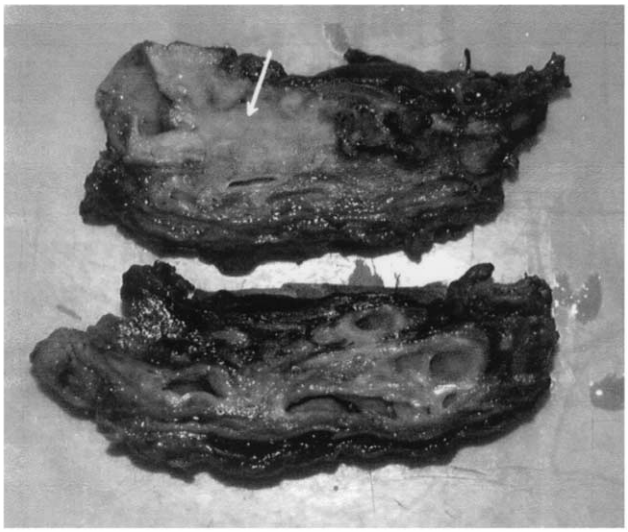

Fig 3. Pathologic specimen of the resected hypoplastic lung (11.6 × 7.5 × 3.4 cm) with the mucoepidermoid carcinoma (2.7 × 2.5 × 1.6 cm) indicated by the arrow.

Comment

Morgagni first described congenital anomalies of the lung in 1762. In 1900, Schneider classified them according to the extent of the deficiency into agenesis, aplasia, and hypoplasia. The latter presents as an ill-formed bronchial tree with a poorly developed alveolar tissue, normally with no defined fissures. Congenital pulmonary hypoplasia (CPH) is not uncommon with an estimated incidence of 1.4 per 1,000 births [2]. When bilateral, CPH is normally associated with neonatal death due to respiratory distress, with a high frequency (more than 10%) of neonatal postmortem examinations revealing the condition [3]. It tends to be associated with multiple congenital malformations. When unilateral, CPH tends to be associated with either maternal oligohydramnios or other anomalies that reduce the available size of the thoracic cavity in utero such as congenital diaphragmatic hernia (more than 40%), kidney abnormalities (25%), and scoliosis [2].

The presence of unilateral isolated lung hypoplasia is a very rare occurrence, presenting in fewer than 10% of the cases of unilateral CPH [2]. It has not been shown to interfere with normal growth or development. In these patients it is usually diagnosed by radiographic evaluation during the management of respiratory tract infections that typically occur from age 3 to 4 years and onward. Although there is an understandable lack of long-term follow-up due to CPH rarity, chronic pulmonary problems have not been reported [4].

Mucoepidermoid carcinoma is a rare malignant tumor of the bronchial tree accounting for 0.2% of the total incidence of lung cancer. The clinical presentation of the case we are reporting is typical of this rare tumor with recurrent episodes of cough, hemoptysis, and features of obstructive pneumonitis. Histopathologic examination shows macroscopically a tumor of bronchial origin and microscopically a combination of squamous cells and mucous glands. Two distinct types can be differentiated according to clinical prognosis and pathologic findings: a low-grade tumor associated with an excellent prognosis in which the bronchial tumor has not invaded lung parenchyma and has little or no mitotic activity in its squamous cells; and a high-grade variety with parenchymal invasion and increased cell mitoses and pleomorphism [5]. The case we present is an example of the high-grade type with pathologic features consistent with the above mentioned.

Although malignant tumors have been described in the context of other lung anomalies, we have failed to find in the English literature a case of lung cancer located in a hypoplastic lung. We recommend a complete radiologic assessment in cases concurrent with congenital abnormalities, as frequently these are multiple. Once they have been excluded, we propose following lung cancer management principles of operability and resectability. If these criteria are satisfied, surgical resection should be the treatment of choice.

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Sclerosing Hemangioma Isolated to the Mediastinum

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Sclerosing hemangioma is an uncommon tumor of unknown histogenesis that generally develops in the lung. We report on a 48-year-old woman with a sclerosing hemangioma that was apparently isolated to the mediastinum. To our knowledge, sclerosing hemangioma arising in the mediastinum has not been previously reported. Potential mechanisms explaining the isolation of sclerosing hemangioma in the mediastinum are discussed.

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Sclerosing hemangioma (SH) of the lung is a rare tumor whose histogenesis is poorly understood. It usually arises in the periphery of the lung as a solitary tumor measuring less than 5-cm diameter [1]. SH of the mediastinum is extremely rare. One case of mediastinal lymph node metastasis from SH of the lung has been reported [2]. We describe a case of SH that apparently isolated to the mediastinum, with no evidence of SH in the lung.

A healthy 48-year-old Japanese woman was admitted for an abnormal shadow on a chest roentgenogram film. Laboratory data were normal, including normal carcinoembryonic antigen, squamous cell carcinoma antigen, and neuron specific enolase. A computed tomographic scan reveald a 7 × 5 cm mass in the posterior mediastinum (Fig 1). Transesophageal ultrasonography illustrated that the mass contained multiple cystic cavities. Magnetic resonance imaging of the chest revealed that the mass in the mediastinum compressed the adjacent left atrium and the right bronchus (Fig 2). Fiberoptic bronchoscopy demonstrated no abnormality, except for external compression of the right bronchus. A provisional diagnosis of bronchogenic cyst was made, and the patient underwent an operation through a right posterolateral thoracotomy. There was no palpable lung mass. The tumor was located in the posterior mediastinum and was in close contact with the pericardium. It was covered by the mediastinal pleura and demonstrated no apparent connection with the lung. Therefore, we could bluntly dissect and isolate the tumor. There was no evidence that the tumor had originated in the lung and invaded the mediastinum (i.e., the tumor appeared to have isolated to the mediastinum).

On gross examination, the tumor measured 7 × 5 × 4.5 cm and was dark red and well circumscribed, but not encapsulated. The cut surface of the tumor consisted of multiple blood-filled cysts with solid components. On microscopic examination, the tumor exhibited four histologic patterns: solid, papillary, sclerotic, and hemorrhagic. The sclerotic area contained calcifications. The tumor cells were interstitial epithelioid cells. Hyperplastic type II pneumocytes lined the surface of the papillary and cystic structures (Fig 3). Mitotic figures were rare in all sections examined. The interstitial epithelioid cells were immunoreactive with epithelial membrane antigen, vimentin, chromogranin, and thyroid transcription factor-1 (TTF-1) and were negative for Factor VIII-related antigen, CD34, and synaptophysin immunostains. There was no evidence of visceral pleura or lung parenchyma. Based on these results, the tumor was diagnosed as a SH isolated to the mediastinum.

The patient's postoperative recovery was uneventful. Five-years postoperatively, the patient was well with no evidence of recurrence.

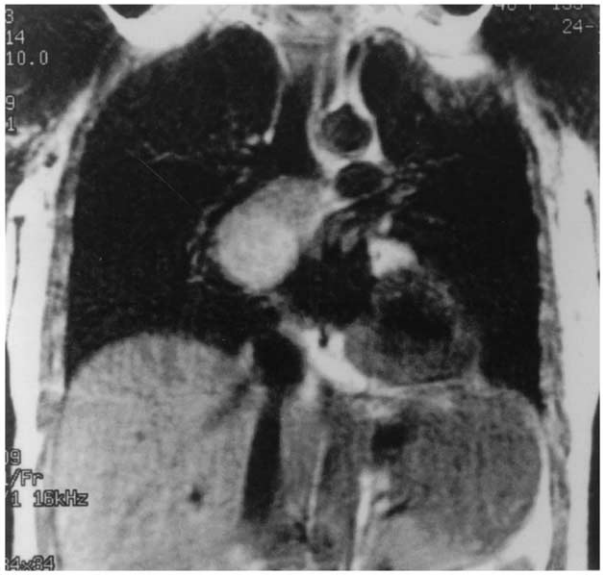

Fig 2. A coronal T1-weighted magnetic resonance image (TR/TE = 909/11) illustrating the tumor below the carina. The tumor compresses the right bronchus.

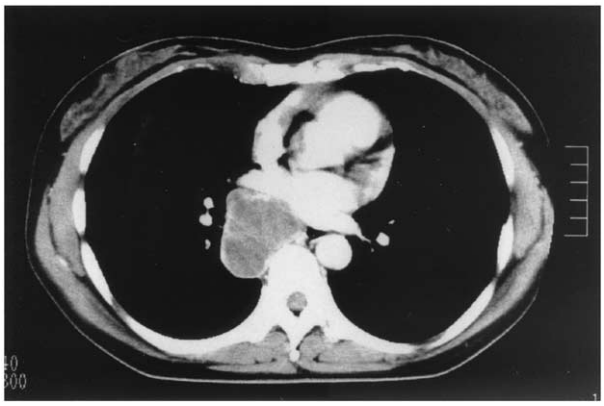

Fig 1. A computed tomographic scan of the chest, illustrating the mass in the posterior mediastinum.

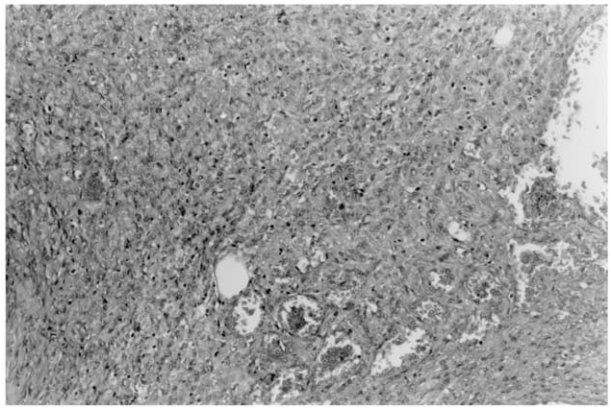

Fig 3. Microscopic features of the resected specimen. Solid and hemorrhagic patterns are observed. Sheets of round tumor cells with eosinophilic or clear cytoplasm are present in the solid structures. Hyperplastic type II pneumocytes line the surface of the cystic structures (hematoxylin and eosin stain, magnification ×100).

Comment

Liebow and Hubbell [3], the first to describe SH, suggested that this benign tumor was an endothelial proliferation; however, electron microscopic and immunohistochemical studies suggest that SH originates from epithelial cells [4, 5]. The exact histogenesis of SH thus remains uncertain [1, 5].

This is the first report to describe SH isolated to the mediastinum. One case of mediastinal SH was found among a series of 100 patients with pulmonary SH, but was considered to have arisen in the periphery of the lung and protrude into the mediastinum, mimicking mediastinal SH [6]. Many investigators consider that SH develops from type II alveolar epithelial cells, although this remains controversial. Positive TTF-1 staining supports the hypothesis that the tumor originated from respiratory epithelial cells [6]. In our patient, however, the tumor developed outside the lung.

Three mechanisms may account for the development of mediastinal SH. The first is metastasis from the lung to the mediastinum, although there was no evidence of tumor in the lung. All previously reported cases of lymph node metastasis from SH had evidence of primary tumor measuring at least 3.5-cm diameter in the lung [2, 6, 7]. The second mechanism is that the tumor could have originated from ectopic lung tissue, such as bronchogenic cysts. However, no ectopic lung tissue was found in the surgical specimen. The final mechanism is that the SH developed from the lung as a pedunculated pleural mass that slowly pulled away from the lung surface.

All three of these potential mechanisms are plausible; however, none can satisfactorily explain the apparent extrapulmonary location of the SH in our patient. Although we consider this to be the first documented case of SH isolated to the mediastinum, immunohistochemical studies suggest that the tumor arose from lung tissue. Our report demonstrates that, although rare, SH should be considered in the differential diagnosis of mediastinal masses.

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Postpneumonectomy Syndrome in Single Lung Transplantation Recipient Following Previous Pneumonectomy

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Successful single lung transplantation following previous pneumonectomy has not been reported in the literature. We report a patient with cystic fibrosis who underwent left single lung transplantation following right pneumonectomy 13 years previously. The outcome was adversely affected by postpneumonectomy syndrome.

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Respiratory failure is the common cause of death in patients with cystic fibrosis. Lung transplantation is an accepted treatment when these patients present with end-stage lung disease. By the nature of the suppurative lung disease, bilateral lung transplantation is generally required to remove the infection, and this is usually achieved by performing a bilateral single lung transplant or a heart and lungs transplantation [1–3]. Exacerbation of respiratory infection and hemoptysis are not uncommon during the course of the disease and when there is massive hemoptysis pulmonary resection could be lifesaving. We report a patient with cystic fibrosis who previously had a right pneumonectomy for recurrent hemoptysis. Subsequently, 13-years later, she underwent a left single lung transplantation. Postpneumonectomy syndrome was a significant and challenging problem following the lung transplantation. She remains alive at 30 months post-transplant.

The patient is a 40-year-old white female with cystic fibrosis diagnosed at birth, who received no treatment until 21 years old. Patient underwent right upper and middle lobectomy when she was 24 years old for recurrent infection and pneumothorax. She underwent a completion pnuemonectomy for recurrent hemoptysis when she was 27 years old. Her respiratory symptoms progressed and, therefore, she was evaluated for lung trans-

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