Fibreoptic bronchoscopy in diagnosis of bronchopulmonary Kaposi's sarcoma

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ABSTRACT Kaposi's sarcoma of the lung in patients with the acquired immune deficiency syndrome is often indistinguishable by clinical and radiographic criteria from opportunistic pneumonia. Pulmonary Kaposi's sarcoma and pneumonia may frequently be present in the same patient. Previous observers have commented on the repeated failure to establish a diagnosis of Kaposi's sarcoma of the lung by fibreoptic bronchoscopy. Thirteen fibreoptic bronchoscopies were performed in a series of 11 patients with thoracic manifestations of AIDS and Kaposi's sarcoma was identified in transbronchial or bronchial biopsy specimens in four patients. This diagnostic yield is comparable to that obtained only by open lung biopsy procedures in previous reports. Fibreoptic bronchoscopy may contribute to the correct management of the patient and facilitate an accurate prognosis by differentiating between opportunistic pneumonia and pulmonary Kaposi's sarcoma.

Disseminated visceral Kaposi's sarcoma has emerged as a common complication of the acquired immune deficiency syndrome (AIDS). The diagnosis of Kaposi's sarcoma of the lung by fibreoptic bronchoscopy has proved difficult; open lung biopsy has been advocated as the only satisfactory diagnostic procedure. We report four patients in a series of 11 in whom pulmonary Kaposi's sarcoma was diagnosed by fibreoptic bronchoscopy.

Patients and methods

Thirteen fibreoptic bronchoscopies were performed on 11 men presenting consecutively with pulmonary complications of AIDS. All the patients were homosexual. Endobronchial Kaposi's sarcoma was seen in one patient as diffuse, red, friable streaks throughout the mucosa of the tracheobronchial tree. Three bronchial biopsies were performed, histological examination of the specimens showed Kaposi's sarcoma. In a second patient numerous discrete, purple, red raised lesions with a smooth surface were seen in the trachea and main bronchi. This appearance was identical to that of Kaposi's sarcoma in other mucous membranes and no biopsy samples were taken. The tracheobronchial mucosa in the other nine patients appeared normal and biopsy samples were not taken.

Four to six transbronchial biopsy specimens were taken in all patients from the lobe that was most affected on the chest radiograph. Each biopsy specimen was embedded in paraffin wax, sectioned at five levels, and stained with haematoxylin and eosin, Perl's stain for haemosiderin, and Gordon and Sweet's impregnation stain for reticulin. Pulmonary Kaposi's sarcoma was found by transbronchial biopsy in a further two patients (table). No evidence of bronchopulmonary Kaposi's sarcoma has been found in the remaining seven patients of whom three have had further bronchoscopies. Three of the seven have died, the longest surviving for 18 months from initial presentation. In the four patients still alive the longest survival has been for 17 months. Necropsies were not performed on any patients.

Discussion

Before the outbreak of AIDS in 1981, Kaposi's sarcoma was a rare neoplasm seen mainly in Ashkenazi Jews in Poland, Italy, and Russia. In the classical form described by Kaposi in 1872 it was a disease of the elderly affecting mainly the skin of the distal limbs. Although it followed an indolent course, up to 30% of patients developed an associated lymphoreticular neoplasm. In areas of east central Africa, the natural history of Kaposi's sarcoma is different. Here it accounts for up to 9% of all malignant tumours and runs an aggressive course among young adults, with widespread disease of lymph nodes and viscera.

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Table Clinical data from four cases of pulmonary Kaposi's sarcoma (KS) diagnosed by bronchoscopy

	Patient No:			
	1	2	3	4
Chest radiograph	Normal	Soft shadows	Reticulonodular shadows, pleural effusions	Soft shadows, bilateral effusions
Pulmonary infection	—	Toxoplasma, cytomegalovirus	—	Candida albicans
Skin	KS	—	KS	—
KS at bronchoscopy	Bronchial	Pulmonary	Pulmonary	Bronchial
Outcome	Died after 4 months	Alive at 1 year	Died after 9 months	Died after 6 months

Although there is no greater prevalence of antibodies to human immunodeficiency virus in Africans with endemic Kaposi's sarcoma, this is the clinical and pathological pattern that is now encountered frequently in patients with AIDS in the United States and Europe. Kaposi's sarcoma is now the second most common AIDS associated disease, and is the presenting feature in 28% of sufferers from AIDS in the United Kingdom and 14% in the United States.

Although the cell of origin is generally agreed to be the pleuripotential angioblast, it is not clear whether the stimulus to sarcomatous change results directly from disturbed immunological mechanisms or from

the consequent predisposition to oncogenic viruses. Ribonucleic acid from the cytomegalovirus, antigens related to cytomegalovirus, and DNA fragments from cytomegalovirus have been found in up to half of tumour biopsy specimens. That cytomegalovirus may have a causative role is further suggested by the observation that immunosuppressed patients who have Kaposi's sarcoma have significantly higher titres of antibody to cytomegalovirus than those without Kaposi's sarcoma. Similarly, high titres were found in African patients and homosexuals with Kaposi's sarcoma even before the outbreak of AIDS. The greater prevalence of Kaposi's sarcoma in the United King

Kaposi's sarcoma in bronchus wall, with multiple irregularly shaped vascular spaces lined by pleomorphic cells, clumps of spindle cells, and extravasated erythrocytes. (Haematoxylin and eosin.)

dom than in the United States may be explained by the greater proportion of homosexuals in the AIDS population of the United Kingdom.

The incidence of Kaposi's sarcoma affecting the lung in patients with AIDS is uncertain. Cohort studies of patients with pulmonary symptoms of AIDS give figures of between 8–33%.^{1,2} The disease is more frequently found at necropsy than is suspected before death. The clinical and radiological picture of bronchopulmonary Kaposi's sarcoma is indistinguishable from that of many opportunistic pneumonias in patients with AIDS. Moreover, it is unusual to find Kaposi's sarcoma in the lungs in the absence of pulmonary infection.³ The symptoms of bronchopulmonary Kaposi's sarcoma are breathlessness, dry cough, haemoptysis, fever, malaise, and weight loss. The radiographic changes of diffuse or reticulonodular shadowing are similarly non-specific; indeed, the chest radiograph may be normal. Pleural effusions, often blood stained, are said to be characteristic, but cytological examination of pleural fluid is unhelpful.¹ Fibreoptic bronchoscopy plays an important part in the investigation of patients with thoracic manifestations of AIDS, but our finding of evidence of pulmonary Kaposi's sarcoma in a third of this small series of cases contrasts with the experience of many previous authors, who have commented on the failure of this procedure to detect Kaposi's sarcoma in the lungs in patients shown by open lung biopsy or at necropsy to have the neoplasm.^{1–4} Open lung biopsy has repeatedly been recommended as the only procedure with an acceptable diagnostic yield.

The reason why Kaposi's sarcoma has proved so elusive at bronchoscopy is uncertain; the explanation may lie partly in the patchy distribution of microscopic foci of neoplasm throughout the lung parenchyma and in the absence of specific tumour markers. Open lung biopsy specimens have shown a predominantly lymphatic distribution in the pleura, septa, and peribronchial region,⁵ this last area being the most accessible to transbronchial biopsy. The histological features are variable; whereas "classical" Kaposi's sarcoma is readily identifiable, early changes comprising only increased numbers of spindle cells,

mesenchymal cells, erythrocytes and haemosiderin deposits may be overlooked (fig 1). Biopsy specimens should be examined specifically for both opportunistic infections and Kaposi's sarcoma. In the tracheobronchial tree Kaposi's sarcoma has a variable appearance, and apart from those lesions that are clearly Kaposi's sarcoma in a patient already known to have the tumour, biopsy specimens should be taken from all areas of abnormal mucosa.

Figures from the Communicable Disease Surveillance Centre show that patients with AIDS who have Kaposi's sarcoma alone have the most favourable prognosis, with a median survival of 21 months from the time of diagnosis. The median survival of those with *Pneumocystis carinii* pneumonia is 12.5 months and patients with both Kaposi's sarcoma and pneumocystis pneumonia have the worst prognosis—only 6.6 months.⁶ Although survival has not been influenced by treatments currently available for Kaposi's sarcoma, the differential diagnosis from opportunistic pneumonia is important in the management of the patient and in determining prognosis.

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