Virchows Arch. A Path. Anat. and Histol. 372, 161 – 165 (1976)

Virchows Archiv A

Case Reports

Pulmonary Blastoma

H.B. Marsden and C.L. Scholtz

Department of Pathology, Royal Manchester Childrens' Hospital,
and Department of Pathology, University of Manchester,
Manchester, M13 9PT, England

Summary. A primary pulmonary tumour in a four year old boy arising from the subpleural zone of the lung is described. It contains both stromal and epithelial elements at the primary site and in the bony metastases. The combination of the age of the patient, the site of the tumour, the stromal and epithelial elements present in the tumour suggest that this is a true pulmonary blastoma. Electronmicroscopy showed the presence of intranuclear viral particles.

Key words: Lung – Blastoma.

Introduction

Primary malignant pulmonary tumours are amongst the rarest tumours in the paediatric age group. In a twenty year period, 1. 1. 1954 to 31. 12. 1973, only four cases were reported to the Manchester Tumour Registry, which is based on approximately one million children under fifteen years, when 2207 malignant tumours were recorded. One was a well differentiated squamous cell carcinoma, one a poorly differentiated carcinoma, the third a primary malignant teratoma and the fourth a pulmonary blastoma. It is the purpose of this paper to describe the latter tumour.

Case Report

A four year old boy presented with general malaise and dyspnoea at rest for two weeks. On examination he had a right pleural effusion and a chest X-ray demonstrated a large solid mass in the right side of the chest displacing the mediastinum to the left. Aspiration of the pleural effusion and lung biopsy demonstrated a predominantly undifferentiated tumour with polyginal and spindle cells and a moderate number of mitoses. There were some rosette-like structures and possibly early tubules but no fully formed ones; however, epithelium was seen.

He was treated with radiotherapy to the tumour and cytotoxic therapy. Initially he improved clinically; however, he ran a course of intermittent respiratory infection and died in respiratory failure nine months after diagnosis.

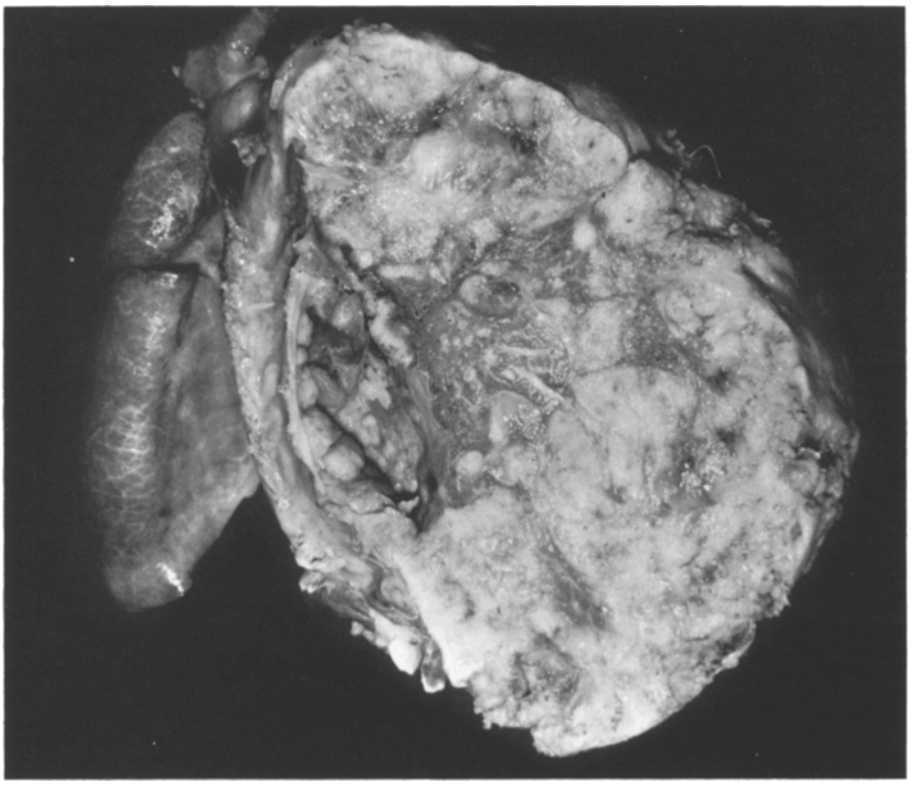

Fig. 1. Pulmonary blastoma: Tumour encasing the right lung with nodules in the residual tissue

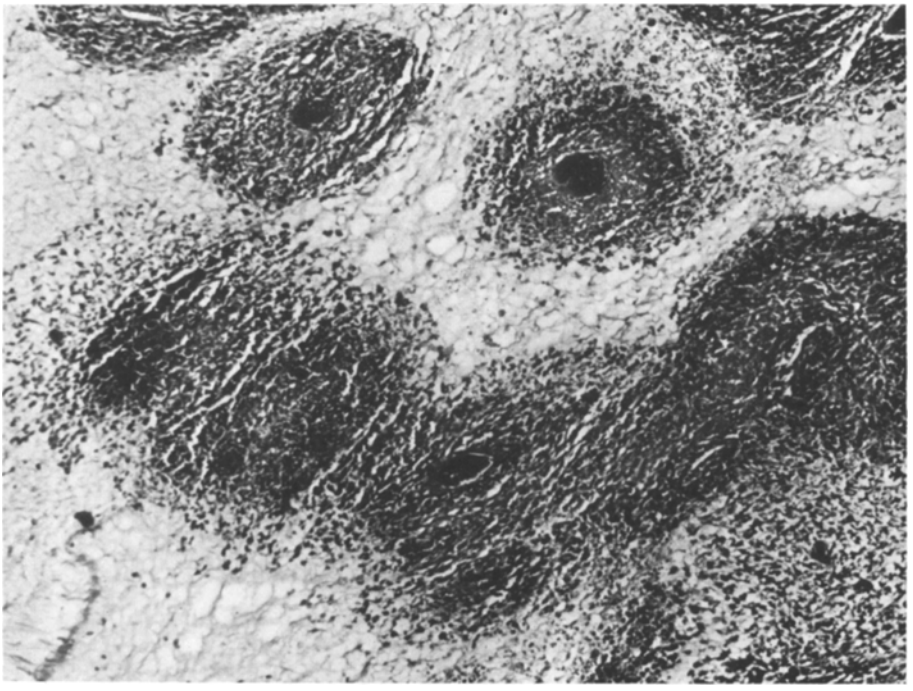

Fig. 2. Pulmonary blastoma: Nodules of tumour around small blood vessels separated by loose mesenchyme. H&E ×75

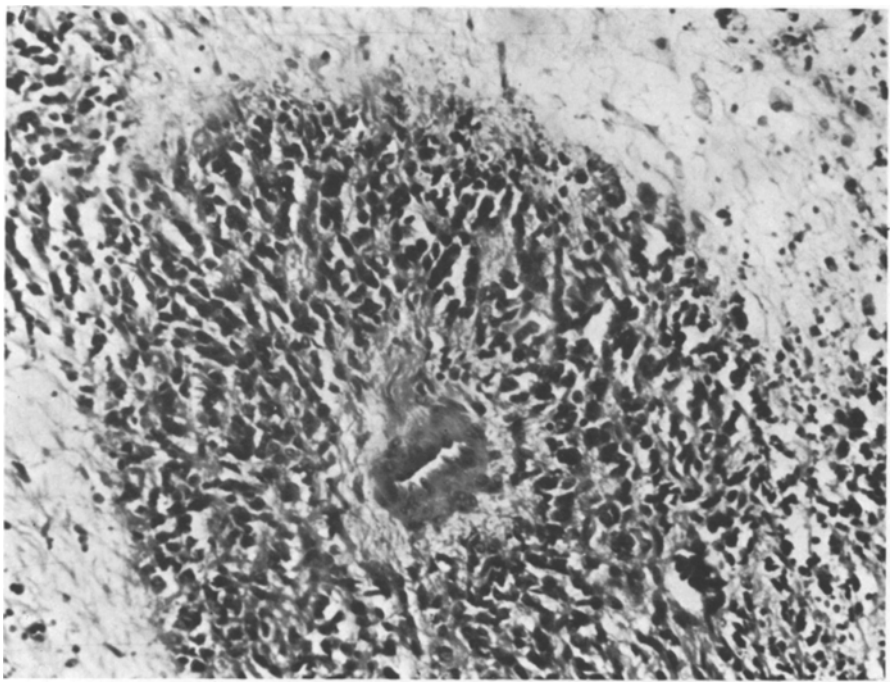

Fig. 3. Pulmonary blastoma: Epithelial clefts are evident in the tumour aggregates. H&E ×250

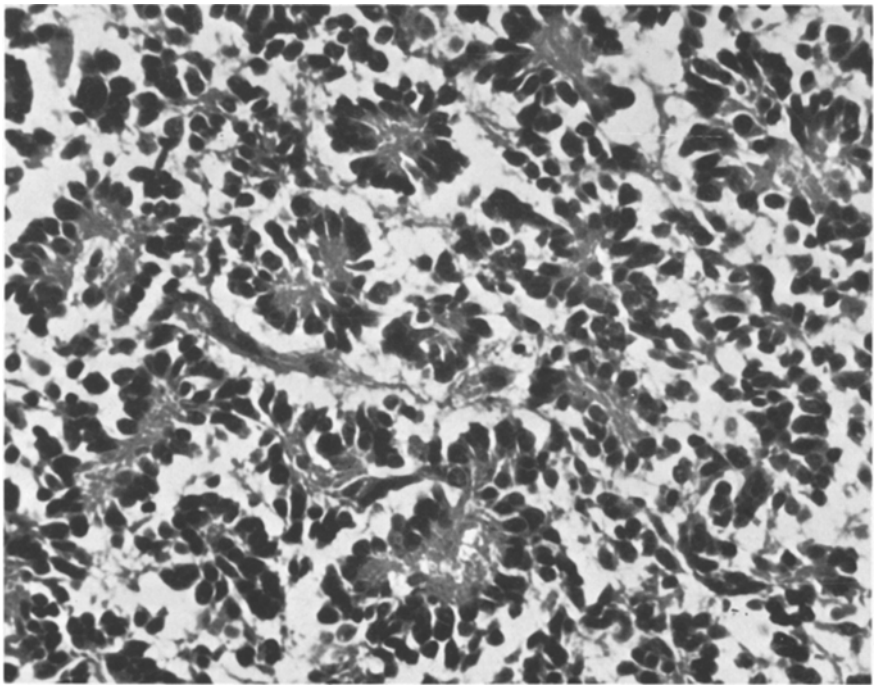

Fig. 4. Pulmonary blastoma: Rosette-like appearance with eosinophilic central zones in a more differentiated area. H&E ×400

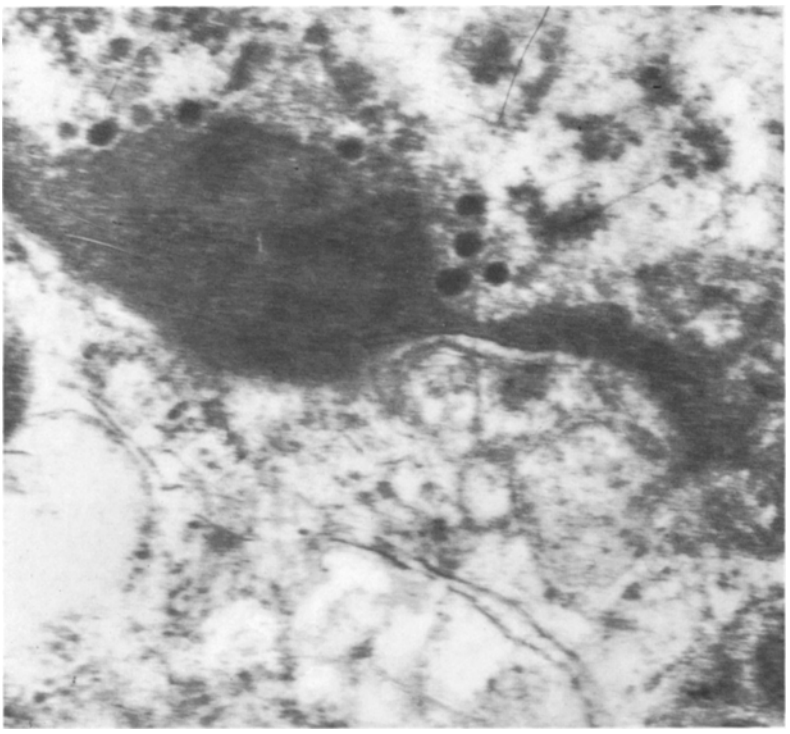

Fig. 5. Pulmonary blastoma: Intranuclear virus particles surrounded by a halo. EM ×40,000

Autopsy Findings. The right lung was almost completely replaced by a yellow grey tumour which encircled the lung, (Fig. 1) and extending through the diaphragm into the intercostal muscles and pericardium. Small areas of lung were present at the hilum which contained nodules of tumour. Secondary deposits of tumour were present in the right iliac bone and vertebral bodies.

Histology. The *tumour* was composed of loose, poorly differentiated mesenchyme arranged around blood vessels separated by more loosely textured tissue (Fig. 2). Epithelial clefts were evident (Fig. 3) which were papillary in some differentiated areas, giving an appearance of rosettes with eosinophilic centres (Fig. 4). Bronchial channels were seen at the periphery of the tumour lined by irregular flattened and cuboidal epithelium. Few mitoses were seen.

The tumour present in the *secondary deposits* in bone was predominantly of the undifferentiated type; however, small papillary areas were seen.

Electronmicroscopy was diagnostically unrewarding; however, intranuclear bodies were identified suggestive of viral particles (Fig. 5).

Discussion

About thirty cases of pulmonary blastoma have been described (Peacock and Whitweel, 1976) but few young patients are included in case reviews of this tumour. This patient is one of the youngest reported under this heading. Only about one quarter have been described in juvenile patients.

Lewis (1973) described a patient aged 115 months and the ages of others reported are 34 months and 5 years 9 months (Willnow and Hofmann, 1974), 11 years (Iverson and Straehley, 1973), 15 years (Nazari et al., 1971), 19 years (Spencer, 1961) and 20 years (Chitambar et al., 1969). If the tumour were embryonic in origin the peak incidence should be similar to other embryonic tumours. The spread of the age incidence could reflect the rarity of the tumour or that several tumours are included in this entity and several authors have suggested that the tumour is a carcinosarcoma (e.g. Barson et al., 1968).

Spencer (1961) suggests that the tumour may originate from undifferentiated mesenchyme at the periphery of the lung which is capable of differentiating into stromal and epithelial constituents. Such a concept is in keeping with the origin of the tumour in the present report. This tumour clearly contains stromal elements. The epithelial elements resemble bronchial channels and their neoplastic features suggest that they are part of the tumour. The absence of smooth muscle around the channels is in favour of a neoplastic origin of the epithelium; however, the fact that the channels are seen only at the peripheral or growing portion of the tumour suggests that they are derived from pre-existing lung tissue.

It is difficult to assess the mode of origin of the true epithelial channels and an origin from existing ones cannot be excluded in spite of atypical epithelial appearances.

Few reports show the tumour arising from the periphery of the lung, most are plaques or intrapulmonary masses (Barson et al., 1968). In this case the tumour is encasing the lung suggesting the whole of the subpleural mesenchyme is involved in the neoplastic process. The metastaic tumour in the vertebrae contains both stromal and epithelial elements showing that tumour emboli are capable of differentiating into both stromal and epithelial elements.

A diagnosis of haemangiopericytoma was considered because of the presence of perivascular tumour collections. This was excluded on reticulin staining and it seems more probable that this orientation is related to nutrition of the tumour cells.

References

Barson, A.J., Jones, A.W., Lodge, K.V.: Pulmonary blastoma. J. clin. Path. **21**, 480–485 (1968)

Chitambar, I.A., Gujral, J.S., Aikat, B.K.: Embryonal sarcoma of the lung. J. thorac. cardiovasc. Surg. **57**, 657–662 (1969)

Iverson, R.E., Straehley, C.J.: Pulmonary blastoma: Longterm survival of a juvenile patient. Chest **63**, 436–440 (1973)

Lewis, J.E. Jr.: Noted as a personal communication by Iverson, R.E. and Straehley, C.J. in 'Pulmonary blastoma: Longterm survival of a juvenile patient'. Chest **63**, 436–440 (1973)

Nazari, J.C., Amir-Mokri, E., Sarram, A., Yaghmai, I.: Pulmonary blastoma. Chest **60**, 187–189 (1971)

Peacock, M.J., Whitwell, F.: Pulmonary blastoma. Thorax **31**, 197–204 (1976)

Spencer, H.: Pulmonary blastomas. J. Path. Bact. **82**, 161–165 (1961)

Willnow, U., Hofmann, V.: Zur Morphologie und Histogenese der primären Lungentumoren des Kindes. Helv. paediat. Acta 425–438 (1974)

Received August 19, 1976