

years the patient has had frequent admissions to hospital with pulmonary problems but is in a stable condition.

Discussion

Cystic fibrosis is an autosomal recessive genetic disorder. Recently the cystic fibrosis locus has been located on chromosome 7 and the nucleotide base sequence of the gene has been determined. The most frequently reported mutation is delta F508.¹ Cystic fibrosis includes chronic obstructive and suppurative lung disease, exocrine pancreatic deficiency and abnormalities of the liver and of the reproductive tract.^{2,3} The diagnosis is based on the detection of increased concentrations of electrolytes in the sweat.⁴ There is often a family history of cystic fibrosis. Expression of the disease varies and there is a little relation between age at diagnosis and age at death. A few elderly patients with cystic fibrosis have been reported,⁵⁻⁸ but our patient is probably the oldest to be diagnosed and reported. This case shows that in patients with appropriate symptoms cystic fibrosis should be considered

whatever their age. A mild clinical course may delay diagnosis and may be related to heterozygosity.

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Removal of endobronchial mucormycosis lesion through a rigid bronchoscope

Sulaiman Al-Majed, Feisal Al-Kassimi, Mohamad Ashour, Mohamad Osman Mekki, Saleem Sadiq

Abstract

A 44 year old man with poorly controlled diabetes mellitus developed endobronchial mucormycosis, which totally obstructed the right lower lobe bronchus. The lesion was removed through a rigid bronchoscope. Two weeks later the bronchus was free of mucormycosis histologically and on culture.

Pulmonary mucormycosis is a rare fungal infection that occurs primarily in patients with poorly controlled diabetes.^{1,2} Acidosis appears to enhance fungal growth.³ *Rhizomucor*, *Rhizopus*, and *Absidia* are the commonly encountered genera. All the organisms have a predilection for invading blood vessels and causing thrombosis and infarction,⁴ thus explaining the high incidence of haemoptysis.

Endobronchial mucormycosis is well des-

cribed and is known to cause severe or fatal haemoptysis and asphyxiation.⁵ Early aggressive surgical resection of airway mucormycosis is therefore recommended.^{5,6}

We report a patient in whom endobronchial mucormycosis was removed endoscopically.

Case report

A 44 year old man was admitted to King Khalid University Hospital, Riyadh, with fever, sweating, cough with grey sputum, and non-pleuritic chest pain of a week's duration. He had had diabetes for 10 years. His diabetes had been poorly controlled since he stopped his regular insulin injections three months before admission. On examination his temperature was 38.8°C and respiratory rate 22/min. There was dullness to percussion in the right lower zone, with bronchial breathing and crackles. The chest radiograph showed shadowing with loss of volume in the right lower lobe. Pneumonia was diagnosed. Plasma glucose was 32.7 mmol/l; there was no ketonuria. Arterial blood gases, with the patient breathing air, were normal. No pathogens were cultured from his sputum and no acid fast bacilli seen. The Mantoux test response to 10 units purified protein derivative was negative.

Treatment was started with intravenous erythromycin and insulin and chest physiotherapy, with no improvement. Repeat chest radiographs showed worsening of the lower lobe shadowing. Fibreoptic bronchoscopy showed a white, cheese like mass occluding the lumen of the right lower lobe bronchus distal to the origin of the apical segment. Multiple

Department of
Medicine
S Al-Majed
F Al-Kassimi
M O Mekki

Department of
Pathology
S Sadiq

Department of
Surgery,
College of Medicine,
King Saud University,
PO Box 2925,
Riyadh, 11461,
Saudi Arabia
M Ashour

Reprint requests to:
Dr Al-Majed

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Section of bronchial wall showing invasion by fungal elements. (Haematoxylin and eosin.)



biopsy specimens from the mass and bronchial mucosa showed granulation tissue with inflammatory cells and abundant non-septate hyphae. Ziehl-Neilson staining was negative. Biopsy tissue grew *Rhizopus* sp on fungal culture. The same fungus was grown from bronchial washings.

Treatment with intravenous amphotericin B was started but had to be withdrawn after a week because of deteriorating renal function. At repeat bronchoscopy (with a rigid bronchoscope) the lesion was unchanged. The mass obstructing the right lower lobe was grasped by the biopsy forceps and removed, which produced a round slough of bronchial wall $2 \times 0.6 \times 0.5$ cm. The mucosa of the lower lobe bronchi was inflamed and oedematous but there were no other localised lesions. Histological sections of the mass showed necrotic bronchial mucosa and fragments of hyaline cartilage, compatible with a partial thickness slough of the bronchial wall.

Histologically, the sloughed tissue and biopsy specimens of mucosa were heavily invaded by non-septate hyphae (figure). Repeat fungal cultures were again positive for *Rhizopus*.

Two weeks later a third set of biopsy specimens taken from the site of the previous mass at the right lower lobe showed normal mucosa free of fungus histologically and on culture. The chest radiograph became normal. Nevertheless, amphotericin B was resumed and a total of 1.2 g was given over 60 days. Repeat bronchoscopy at four and six months showed no abnormalities and his chest radiograph remained normal. No fungus was found on a subsequent sputum culture. The patient was well at follow up after one year.

Discussion

Pulmonary mucormycosis occurs in two forms. The more common form produces parenchymal disease with consolidation of the lung and a rapidly progressive clinical course leading to respiratory failure. The other form, endobronchial disease, affects the large airways predominantly.⁶ This form often presents insidiously but it may have devastating consequences when it causes acute upper airways obstruction⁷ or fatal haemoptysis resulting from infarction of the airways.^{6,8} Mucormycosis in both forms has a high mortality; about half the patients reported have died from massive haemoptysis.⁵ The parenchymal form has the worse prognosis, with a fatal outcome in most cases.^{9,10}

It is generally accepted that surgical resection of the infected part and surrounding devitalised tissues should be performed whenever possible.⁵⁻⁷ This view is strengthened by the fact that amphotericin B does not appear to penetrate to the bronchus effectively.¹¹ Of the six reported patients with endobronchial mucormycosis who have survived, only two were cured by amphotericin B⁵; the others had surgical resection with or without amphotericin B.

In our case, the airway mucormycosis was removed through a rigid bronchoscope. Although the relative contributions of the procedure and the subsequent amphotericin B cannot be determined, the fact that the many biopsy specimens taken after the procedure showed no hyphae and grew no fungi suggests that the endobronchial removal played a large part in the successful outcome.

In view of the high incidence of massive haemoptysis associated with these lesions, the treatment of choice should be surgical resection in conjunction with amphotericin B. Bronchoscopic removal of endobronchial mucormycosis should be considered in patients not fit for surgery. As the procedure could trigger massive haemoptysis, facilities for surgical intervention should be available on site.

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