Thorax 1985;**40**:952–953

Short reports

A case of alveolar microlithiasis: observation over 22 years and recovery of material by lavage

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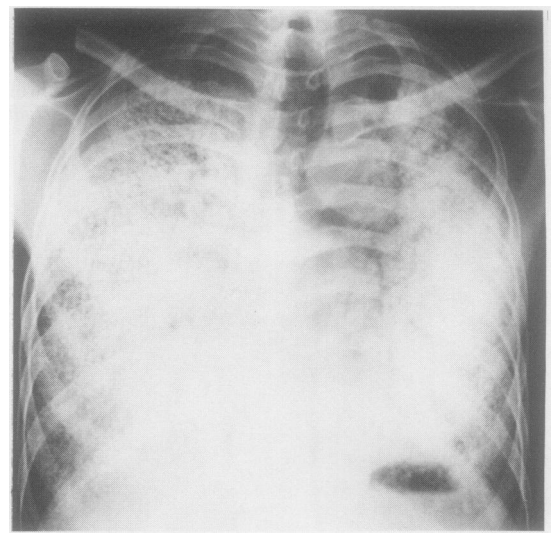
About 150 cases of alveolar microlithiasis have so far appeared in published papers. Although some patients present with progressive respiratory symptoms, the diagnosis is usually made as an incidental radiographic finding.[1] The management of the condition has been merely supportive in all reported cases. We describe the natural history of the disorder in an unusually well documented case that was first reported in 1962,[2] and discuss the possible role of bronchoalveolar lavage in its management. To compare progression in this case with previous reported experience a review of the published cases has been undertaken.

Case report

In 1962 a 13 year old Pakistani boy was attending a tuberculosis contact clinic when a chest radiograph showed the typical appearance of alveolar microlithiasis. He was at that time symptomless and there was no evidence of pulmonary tuberculosis. He was lost to follow up until 1978, when further review found him to be still symptomless, although there was some increased shadowing on the chest radiograph.

He was seen again in December 1982, having spent the intervening period in Pakistan. On this occasion he complained of increasing dyspnoea and a productive cough. On examination in 1982 there was considerable finger clubbing and he was cyanosed and dyspnoeic at rest, but examination of the chest showed nothing remarkable. A loud pulmonary second sound and right ventricular heave indicated pulmonary hypertension. The chest radiograph (figure) showed a further increase in pulmonary shadowing with bilateral apical pneumothoraces. Other investigations showed arterial hypoxaemia (Pao_2 5.9 kPa (44 mm Hg)), right ventricular hypertrophy, and strain pattern on the electrocardiogram, and bilateral pleural thickening and high density deposits in both lungs on computed tomography of the thorax. No evidence of abnormal calcium metabolism was found. The deterioration in the patient's pulmonary function over 22 years is shown in the table.

During February and March 1983 three bronchoalveolar lavages were undertaken under general anaesthesia according a method described in the management of alveolar proteinosis.[3] With the use of a total of 22 litres of buffered saline lavage fluid, 14.5 g of solids were removed from the lungs. The material recovered, which was readily visible to the naked eye, had the appearance of fine sand. Spectrophotometry showed the material to be composed of mainly calcium and phosphorous, with a smaller amount of magnesium.



Chest radiograph, 1982.

Unfortunately the patient's condition did not improve after bronchoalveolar lavage. He became increasingly dyspnoeic, with increasing arterial hypoxaemia. In May 1983 arrangements were made for him to be considered for a heart-lung transplant but before the full assessment for this procedure could be undertaken he developed a large left sided pneumothorax. Full reinflation of the lung could not be obtained despite repeated tube drainage, and he died in June 1983. Postmortem examination showed typical changes of alveolar microlithiasis with complicating cor pulmonale.

Changes in lung function test results over 22 years (the percentages of predicted normal values in parentheses)

	1962	*1978*	*1983*
Age (y)	13	29	34
Height (cm)	140	166	166
Vital capacity (l)	1.63 (48)	2.14 (49)	1.3 (32)
Total lung capacity (l)	2.16 (60)	2.37 (40)	2.07 (35)
Forced residual capacity (l)	1.31 (49)	1.27 (41)	1.32 (43)
Residual volume (l)	0.56 (95)	0.23 (14)	0.72 (44)
FEV_1 (l)	1.57 (51)	2.14 (61)	1.31 (39)
Forced vital capacity (l)	1.63 (60)	2.19 (50)	1.34 (32)

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Accepted 13 May 1985

Discussion

An accurate assessment of the natural history of alveolar microlithiasis is difficult, especially as its cause is unknown and in many cases the diagnosis is made as an incidental radiographic finding in symptomless patients. There seems little doubt, however, from the reported cases that the disorder is progressive, both radiologically and clinically. This patient had a long asymptomatic phase but then a slow, relentless progression of disease, ultimately developing respiratory failure and cor pulmonale and dying at the age of 34 years.

We have reviewed 19 previous reports containing 71 cases of alveolar microlithiasis (36 male, 35 female). Twenty of the 71 patients had died at the time of publication, the mean age at death being 41.7 years (range newborn to 80 years). These fatal cases had been followed up for a mean period of 7.1 (range 0–25) years from the initial diagnosis. There were 51 live patients (mean age 29.1, range 2–59 years), 27 of whom had had no follow up. All 27 were symptomless at the time of publication apart from two patients with mild dyspnoea. The other 24 patients had been followed up for a period of 6.7 (range 1–16) years from diagnosis. Five of these patients complained of increasing dyspnoea and a further two had cor pulmonale. The most common cause of death was respiratory failure and cor pulmonale. Recurrent pneumothoraces in this condition have been previously described.[4] These figures suggest that although this condition is often associated with a long asymptomatic period many patients eventually die from its progression and associated complications.

Bronchoalveolar lavage is of proved benefit in alveolar proteinosis and although the concretions in microlithiasis are mineral (calcium and magnesium phosphates and carbonates) rather than proteinaceous, and may therefore be more difficult to remove, this report shows that it is possible to extract material from the lungs in this condition. Although lavage did not benefit this patient (as the removal of 14.5 g was likely to be insignificant in relation to the total microlith load) further investigation of the use of this technique, before the terminal stage of this condition, is required.

Patients with microlithiasis are often young and, apart from their lung and associated heart disease, well. We suggest that heart and lung transplantation should now also be considered in the management of such patients.

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