Case reports

A commentary on the following case reports appears on pp 812–3.

Three cases of pulmonary aspergilloma in adult patients with cystic fibrosis

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Abstract

Pulmonary aspergillomas usually occur when Aspergillus fungi colonise lung tissue previously damaged by disease. Pulmonary aspergillomas in three adult patients with cystic fibrosis are reported— an association not previously described. At the time of diagnosis all three patients had previous long colonisation with Aspergillus fumigatus and severe advanced destructive lung disease with lung function less than 25% of the predicted normal values. It is likely that, with increasing survival in cystic fibrosis, more adult patients will develop aspergillomas during the protracted phase of end stage lung disease that characterises the terminal years of this condition. (Thorax 1995;50:805–806)

Keywords: Aspergillus, cystic fibrosis, aspergillosis.

Evidence of sensitisation to the fungus Aspergillus fumigatus is common in patients with cystic fibrosis. We describe here three adult patients who developed pulmonary aspergillomas many years after becoming sensitised to A fumigatus, an association to our knowledge not previously described. All three patients had very poor lung function at the time of the diagnosis of an aspergilloma and two of the three patients have since died.

Case reports

PATIENT 1

MB, a woman, was diagnosed with cystic fibrosis at 12 years of age by a positive sweat test. In 1968, at the age of 14, she presented with haemoptysis at which time sputum culture grew Staphylococcus aureus. Chest radiography showed nodular cystic changes consistent with cystic fibrosis. Contrast bronchography revealed proximal bronchiectasis in both upper lobes. She presented again in 1986 with wheeze, dyspnoea, purulent sputum, and pulmonary infiltrates. She had positive skin prick test to extracts of A fumigatus, peripheral eosinophilia, and sputum examination revealed fungal hyphae. The total IgE antibody titre was raised at 275 kU/l (0–120 kU/l), and tests for specific IgE antibodies to A fumigatus measured by the radioallergosorbent method (RAST) were positive as were serum IgG antibodies to A fumigatus detected by enzyme linked immunosorbent assay (ELISA). A diagnosis of allergic bronchopulmonary aspergillosis was made and she was commenced on inhaled and systemic steroids. In 1991 her respiratory status deteriorated and chest radiographs now revealed a newly acquired rounded 5 cm diameter lesion in the right upper lobe characteristic of an aspergilloma. A computed tomographic (CT) scan of the thorax confirmed the presence of an aspergilloma in the right upper lobe (fig 1). Her general condition continued to deteriorate and she died in May 1992 at the age of 38 years.

PATIENT 2

ML, a man, was diagnosed with cystic fibrosis by a positive sweat test in 1972 at the age of four. In May 1990 he presented with a lower respiratory tract infection. Sputum culture grew Pseudomonas aeruginosa and A fumigatus. Chest radiography showed extensive radiological changes of cystic fibrosis with proximal bronchiectasis, severe emphysema, and a widespread nodular cystic infiltrate. The total IgE was raised at 266 kU/l (0–120 kU/l) and the peripheral eosinophil count was raised at 600 cells/mm³. Serum IgG antibodies to A fumigatus (ELISA) were strongly positive. However, skin prick test and specific IgE antibody titres to extracts of A fumigatus (RAST) were negative. He required frequent admissions for respiratory tract infections between 1990 and 1993, when he developed respiratory failure. During this terminal phase a clear cut aspergilloma was seen.
Figure 2. Plain chest radiograph showing an aspergilloma in the right upper lobe of patient 2.

in the right upper lobe of a chest radiograph (fig 2). His condition deteriorated rapidly and he died shortly afterwards at the age of 25 years.

PATIENT 3
BH, a man, was diagnosed with cystic fibrosis in 1979 at the age of nine. In 1987 he developed a recurrent right sided pneumothorax requiring intercostal drainage. During that admission his chest radiograph showed extensive chronic left lower lobe atelectasis. Bronchoscopy was performed revealing copious purulent secretions on the left side. These were aspirated with a subsequent partial radiological improvement. Sputum culture grew Staphylococcus aureus and Haemophilus influenzae species. He was next admitted in February 1993 with a six month history of progressive dyspnoea. Chest radiography showed complete atelectasis of his entire left lung with compensatory hyperexpansion of his right lung. Two aspergillomas were present in the atelectatic left lung. Bronchoscopy was performed and a large volume of purulent secretions was suctioned from the left side. Sputum culture grew Pseudomonas aeruginosa and Aspergillus fumigatus. Serum specific IgG antibody titre to A fumigatus (ELISA) was significantly raised. The specific IgE antibody titre to A fumigatus (RAST) was also positive. The peripheral eosinophil count was normal. He recovered sufficiently to be discharged home but his respiratory status remains poor in April 1994 at the age of 24 years.

Discussion
This report documents for the first time the occurrence of pulmonary aspergilloma in three patients with end stage pulmonary disease due to cystic fibrosis. This extends the spectrum of chronic fibrotic conditions which may predispose to aspergilloma formation, such as tuberculosis, sarcoidosis, pneumocystis, and ankylosing spondylitis.1 It is surprising that such an association with cystic fibrosis has not been recorded previously. As many as 60% of patients with cystic fibrosis may have positive sputum cultures for fungi2 and precipitating antibodies to A fumigatus may be detected in 30% of patients.3 Furthermore, a firm diagnosis of the clinical-radiological syndrome of allergic bronchopulmonary aspergillosis has been observed in about 10% of patients with cystic fibrosis.4 Despite this, we have found no previous reports of pulmonary aspergilloma developing in this group of patients.

Cystic fibrosis is a condition in which secretion of abnormal mucus results in plugging of the distal airways leading to chronic pulmonary infection and destruction of lung tissue. The presence of necrotic tissue and lung cysts provides an ideal environment for the colonisation of the airways by A fumigatus and the development of related conditions such as allergic bronchopulmonary aspergillosis and aspergilloma. This colonisation may also be facilitated by the frequent use of both oral and inhaled corticosteroids which may be required to control symptoms in these patients, by the advent of diabetes mellitus, and by the associated poor nutritional status common to this disease.

The development of an aspergilloma represents an additional potential hazard for patients with cystic fibrosis. Although many patients may for a time remain unaffected by aspergilloma, there is clearly potential for developing severe haemoptysis which may require emergency measures such as bronchial artery embolisation. Furthermore, the presence of an aspergilloma may be a significant impediment for patients being considered for lung transplantation. The possibility of effective treatment is unfortunately limited. Patients with end stage cystic fibrosis are unlikely to be considered for surgical intervention. While the results of both intracavitary instillation and systemic treatment with antifungal agents has been disappointing,5 these measures may need to be evaluated for efficacy in the unique setting of cystic fibrosis.

In conclusion, as the median age of survival of patients with cystic fibrosis rises and with the increased use of CT scanning for clinical assessment, it is likely that the diagnosis of pulmonary aspergilloma will be made with greater frequency in adult patients with cystic fibrosis.

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