

Kaposi's sarcoma of the lung: radiography and pathology

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Kaposi's sarcoma is a neoplasm of multifocal origin apparently derived from fibroblastic and vascular elements and predominantly involving the skin. In 10 to 20% of patients with the neoplasm, associated lesions may occur in the lymph nodes, gastrointestinal tract, liver, lungs, heart, bone, and spleen in order of decreasing frequency. Kaposi's sarcoma in a previously healthy person occurring in the internal organs without cutaneous lesions is exceedingly rare. Such a case, with predominantly thoracic involvement is presented here.

Case report

A 42-year-old white male was admitted to hospital complaining of increasing fatigue, dyspnoea, and haemoptysis. Four months ago he had noted easy fatiguability, malaise, and a cough with clear sputum production. He was a 35 pack year smoker previously in excellent health. A chest radiograph at that time was interpreted as normal. Six weeks before admission he had experienced severe substernal tightness with associated sweating, nausea, vomiting, and moderate dyspnoea, but an electrocardiogram did not suggest myocardial damage. The chest film showed left ventricular enlargement and a diffuse coarse reticulonodular infiltrate uniformly distributed through both lungs. Digitalis, diuretics, and a low salt diet were given. Though the radiographic findings did not change, all his symptoms except the dyspnoea abated and he was sent home. Three weeks before admission he became more tired and short of breath, and noticed blood-tinged sputum.

At the time of admission he had a respiratory rate of 24 per minute and a normal pulse and blood pressure. He was afebrile. There were mid-inspiratory crackles in the lower and mid-zones. No other clinical abnormality was found and there were no skin lesions.

Haemoglobin was 13.3 gm%, haematocrit 39.7%, total leucocyte count 16,100/mm³ with 80% segmented neutrophils and 4% band forms. The sputum was grossly bloody. Gram stain showed mixed flora and the smear for tuberculosis bacilli was negative. The erythrocyte sedimentation rate was 15 mm per hour. The arterial blood gases on room air showed a pH of 7.50, Pao₂

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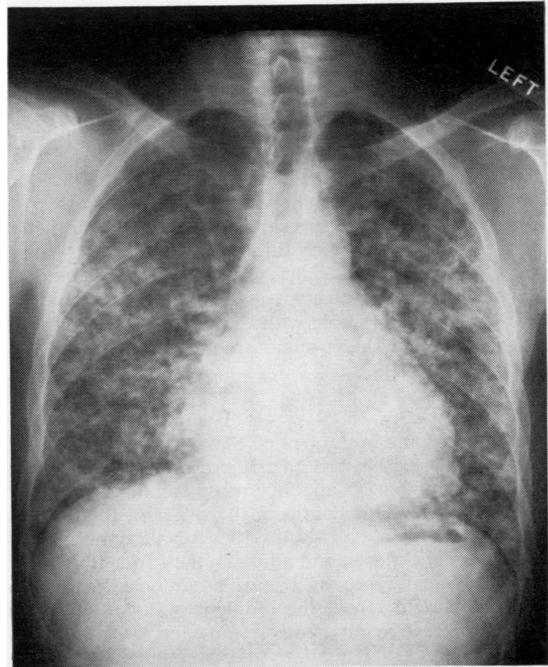


Fig 1 PA chest film on admission shows marked progression of the pulmonary infiltrate.

of 50 mmHg, and a Pao₂ of 37 mmHg. The chest film (fig 1) showed further increased cardiomegaly with an increased patchy, predominantly nodular infiltrate. The nodules were poorly margined and showed a tendency to coalesce.

Fibreoptic bronchoscopy was normal except that blood was coming from all the subsegmental bronchi. The bronchoscopic aspirates proved negative for acid-fast bacilli, fungi, and cytology. The transbronchial lung biopsy disclosed moderately thickened, congested alveolar septa containing occasional haemosiderin granules with infiltration by fibroblasts, histiocytes, and macrophages. No tumour cells were seen in the specimen. Skin tests showed him to be anergic.

The patient's dyspnoea worsened and haemoptysis continued. Arterial blood gases on 50% FiO_2 showed a pH of 7.46, Paco_2 of 43 mmHg, and Pao_2 of 57 mmHg reflecting a shunt of approximately 12%. He was intubated and placed on a mechanical ventilator, but deteriorated and died on the ninth hospital day.

At necropsy the left lung weighed 1200 gm and the right 1400 gm. There were 300 ml of serosanguineous fluid in the right pleural cavity and 400 ml in the left. The major pulmonary arteries were normal. Discrete dark red haemorrhagic nodules measuring 0.4-0.6 cm were found throughout the lung and pleura but the bronchial tree was uninvolved. These nodules were also found on the parietal pericardium, subepicardial fat and right atrium, and were seen in some areas to compress the coronary arteries, but the heart valves and ventricular myocardium were normal. A single nodule each was found in a mediastinal lymph node and the thyroid. The skin was not involved.

Microscopically, the haemorrhagic nodules in the lung varied from a lake of blood traversed by thin fibrous septa covered by neoplastic epithelium to solid nests of spindle-shaped tumour cells containing indefinite blood channels, scattered erythrocytes, and haemosiderin. Occasionally a larger arteriole showed its endothelial lining to be interrupted by tumour cells. The intervening lung showed varying stages of interstitial pneumonia and extensive intra-alveolar haemorrhage. The above findings were compatible with the diagnosis of Kaposi's idiopathic haemorrhagic angiosarcoma (fig 2).

Discussion

Kaposi's sarcoma in the United States accounts for 0.6% of all cancers, and is particularly prevalent in individuals of Italian and Jewish ancestry most commonly in their seventh decade. This contrasts sharply with the African incidence where it constitutes 10% of all tumours and affects mostly children and adults in their fourth decade.¹

Four clinical types of Kaposi's sarcoma have been described ranging from the indolent nodular type with slowly progressive cutaneous lesions and late manifestations in the viscera, if at all, to the disseminated aggressive type, which involves many systems and uncommonly involves the skin.² The pathological manifestations in the thoracic cases have been well documented.^{2,3} Our case is an example of the disseminated aggressive form involving the lungs with a few lesions in other viscera and with no cutaneous lesions. He had the very short course characteristic of this form of the disease, with progressive hypoxaemia probably due to shunting from extensive infiltration of lung.

The radiographic pattern in the chest seems to be similar in our case to other reports in the literature.^{2,4}

An increasing association of Kaposi's sarcoma has been noted with Hodgkin's disease, lymphosarcoma, lymphocytic leukaemia, myeloid leukaemia, multiple myeloma, and a variety of epithelial carcinomas, as well as with immunosuppressive therapy.⁵ In organ transplant recipients, Kaposi's sarcoma accounts for 4.9% of all de

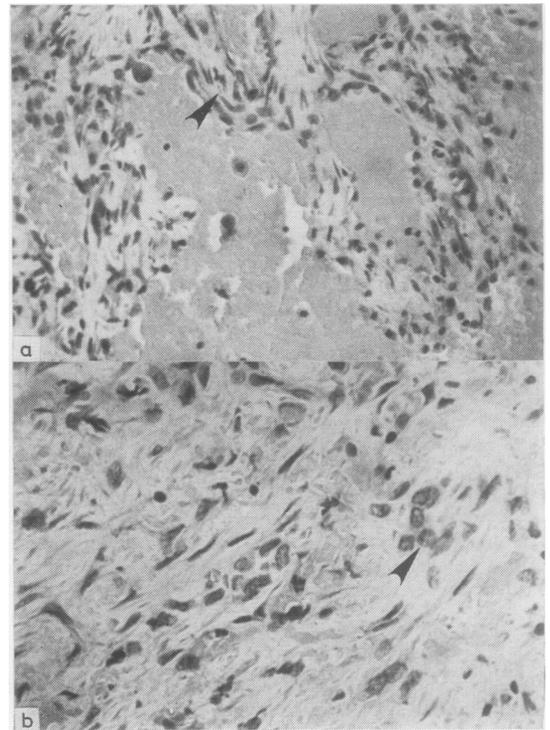


Fig 2 (a) Microscopic appearance of the tumour in the lung demonstrating whorled masses of spindle-shaped malignant cells (arrow) surrounding lakes of erythrocytes (magnification $\times 200$). (b) Higher power view showing malignant cells coursing through lung parenchyma with destruction of pulmonary capillary (arrow) (magnification $\times 600$).

novo neoplasms. In 45% of the cases the internal organs were involved and 30% had a preceding *Herpes simplex* infection. Interestingly, after cessation or partial reduction of the immunosuppressive agents, the tumour in some cases has regressed.

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