Pulmonary blastoma is a rare tumour of the lung composed of immature mesenchyma and epithelium that morphologically mimics the embryonal lung structure. The first report of pulmonary blastoma was made in 1945 [1], and only 50 cases of pulmonary blastomas have been reported to date. The histogenesis of pulmonary blastoma is still unknown because of its rarity. Recently, several pathogenetic possibilities have been proposed, but no definite conclusion has been reached.

Case report

A 61 yr old female smoker (7–8 cigarettes-day⁻¹ for 15 yrs) had suffered from systemic sclerosis for 10 yrs. The onset signs were Raynaud’s phenomenon and puffy fingers, and systemic sclerosis was confirmed with cutaneous biopsy which showed fibrotic changes in the skin and subcutaneous tissue. No features of the calcinosis, Raynaud’s phenomenon, oesophageal dysmotility, sclerodactyly, telangiectasia (CREST) syndrome were seen during the clinical course. She had received D-penicillamine and corticosteroid treatment in the dermatology department for 3 yrs. She was referred to our department on May 22, 1995, with a one-month history of cough producing bloody sputum.

She was afebrile and had no dyspnoea or cyanosis. Her blood pressure was 150/72 mmHg and cardiac frequency 90 beats·min⁻¹ with a regular rhythm. The heart and breath sounds were normal. There were no abnormal neurological findings. The neck and axillary lymph nodes on the left side were firm and swollen. The abdomen was soft with no organomegaly. Cutaneous scleroderma was present on the extremities, face and trunk. Chest radiography on ad-mission revealed multiple abnormal opacities in both lung fields (fig. 1a). A computed tomography (CT) scan of the chest showed a 3.4×6.6 cm tumour mass in the left S9 area, which was designated as primary lesion, with multiple metastatic lesions (fig. 1b). Several peripherally sited metastatic lesions have pleural involvement. Fibrotic changes were also recognized in both lower lung fields. The haemoglobin level was 9.2 g·dL⁻¹ and lactate dehydrogenase was 467 U·L⁻¹. Antinuclear antibodies were strongly positive (>1280, nucleolar type), but anti-deoxyribonucleic-acid, anti-ribonucleoprotein (RNP), and anti-scleroderma (Scl)-70 antibodies were negative. Hepatitis B virus (HBs) antigen and hepatitis C virus (HCV) antibody were negative. Liver function parameters were within normal limits. Levels of tumour markers associated with lung cancer were high: carcinoembryonic antigen (CEA) was 551.7 ng·mL⁻¹ (normal limit <3.0 ng·mL⁻¹), sialyl lewis-i antigen (SLX) was 120 U·mL⁻¹ (normal limit < 37 U·mL⁻¹), cytokeratin 19 fragment (CYFRA) was 29.6 ng·mL⁻¹ (normal limit <1.0 ng·mL⁻¹). In addition, the elevation of serum α-fetoprotein (AFP) was remarkable, >80,000 ng·mL⁻¹ (normal limit <10 ng·mL⁻¹).

Fibreoptic bronchoscopy revealed a polypoid lesion in the left main bronchus and stenosis distal to that site. The biopsied specimen of the polypoid lesion revealed the well-differentiated foetal adenocarcinoma variant of pulmonary blastoma (fig. 2). Transbronchial lung biopsy, performed through the left B2 area, showed the same histological findings as the polypoid tissue. There were no abnormalities in any other organs including the liver, ovaries or mediastinum. Immunohistochemical staining of the biopsied specimen by an anti-AFP monoclonal antibody demonstrated many positively stained tumour cells (fig. 3).

We diagnosed this case as AFP-producing pulmonary blastoma complicated with systemic sclerosis. The serum α-fetoprotein level was elevated at presentation and the biopsied tumour stained positively against anti-α-fetoprotein antibody. The α-fetoprotein produced autonomously by tumour cells was of yolk-sac origin. Although the pathogenesis of pulmonary blastoma has not been clarified, we suggest that this pulmonary blastoma is a type of yolk-sac tumour. Eur Respir J 1998; 11: 1185–1187.

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Pulmonary blastoma is a rare lung neoplasm, characterized by immature mesenchyma and epithelium that resembles the embryonal pulmonary structure of between 10–16 weeks’ gestation [2, 3]. Only approximately 50 cases of pulmonary blastomas have been reported to date [4]. Recently, pulmonary blastoma was classified into three subtypes: 1) those with a mesenchymal and epithelial appearance (biphasic blastoma); 2) those with only a sarcomatous component, occurring in children <15 yrs of age with an extremely poor prognosis (pleuropulmonary blastomas); and 3) those with a mainly malignant glandular structure of embryonal cells (well-differentiated foetal adenocarcinomas) [4, 5]. Histologically, the present case belongs to the third type of pulmonary blastoma.

The present case had an atypical manifestation of pulmonary blastoma coexistent with systemic sclerosis. However, the relationship between pulmonary blastoma and systemic sclerosis is obscure [6]. We consider this case to have the features of two different conditions.

The present case also had a remarkably high serum AFP level and the glandular cells of blastomas stained positively against anti-AFP monoclonal antibody. This case was diagnosed as an AFP-producing pulmonary blastoma. AFP is an oncofoetal antigen that is produced from the foetal liver and yolk sac [7] and can be quantitated for confirmation of the diagnosis of hepatocellular carcinoma or germ-cell tumours [8]. Recently, Tsuchida et al. [9] detected differences in lectin affinity between AFP produced by the foetal liver and AFP produced by the yolk-sac. In the present case, the concanavalin A (ConA) nonreactive fraction rate for serum AFP was 60% and the reactive fraction was 40%. The lentil agglutinin (LCA) strongly
reactive and weakly reactive subfractions were 31% and 55% respectively (table 1). These results indicated that the AFP of the present case was of yolk-sac origin.

The aetiology of pulmonary blastoma remains obscure because of its rarity. Several hypotheses have been proposed concerning its histogenesis. According to the first theory, supported by Bensch et al. [10], pulmonary blastoma is a true carcinosarcoma. A second theory is that pulmonary blastoma originates in the bronchial epithelium [11]. The resemblance of epithelial components of the pulmonary blastoma to the foetal lung granular structure encourages this hypothesis of the endodermal origin of epithelial elements. The third theory proposed by Davis et al. [12] suggests that pulmonary blastoma is the malignant change of hamartoma. However, pulmonary blastomas with germ-cell differentiation [13, 14] have recently been reported. In these reports, they found the blastomatous areas in the germ-cell component and emphasized the relationship between pulmonary blastoma and yolk-sac tumour histologically. This fourth hypothesis suggests that pulmonary blastoma is a variant of germ-cell malignancy. A high serum concentration of AFP was also recognised in these cases, as in our case. However, immunostaining and the lectin affinity for AFP were not analysed in these reports. In this report, we immunohistochemically detected autonomous AFP production by tumour cells. In addition, this is the first reported case of pulmonary blastoma in which the lectin affinity of AFP was analysed and which indicated that the AFP was of yolk-sac origin.

While it may be argued that the histochemical findings of yolk-sac tumour are not sufficiently established to allow a definitive demonstration of the similarities of immunohistochemistry and lectin affinity with pulmonary blastoma, we submit that our hypothesis is sufficiently compelling to elicit further investigation by investigators with access to more tumours. As such, the present case may shed some light on the path by which further insight into these intriguing tumours might be gained.

### Table 1. – Lectin affinity of serum α-fetoprotein

<table>
<thead>
<tr>
<th></th>
<th>Foetal liver AFP %</th>
<th>Yolk-sac AFP %</th>
<th>Present case</th>
</tr>
</thead>
<tbody>
<tr>
<td>ConA reactive</td>
<td>90–95</td>
<td>45–55</td>
<td>40</td>
</tr>
<tr>
<td>ConA nonreactive</td>
<td>5–10</td>
<td>45–55</td>
<td>60</td>
</tr>
<tr>
<td>LCA</td>
<td>Strongly reactive and nonreactive dominant</td>
<td>Strongly reactive and weakly reactive dominant</td>
<td>Nonreactive 14</td>
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<tr>
<td></td>
<td>Strongly reactive</td>
<td>Nonreactive 14</td>
<td>Weakly reactive 55↑</td>
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<td></td>
<td>Nonreactive 14</td>
<td>Nonreactive 14</td>
<td>Weakly reactive 55↑</td>
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<tr>
<td></td>
<td>Strongly reactive and nonreactive dominant</td>
<td>Strongly reactive and weakly reactive dominant</td>
<td>Nonreactive 14</td>
</tr>
</tbody>
</table>

Shown are the reactive/nonreactive fractions as % values of total serum α-fetoprotein (AFP). ConA: concanavalin A; LCA: lentil agglutinin; ↑: increase.

### References