Ann Thorac Surg
2003;75:1020–1

CASE REPORTS

Mucoepidermoid Carcinoma in Unilateral Hypoplastic Lung: A Rare Tumor in a Rarer Condition

Antonio E. Martin-Ucar, FRCS, and Gaetano Rocco, MD, FECTS

Department of Thoracic Surgery, Northern General Hospital, Sheffield, United Kingdom

We present a rare case of a mucoepidermoid carcinoma arising from a unilateral isolated hypoplastic lung treated with a left pneumonectomy. The rarity of the condition is described and both pathologies discussed.

(Ann Thorac Surg 2003;75:1020–1)

Unilateral isolated hypoplastic lung is a rare congenital abnormality. In young adults the pathology is found during radiologic evaluation of recurrent respiratory tract infections with or without hemoptysis. Associated pathologies can occur within the background of congenital pulmonary abnormalities.

A 23-year-old man of South-Asian origin presented to his general practitioner with a long-standing history of irritative cough, hemoptysis, and malaise. He recalled his first episode of hemoptysis at age 5 and his early youth was characterized by symptoms of recurrent respiratory tract infections. A chest radiograph revealed mediastinal deviation toward the left side with lung markings occupying the entire left pleural cavity. The right lung field was seen as normal.

A computed tomography (CT) scan of his chest and upper abdomen confirmed the mediastinal shift but also revealed the presence of a partly aerated small left lung and compensatory enlargement of the right lung that occupied most of the left hemithorax (Fig 1). No associated abnormality was detected in addition to the hypoplastic left lung. The vascular supply and drainage of the lung appeared small but with normal anatomy when intravenous contrast was injected.

The investigations were completed with fiberoptic bronchoscopy and simultaneous bronchogram under sedation, showing a hypoplastic left lung with a patent although smaller left bronchial tree and a partly aerated parenchyma (Fig 2). Owing to the abnormal appearances of the bronchial mucosa, biopsies were performed during the investigation. The pathologic examination revealed mucoepidermoid carcinoma.

As there was no evidence of extrapulmonary malignancy in the workup and the patient fulfilled the operability criteria with an FEV1 of 2.6 L, surgical treatment was elected. This consisted of a left pneumonectomy of the hypoplastic left lung through a 15-cm posterior thoracotomy as described by Overholt [1] with partial excision of the posterior angle of the fifth and sixth ribs. Although adhesions between the abnormal lung and both pericardium and parietal pleura were encountered, these were divided by a combination of blunt and sharp dissection, proceeding to a pneumonectomy dividing the pulmonary artery intrapericardially and the pulmonary veins and main bronchus in a standard fashion.

The patient's recovery was uneventful and he was discharged from hospital on the seventh postoperative day. At 6-week follow-up he remained well and was back to his normal activities. The histopathologic examination confirmed the presence of a completely excised mucoepidermoid carcinoma arising in the context of an hypoplastic lung (Fig 3). The nodal stations sampled did not shown metastatic deposits.

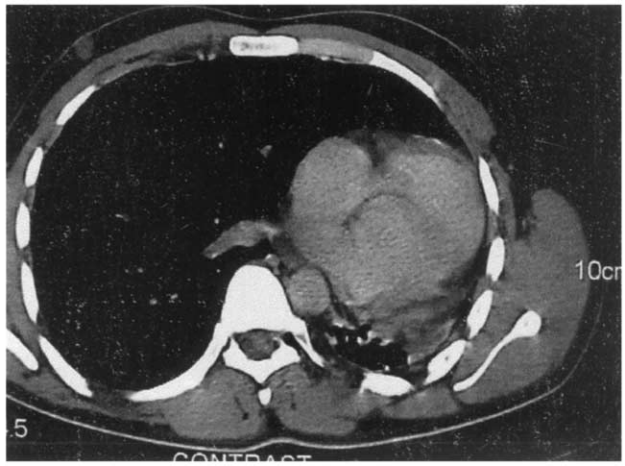

Fig 1. Computed tomography scan of the chest. The right lung occupies most of the left hemithorax with a dramatic mediastinal shift. A small aerated left lung lies posterior to the heart.

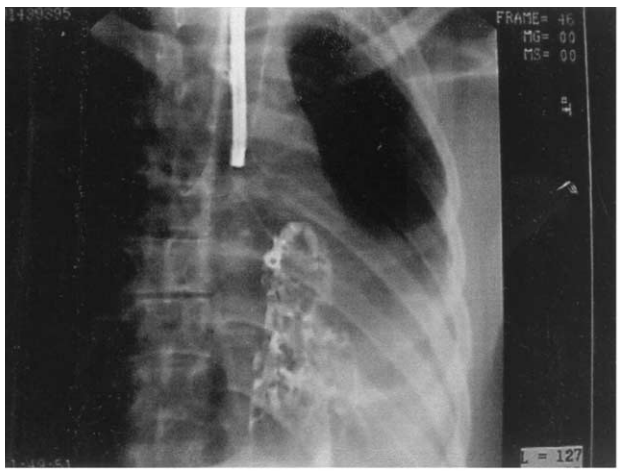

Fig 2. Bronchogram performed during a fiberoptic bronchoscopy shows a left lung hypoplasia. Biopsies from the left bronchial tree revealed mucoepidermoid carcinoma.

Accepted for publication Aug 22, 2002.

Address reprint requests to Dr Rocco, The Price-Thomas Unit, Northern General Hospital, Herries Road, Sheffield S5 7AU, UK; e-mail: gaetano.rocco@btopenworld.com.

Published by Elsevier Science Inc

0003-4975/03/$30.00
PII S0003-4975(02)04770-7

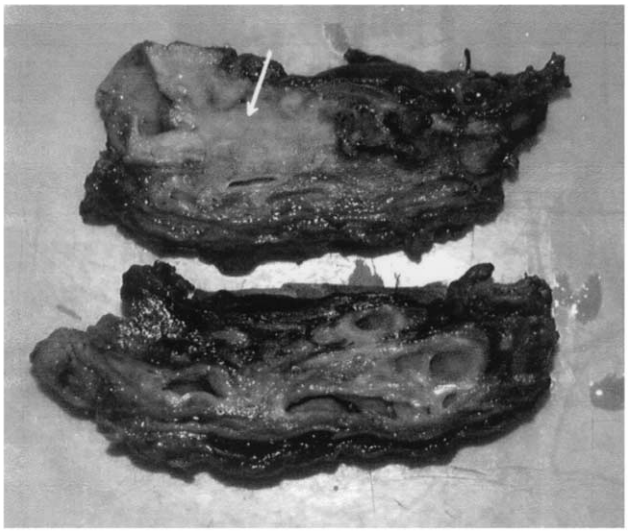

Fig 3. Pathologic specimen of the resected hypoplastic lung (11.6 × 7.5 × 3.4 cm) with the mucoepidermoid carcinoma (2.7 × 2.5 × 1.6 cm) indicated by the arrow.

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.

References

1. Overholt RH. The technique of pulmonary resection. Springfield, IL: CC Thomas, 1949.
2. Knox WF, Barson AJ. Pulmonary hypoplasia in a regional perinatal unit. Early Hum Dev 1986;14:33–42.
3. Page DV, Stocker JT. Anomalies associated with pulmonary hypoplasia. Am Rev Respir Dis 1982;125:216–21.
4. Phelan PD, Olinsky A, Robertson CF. In: Respiratory illness in children. Blackwell Scientific, 1994:357–93.
5. Hasleton PS. In: Spencer's pathology of the lung. McGraw-Hill, 1996:923–5.

Sclerosing Hemangioma Isolated to the Mediastinum

Kazuhiro Sakamoto, MD, Masato Okita, MD, Hiroshi Kumagiri, MD, Shunji Kawamura, MD, Koichiro Takeuchi, MD, and Riichiro Mikami, MD

Department of Respiratory Surgery, Yokohama Rosai Hospital, Yokohama, Japan

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.

(Ann Thorac Surg 2003;75:1021–3)

Accepted for publication Aug 24, 2002.

Address reprint requests to Dr Sakamoto, First Department of Surgery, Yokohama City University School of Medicine, 3-9 Fukuura, Kanazawa-ku, Yokohama 236-0004, Japan; e-mail: saka784@lycos.jp.

Published by Elsevier Science Inc

0003-4975/03/$30.00
PII S0003-4975(02)04365-5