a 10-year period. The patient was diagnosed with Crohn's disease in 1979. The disease was initially confined to the large intestine but despite intensive medical treatment she required a total colectomy with ileostomy formation and, 2 years later, she was treated with a proctectomy.

In 1996 she developed a problematic nocturnal cough with associated early morning sputum expectoration and intermittent wheeze, which did not respond to a trial of inhaled bronchodilators. In 1998 she was admitted to hospital with a severe left lower lobe pneumonia, which responded well to oral ampicillin and erythromycin. She subsequently complained of coughing during meals but denied any indigestion or heartburn. There was no background of atopy, or history of asthma or rhinitis.

Systemic examination was unremarkable. Review of previous chest radiographs revealed recurrent changes of left lower lobe consolidation (*Figure 1*). Initial investigations including spirometry, biochemical screen, full blood count, immunoglobulin subclasses, aspergillus precipitins and sputum culture for bacteriology and tuberculosis were normal.

An initial diagnosis of minor localized bronchiectasis was made and the patient was referred to the physiotherapy department for postural drainage. Because of the persistent nocturnal cough, gastro-oesophageal reflux with aspiration was considered and a barium swallow arranged.

The barium swallow (*Figure 2*) demonstrated a fistula between the oesophagus and the left lower lobe bronchus arising approximately 10 cm from the gastro-oesophageal junction. Multiple 'rose-thorn' ulcers were also seen in the mid- and lower oesophagus. The barium, which passed into the left lower bronchial tree, did not demonstrate bronchiectatic changes (*Figure 3*). This was confirmed with high resolution computer tomography, which revealed some localized fibrosis in the left lower lobe, but did not show any evidence of bronchiectasis.

At endoscopy a 4 mm defect was seen at 32 cm, which was biopsied. The histology showed hyperplastic squamous epithelium with inflammatory changes but no features of malignancy.

The patient gained considerable relief of her symptoms from the use of antacid suspensions and propping the head of her bed at night. She discontinued the proton pump inhibitor owing to side effects, but has remained relatively symptom free during the last year. She has consistently refused enteral feeding and surgical intervention despite being aware of the potential risks of complications from the oesophago-bronchial fistula.

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1992), whereas in resected specimens the chronic inflammatory cell infiltrate and giant cells are seen to extend through all layers of the oesophagus (Cynn et al, 1975).

Because of the transmural nature of the disease, fistulas to other organs can occur but oesophago-bronchial fistulas are rare. The authors are aware of only nine reported cases in the literature (Achenbach et al, 1956; Cynn et al, 1975; Haeremai et al, 1982; Davidson and Sawyer, 1983; Steel et al, 1988; Mathis et al, 1994; Malchow et al, 1996; Rholl et al, 1998; Ho et al, 2002). The most common site of fistula communication has been intrapulmonary, with the bronchial tree affected in five cases (Achenbach et al, 1956; Haeremai et al, 1982; Steel et al, 1988; Ho et al, 2002) and the trachea in one (Malchow et al, 1996). In two cases the tracts remained intramural (Cynn et al, 1975, Davidson and Sawyer, 1983) within the body of the oesophagus, with the final case describing an oesophago-gastric fistula (Cynn et al, 1975). Of the nine cases of fistula formation in the oesophagus, only one demonstrated giant cells, with the rest showing inflammatory cells only (Achenbach et al, 1956).

Isolated oesophageal Crohn's disease is very rare and other granulomatous conditions such as sarcoidosis, fungal disease and tuberculosis have to be considered. The presence of Crohn's disease at other sites in the GI tract helps support the diagnosis of Crohn's disease of the oesophagus when suspected. In this patient, although the endoscopic biopsies showed no specific features of active Crohn's, the radiology proved diagnostic with the demonstration of fistulation and adjacent deep ulceration in the mid and lower oesophagus in the form of 'rose-thorn ulcers'. It would be reasonable to assume that she developed the oesophago-bronchial fistula around the time of extensive fistulae formation in the lower GI tract and in the absence of immunosuppression it is postulated that the fistula was responsible for her recurrent respiratory tract infections.

Management of fistulas in Crohn's disease is difficult and is tailored to the severity and location. Therapeutic options include steroids, antibiotics for local perianal disease, immunomodulators, such as azathioprine or 6-mercaptopurine, anti-tumour necrosis factor alpha (TNF-alpha) agents (infliximab), enteral feeding and surgery. However, because of the infrequency of oesophageal involvement in Crohn's disease and the limited data on oesophageal fistula, there is a lack of evidenced-based knowledge with respect to the best treatment available.

The patient in this report is unusual in that she has remained extremely well without any specific therapy over a period of 5 years, and has had no significant respiratory infections since the fistula was demonstrated. This raises the possibility of spontaneous closure, although the patient has declined further investigations to explore this possibility.

Figure 2. Single contrast barium swallow study demonstrating an oesophago-bronchial fistula (white arrow) with multiple 'rose-thorn ulcers' characteristic of Crohn's disease in the distal oesophagus (black arrows).

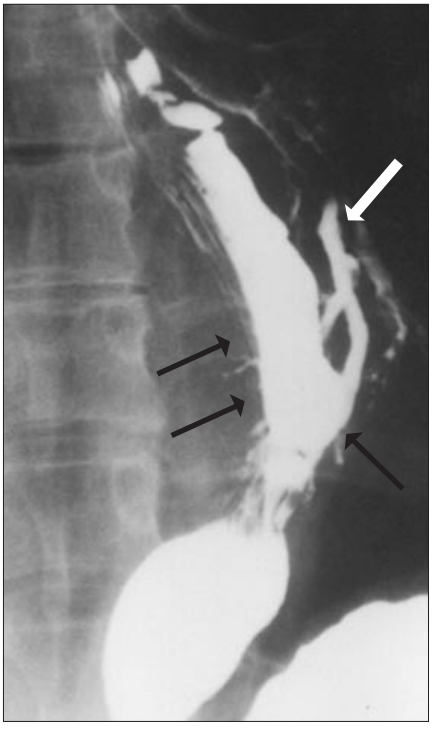
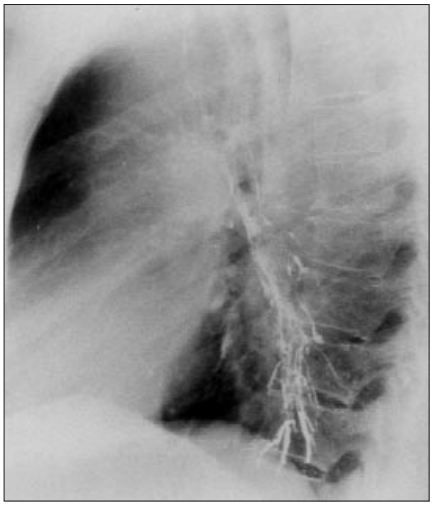


Figure 3. Left lateral chest radiograph demonstrating the presence of barium in the left lower bronchial tree.



CONCLUSION

Involvement of the oesophagus in Crohn's disease is much less common than the remainder of the GI tract and, although oesophageal-bronchial fistula in Crohn's disease is extremely rare, it should be suspected in patients with persistent respiratory symptoms. Non–surgical treatment of fistulation in the past has largely been unsuccessful, and the role of anti TNF-alpha agents is as yet unclear. **HM**

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