PRIMARY LYMPHOCYTIC LYMPHOMA (LYMPHOSARCOMA) OF THE LUNG

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SYNOPSIS

A case of primary lymphosarcoma of the lung in a woman aged 57 years is presented. This is the first documented case in which bilateral resection of the lung was performed.

INTRODUCTION

Primary lymphosarcoma of the lung is a rare disease. In 1947, Churchill, and Spatt and Grayzel reported the first two cases that were successfully resected. A total of seven cases seen at the Brompton Hospital, London, over a period of 20 years were analysed recently by Rees (1973) and so far about 120 cases have been reported in the literature.

In view of the limited clinical experience and uncertainty regarding its course and management, a case of primary lymphosarcoma of the lung is reported. This is the first documented case in which bilateral resection was done.

CASE REPORT

The patient, a 57 year old Chinese woman, was first seen in January 1973 with a history of cough productive of a little whitish sputum of two years' duration. There was no associated fever, staining of her sputum nor loss of weight. She did not smoke and there was no occupational or industrial history of note.

Clinically her general condition was good. There were no abnormal physical findings. A chest radiograph (Fig. 1) showed consolidation of the middle lobe and lingula. Routine blood and urinalysis were normal; her erythrocyte sedimentation rate was 50 mm per hour. The tuberculin test to 1 TU PPD was 5 mm. Bronchoscopy revealed normal findings and the brochial aspirate was negative for malignant cells. A bilateral bronchogram showed patent bronchi to the middle lobe and lingula. Serum protein electrophoresis and immunoglobulin estimation were normal.

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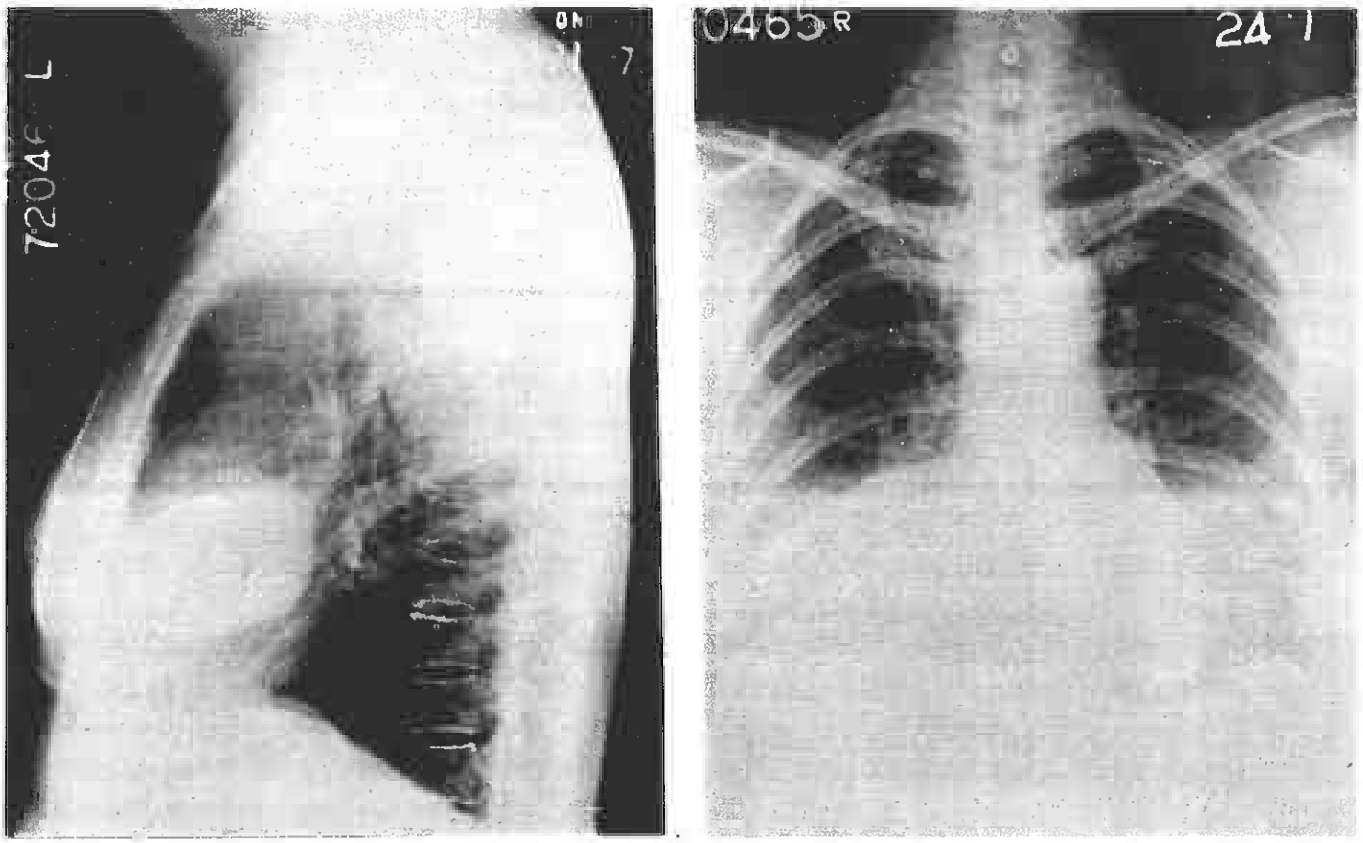

Fig. 1. Chest radiograph showing consolidation of middle lobe and lingula.

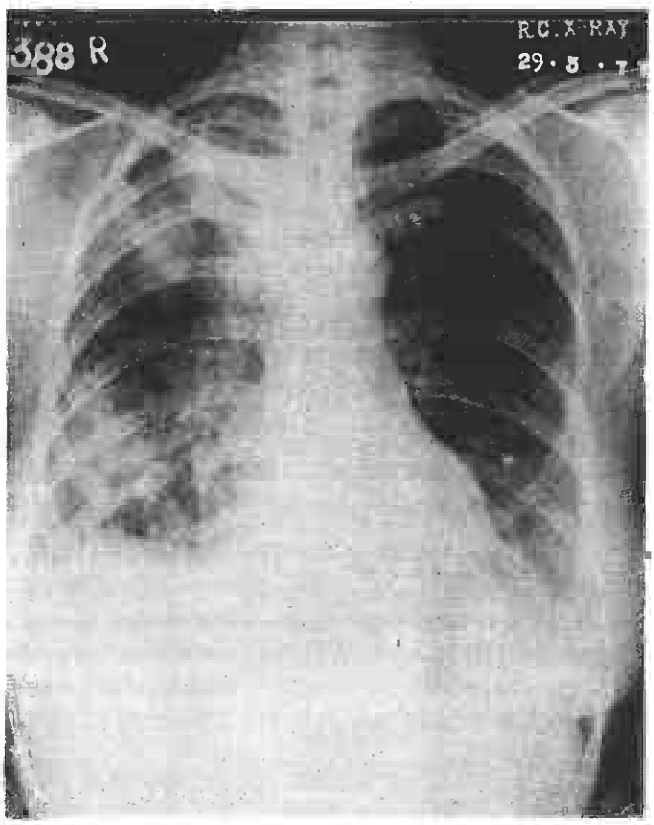

Fig. 2. Chest radiograph showing opacities in the right lung and left lower lobe.

A tentative diagnosis of primary lymphosarcoma of the lung was made but this could not be established as she refused an exploratory thoracotomy.

She was followed up until May 1973 when she presented with fever and right sided chest pain for two days, and signs of a right plueral effusion. The chest radiograph showed a loculated effusion but repeated aspirations yielded no fluid. Her fever settled with a course of antibiotics. She was also found to be a mild diabetic.

She finally consented to a right thoracotomy in June 1973. The middle lobe was found to be uniformly firm and of rubbery consistency. Some loose adhesions were present between the middle and lower lobes. A small pocket of sterile fibrinous exudate, possibly old loculated empyema, was present in the mid zone posterolaterally. Some soft hilar nodes were felt. A middle lobectomy was performed and the loculated exudate removed.

Section of the resected lobe showed a homogenous whitish surface with a rim of lung tissue at the periphery. Histologically the appearances were those of a lymphocytic lymphoma. The tumour was composed of a uniform population of

darkly staining cells with round or oval nuclei and a central nucleolus with very little cytoplasm.

Three weeks later a left thoracotomy was performed. The lingula segment of the upper lobe was found to be infiltrated with a solid tumour. No adhesions were present and the hilar lymph nodes were not enlarged. A lingulectomy was done. Examination of the resected lobe showed a similar histological picture to the middle lobe. There was no invasion of the major vessels.

Her post operative course was uneventful. She remained well over the next three years. In September 1976, she complained of cough again. Her chest radiograph revealed a pneumonic shadow in the right upper lobe. This failed to respond to a course of antibiotics. Her condition gradually deteriorated with increased cough and progressive dyspnoea. There was extension of the opacities to the right and left lower lobes (Fig. 2). Investigations revealed a hemoglobin of 9.4 g%, normal white cell and platelet counts and an ESR of 137 mm per hour. Sputum cultures for pyogenic, fungus and acid fast bacilli were negative. Cryptococcal antigen was not detected in the blood and the complement fixation titre for cytomegalovirus was not significantly raised. The liver function tests were normal except for a raised alkaline phosphatase of 189 units. Serum protein electrophoresis showed albumin 2.9 g%, α_1 globulin 0.4 g%, α_2 globulin 0.9 g%, β globulin 0.7 g% and α globulin 0.9 g%. Immunoglobulin estimation revealed IgG 750 mg%, IgA 144 mg% and IgM 251 mg%. Bone marrow examination was reported as showing a non-specific reactive hyperplasia; the direct Coomb's test was negative.

She developed hepatomegaly and bilateral enlargement of supraclavicular lymph nodes in April 1977. Biopsy of the right supraclavicular lymph node revealed a poorly differentiated lymphocytic lymphoma. She was started on Cyclophosphamide, Vincristine and Prednisolone, but failed to maintain her initial response to chemotherapy and died in June 1977.

DISCUSSION

Controversy exists regarding the diagnostic criteria for primary lymphosarcoma of the lung. Papaioannou and Watson (1965) state that the disease "should be confined to one lung with or without hilar involvement but without mediastinal spread". This view has been regarded an "an arbitrarily harsh distinction" by Rees (1973) who also considers as "unrealistic" the exclusion of those in whom the contralateral lung is involved "when the tumour's properties of transgressing interlobar tissues and overlying pleura are well known". Pulmonary involvement secondary to generalised disease occurs in about 7% of patients with malignant lymphoma (Robbins, 1953). To exclude cases of malignant lymphoma of the usual type in which dissemination already has occurred but is not apparent at the time of diagnosis, Saltzstein (1969) considers that there must be no evidence of dissemination for at least three months after diagnosis. He also excludes cases in which a diagnosis was made at autopsy only.

The tumour is believed to originate from the lymphoid tissue present under the pleura, around the bronchi and pulmonary vessels or within the parenchyma itself. As it usually does not infiltrate or obstruct the lumen of vessels or bronchi, diagnostic procedures such as bronchoscopy and exfoliative cytology are rarely rewarding.

About 50% of cases are asymptomatic being detected on routine radiographs of the chest. Patients who do have symptoms present with cough, chest pain, weakness, fatigue and weight loss. Rabbiah (1968) in an analysis of 94 reported cases, found that the most common radiological presentation was an infiltrative homogeneous pneumonic area of consolidation (45.4%), with 20.4% showing a fairly localised area of density and 17% presenting as a 'coin lesion'. The most common site of the tumour was in the right upper lobe.

Adequate surgical resection is considered the treatment of choice and "prophylactic" radiation therapy does not seem to be indicated (Papaioannou and Watson, 1965). Although some routinely add postoperative radiotherapy, (Havard et al, 1962), the addition of postoperative radiotherapy does not seem to add appreciably to long-term survival (Rees, 1973). Papaioannou and Watson (1965) report a five year survival rate of 44% following surgery alone, 42% after radiotherapy alone and 40% after treatment with both. Chemotherapy is usually reserved for disseminated disease, but poor responses are not uncommon as noted by Rees (1973) and in this patient.

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