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Idiopathic Bronchiectasis: A Case-Based Review of Modern Management

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Case Report

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ABSTRACT

While bronchiectasis was first described more than two centuries ago, it is only in the last decade that there have been major advances in our understanding of the condition and in its treatment. Recent evidence has shown that the "vicious cycle hypothesis" of airway inflammation and damage remains valid. The concept of "treatable traits" provides a useful framework on which to base the management of this condition and the co-morbidities that need to be actively managed. In all patients recently diagnosed, a search should be undertaken for specific aetiologies. A number of factors including chronic infection by *Pseudomonas aeruginosa*, have been shown to be associated with increased morbidity and mortality. While airway clearance strategies remain the mainstay of therapy, other treatments including nebulised hypertonic saline and inhaled antibiotic treatment are of demonstrable benefit. These treatments are discussed in the context of a specific case.

INTRODUCTION

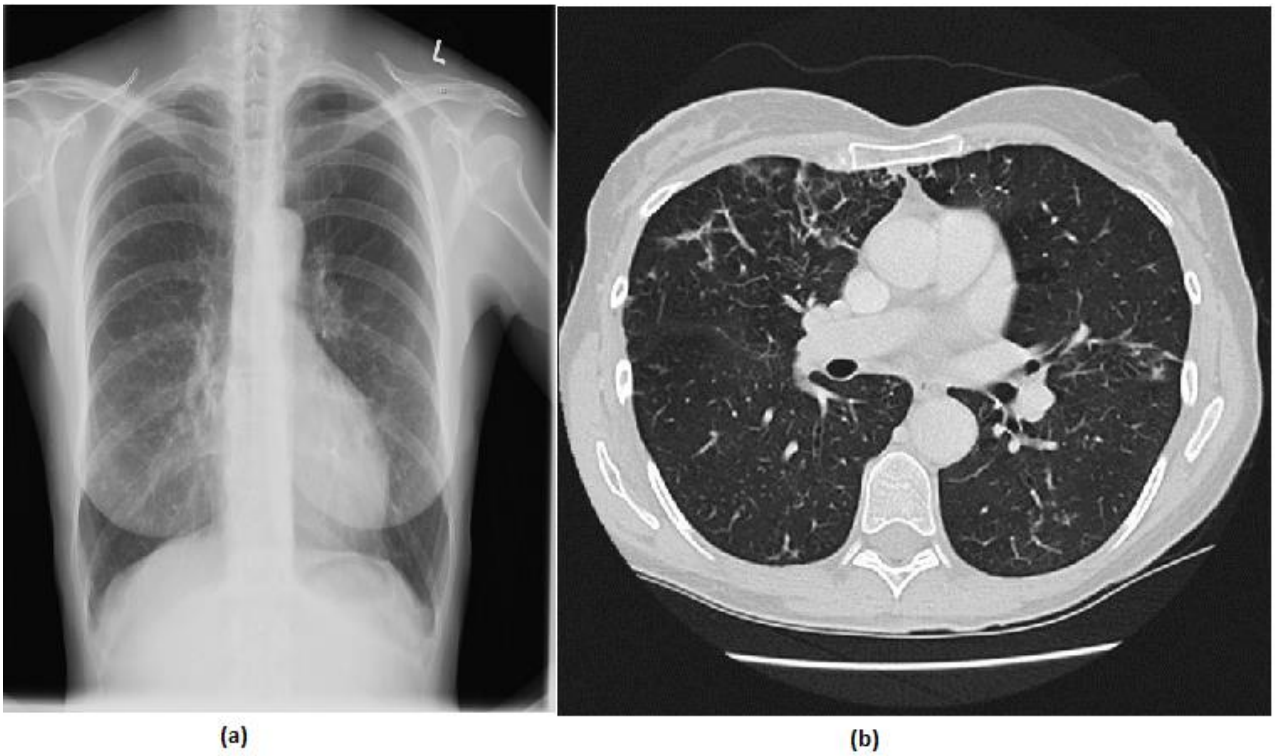
Some aspects of the modern management of idiopathic bronchiectasis will be discussed in the context of a specific case. The discussion is not intended as a comprehensive review of bronchiectasis and its management; for this the reader is directed to other relevant publication [1-4].

CASE PRESENTATION

A 65 year old European woman, BMI=18, presented with a chronic cough productive of 20 ml of muco-purulent sputum per day. Her cough and sputum production had worsened in the last few months. She had not had haemoptysis. Her appetite was reduced and she had lost 4 kg in the last year. She had never been hospitalised for bronchiectasis but had required two courses of oral antibiotics per year for episodes of increased cough and sputum. She was a life-long non-smoker. She had a history of rhino-sinusitis but no symptoms of gastro-oesophageal reflux or other co-morbidities. Clinically she was not clubbed and on auscultation of the chest there were scattered inspiratory medium crackles on the right.

Her initial chest radiograph and an image from HRCT scan of the chest are shown in Figure 1. The HRCT scan showed bronchiectasis particularly involving the right middle lobe and lingula of the left upper lobe. Levels of immunoglobulins G, A and M were normal, Aspergillus serology was negative and sweat chloride and nasal NO were within normal ranges. Culture of sputum grew *Pseudomonas aeruginosa*. Sputum was smear negative but culture positive for *Mycobacterium avium* (MAIC). Spirometry revealed an FEV1 of 2.70l and FVC of 3.54l; 100% and 112% predicted.

Figure 1. (a) Chest radiograph and (b) HRCT scan, at presentation.



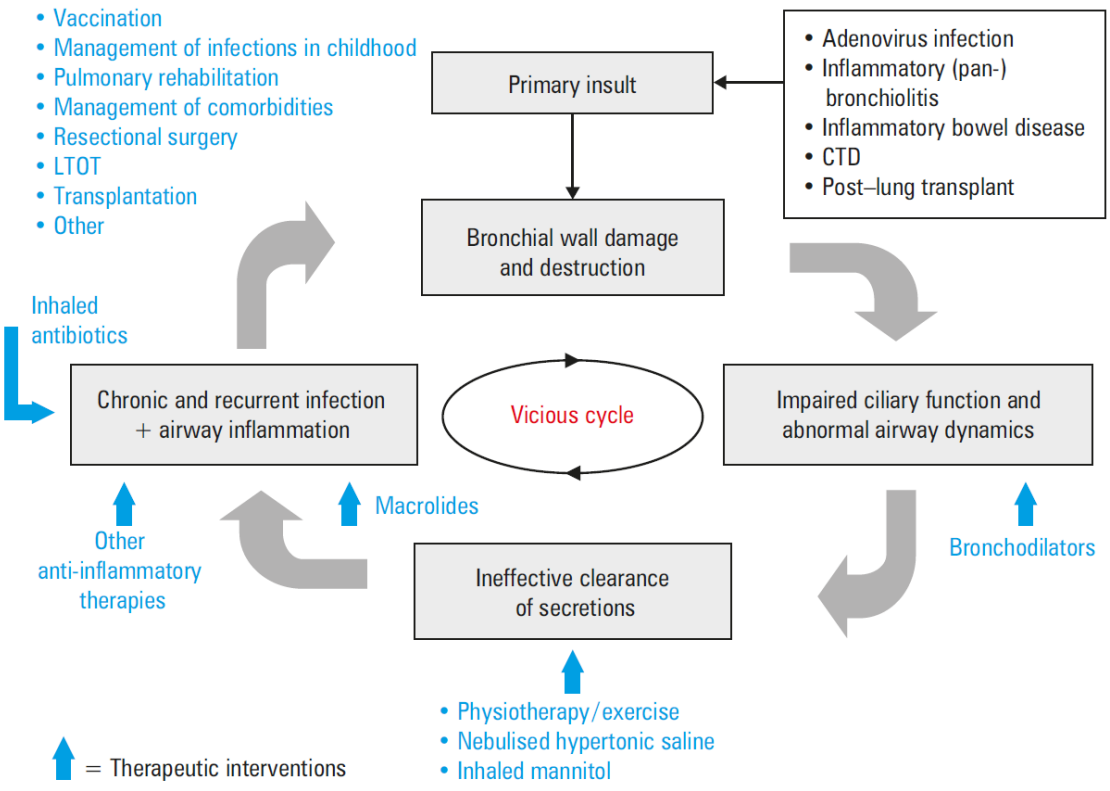
RESULTS AND DISCUSSION

Although bronchiectasis has been recognised for more than two centuries, it is only in the last decade that there have been major advances in our understanding of the disease and its treatment [1-4,6].

Nowadays the diagnosis is conformed radiologically: specifically on HRCT scan of the chest with the most common criteria being the luminal diameter of the airway being greater than the diameter of the accompanying branch of the pulmonary artery (B:A ration >1). However it is important to remember that bronchiectasis is a clinical syndrome and not merely a radiologic diagnosis; many patients with other forms of airways disease (COPD and chronic asthma) may have radiologic evidence of bronchial dilatation but do not have the symptoms of chronic cough and sputum production.

Recent evidence indicates that the "vicious cycle" hypothesis of airway wall damage, impaired clearance of secretions, chronic infection and inflammation and worsening pulmonary damage, proposed by Cole many decades ago [5], remains valid (See Figure 2).

Figure 2. The "vicious cycle of bronchiectasis and modern therapeutic interventions (reproduced from Reference 6 with permission).



As with other airway diseases, the concept of "treatable traits" has been applied to the management of bronchiectasis; these have been categorised as pulmonary (manifestations), aetiological (factors), extra-pulmonary manifestations, and environmental/lifestyle factors [3]. While this approach merely formalises the approach to

management taken by many physicians, it does provide a worthwhile framework to approach the management of this condition.

While most cases of bronchiectasis are considered to be the sequelae of childhood infections (such as adenovirus bronchiolitis), a specific aetiology is generally not confirmed and the disease is labelled “idiopathic”. Recommended investigations, to diagnose a specific cause for the disease, include quantitative immunoglobulin levels (IgA, G and M, but not IgG sub-classes), sweat chloride (to identify cystic fibrosis), nasal nitric oxide (as a screen for primary lung dyskinesia) and Aspergillus serology (for allergic broncho-pulmonary Aspergillosis) [1]. Bronchiectasis may be a feature of certain connective tissue diseases (particularly rheumatoid disease) and inflammatory bowel disease (ulcerative colitis but also Crohn’sdisease).

This patient had the comorbidity of chronic rhino-sinusitus which was specifically managed. This condition have contributed to her symptoms of cough and sputum, and may aggravate her lower respiratory tract disease. She had no symptoms of gastro-oesophageal reflux; upper gastro-intestinal disease may aggravate lower respiratory tract disease either directly by micro-aspiration or indirectly via vagal reflexes. With input from a dietician and the provision of nutritional supplements, her nutritional status was improved.

Her pulmonary manifestations are outlined in the case history above and the range of potential therapeutic interventions are illustrated in Figure 2. Bronchiectasis is associated with abnormal mucus and impaired muco-ciliary clearance. In patients with chronic sputum production, chest physiotherapy/airway clearance strategies remain the mainstay of management, although a variety of physiotherapy techniques and devices exist [6,7]. Inhalation of nebulised hypertonic (6%-7%) saline is a cheap, well-tolerated therapy that improves the physical properties of sputum and “may improve quality of life, outcomes and sputum clearance in individuals with bronchiectasis” [1,8,9].

This patient received instruction from a respiratory physiotherapist regarding airway clearance strategies to be performed on a daily basis and was also started on nebulised hypertonic saline.

Chronic *Pseudomonas aeruginosa* infection is associated with an increased exacerbation rate, poorer quality of life, greater impairment of pulmonary function and increased mortality. For those reasons, if this was the first culture of Pseudomonas it would be reasonable to attempt eradication with two weeks of oral ciprofloxacin, as the first of several alternatives [1]. Randomised controlled trials of chronic suppressive inhaled antibiotic therapy for chronic Pseudomonas infection have been disappointing overall. One trial with a positive result in terms of microbiological and patient-centered outcomes, used the intravenous form of gentamicin delivered by nebuliser [10]. This treatment is not indicated routinely but could be considered in this patient if they had continued morbidity such as frequent requirements for oral or intravenous antibiotics.

Although three large randomised controlled trials long term macrolide therapy in bronchiectasis, have shown benefit in terms of reduced rate of exacerbation and other outcomes, such therapy is contraindicated in this patient because of the culture of a Non-Tuberculous Mycobacterium (NTM) [11-13].

NTM infection is increasingly recognised in patients with bronchiectasis, and this patient fits a particular NTM phenotype known as Lady Windermere Syndrome; typically thin, postmenopausal European females with

bronchiectasis in the right middle lobe and/or lingual [14,15]. This patient had no radiologic evidence of pulmo-cavitatory NTM disease, was smear negative and there were no indications for specific anti-mycobacterial therapy. Such treatment is prolonged, associated with adverse effects and variably effective [15]. As such it is a clinical decision not to be entered into lightly. In most cases, including this patient, the infection can be managed with airway clearance (and nebulised hypertonic saline) [16,17]. Although commonly used, there is no evidence for a benefit with inhaled corticosteroids in idiopathic bronchiectasis, and with use of such treatment the risk of NTM infection is increased.

CONCLUSION

The management of bronchiectasis was discussed in relation to a specific patient; this included investigations to determine a specific etiology, the management of comorbidities, the importance of airway clearance and other strategies to improve muco-ciliary clearance including the use of nebulised hypertonic saline and the use of nebulised anti-Pseudomonal antibiotics.

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