Case Report

Surgical Treatment of Endobronchial Leiomyosarcoma with Right Main Bronchus Total Obstruction: A Case Report

Ming-Ching Lee, MD,[1] Chung-Ping Hsu, MD, FCCP,[2] and Jiun-Yi Hsia, MD, FCCP[1,3]

Endobronchial leiomyosarcoma is an unusual tumor of the respiratory tract. Clinically, patients may present with intermittent coughing, chest pain, dyspnea, hemoptysis, and fever until late in the course of the disease because of total obstruction of the main airway. In this paper, we report the case of a 51-year-old male with endobronchial leiomyosarcoma who presented with acute respiratory distress as a result of total obstruction of the right main bronchus and suffocation after massive hemoptysis. After intraoperative bronchoscopic assessment and bronchotomy, an elongated endobronchial tumor was found that arose from the right middle lobe (RML) bronchus with intraluminal extension upward into the right main bronchus. He underwent RML and right lower lobe (RLL) bilobectomy and had a rapid and uneventful recovery. (Ann Thorac Cardiovasc Surg 2008; 14: 105–108)

Key words: leiomyosarcoma, endobronchial, hemoptysis

Introduction

Primary endobronchial leiomyosarcomas are a rare malignancy in adults, with only 13 cases in patients more than 20 years old having been reported. To complete resection and prolong survival, sleeve lobectomy, pneumonectomy, or carinal resection may be necessary, depending on the anatomic location and size of the tumor. Herein we present a patient with a primary endobronchial leiomyosarcoma who presented with acute respiratory distress and was successfully treated with right middle lobe (RML) and right lower lobe (RLL) bilobectomy.

From [1]Division of Thoracic Surgery, Department of Surgery, Taichung Veterans General Hospital, Taichung; and [2]School of Medicine, National Yang-Ming University, Taipei; and [3]Chung Shan Medical University, Taichung, Taiwan

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Address reprint requests to Jiun-Yi Hsia, MD, FCCP: Division of Thoracic Surgery, Department of Surgery, Taichung Veterans General Hospital, No. 160, Sec. 3, Taichung-Kang Rd., Taichung, Taiwan.

Case Report

A 51-year-old man suffered from an intermittent cough, dyspnea, and painful sensations on the right side of the chest wall for one year. These symptoms worsened one week prior to admission and were accompanied by hemoptysis and intermittent fever up to 39°C. After visiting our emergency room, the patient was found to have acute respiratory distress and suffocation by hemoptysis, and oroendotracheal tube intubation with mechanical ventilator support was performed to oxygenate his blood. The chest radiography demonstrated almost total opacification of the right lung with a mediastinal shift to the same side (Fig. 1A). Computed tomography revealed an endobronchial tumor in the right main bronchus, causing complete atelectasis in the RML (Fig. 1, B and C) and partial collapse in the RLL and right upper lobe (RUL), secondary to bronchial occlusion. There was no evidence of an enlarged mediastinal lymph node or pleural effusion formation. Laboratory examination showed a hemoglobin level of 13.3 g/dL, leukocyte count was elevated to 15,800/mm^3, and C-reactive protein was 17.6 mg/dL. The arterial blood gas analysis revealed PaO_2 = 71 mmHg and $PaCO_2$ = 35.3 mmHg. Mycobacterium tuberculosis was

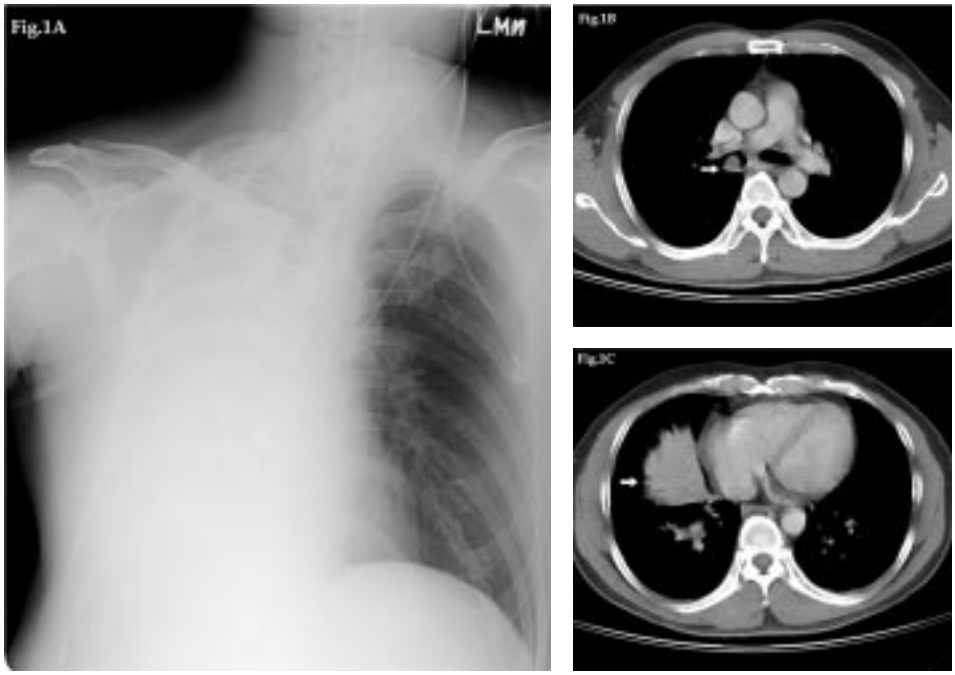

Fig. 1. (**A**) Anterior-posterior (A-P) chest X-ray reveals opacification of the right lung with mediastinal shift to right side. (**B**) Chest-computed tomography (CT) shows endobronchial tumor in right main bronchus (arrow). (**C**) Complete atelectasis in right middle lobe (RML) secondary to bronchial obstruction (arrow).

not found in the sputum smear or culture. Fiberoptic bronchoscopy showed that the orifice of the right main bronchus was totally obstructed by a reddish pedunculated tumor (Fig. 2), but the bronchoscopic biopsy was abandoned to prevent further tumor bleeding. Because of persistent tumor bleeding, surgical intervention was arranged and a right side pneumonectomy planned.

During exploration in right-sided postlateral thoracotomy, the right main bronchus seemed intact and the tumor seemed to be an endobronchial lesion arising from the right intermediate bronchus (RIMB). So a bronchotomy of RIMB was performed, and an 8×2×2 cm elongated whitish endobronchial tumor arose from the RML bronchus with an intraluminal extension upward into the RIMB until the right main bronchus was seen. The intraoperative bronchoscopic assessment revealed that no active lesion was noted in the right main bronchus, and the tumor infiltrated the peripheral RML bronchus. It also caused a consolidation of RML and RLL secondary to bronchial obstruction. Moreover, obstructive pneumonitis with purulent discharge in RUL was seen. After sucking out the purulent discharge from the RUL bronchial orifice by intraoperative bronchoscopy, the RUL was reinflated successfully. Thereafter, a bilobectomy of RML and RLL with an intercostal muscle flap buttress of the bronchial stump was performed. Upon microscopic examination, the endobronchial tumor was found to consist

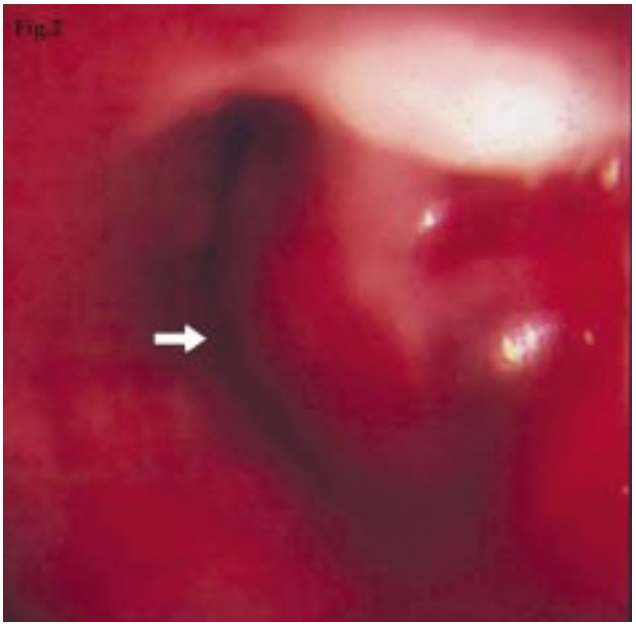

Fig. 2. On bronchoscopy, a reddish pedunculated tumor (arrow) obstructs the right main bronchus.

of spindle-shaped cells containing fusiform and hyperchromatic nuclei. Occasional mitotic figures were also observed under a high-power field (Fig. 3). Because the tumor cells surrounded the whole lumen of RML bronchus, including the membrane portion of bronchus, bronchial cartilage portion, and vessels surrounding bronchus, the definitive arising site of the tumor is difficult to identify. The immunohistochemistry stain showed moderately differentiated tumor cells expressing vimentin and smooth muscle actin (M851). These pathological features allowed the diagnosis of endobronchial leiomyosarcoma. The postoperative course was uneventful. After discharge, the patient was followed up periodically for 8 months, and neither distant metastasis nor local recurrence was found.

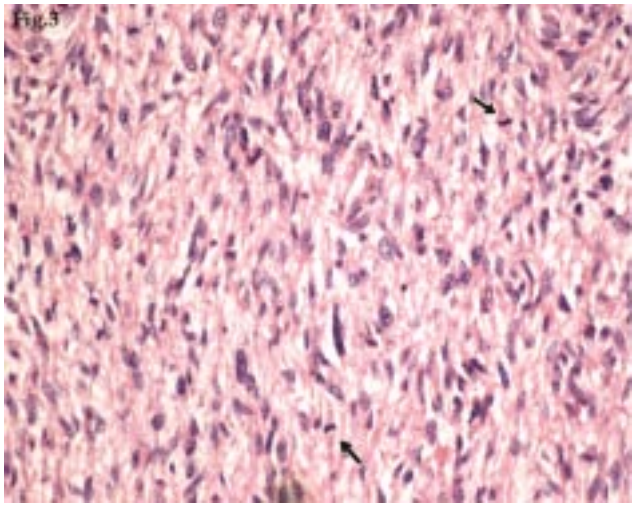

Fig. 3. Microscopically, the tumor consists of spindle cells containing fusiform and hyperchromatic nuclei. Occasional mitotic figures are also seen (arrows). H & E (×200).

Discussion

Primary endobronchial leiomyosarcoma is a rare malignancy of the lung. Since 1967, only 13 adult cases of it have been reported in relevant literature (Table 1).[1–12] These reports documented the disease in 7 males and 6 females, with ages ranging from 20 years to 69 years. Moreover, 16 cases of endobronchial leiomyosarcoma occurring in childhood have been reviewed and published by Takeda et al. in 2004.[13] Clinical symptoms are characterized by cough, fever, wheezing, hemoptysis, dyspnea, chest pain, and even expectoration of tumor fragments. Theoretically, chest radiographic findings of endobronchial tumors will present as obstructive pneumonitis, partial or total atelectasis of affected lung secondary to bronchial obstruction, and finally mediastinal shift resulting from lung volume reduction. In the presented case, these chest radiographic findings resulted from an elongated endobronchial tumor with a total obstruction of the right main bronchus. Furthermore, the total obstruction of the right main bronchus resulted in a mediastinal shift to the right side and led to the symptoms of acute respiratory failure.

A definitive preoperative diagnosis of endobronchial leiomyosarcoma is extremely difficult. In the literature reviewed, only five of 14 (5/14, 37%) cases had a definite preoperative diagnosis. One was proved by pathologic examination of expectoration of tumor fragment, and the others were achieved by bronchoscopic biopsy. Because bronchoscopic biopsy may yield insufficient material for diagnosis or cause bleeding, this procedure was abandoned in our case.

The only curative therapy for primary leiomyosarcoma of the lung is radical resection. In the case of Giuseppe Muscolino et al., a local recurrence of endobronchial leiomyosarcoma was noted after Nd:YAG laser therapy 2 months later. This patient has stayed alive and disease-free 6.5 years after operation.[9] However, to save uninvolved pulmonary parenchyma and to preserve pulmonary function, intraoperative bronchoscopic assessment and bronchotomy are necessary.[14] In the presented case, the right upper lobe was rescued after the accumulated purulent was sucked out via bronchotomy and reinflated after bronchoplexy and the intercostal muscle flap without any air leakage. The patient was weaned from mechanical support immediately after the operation.

In conclusion, although primary endobronchial leiomyosarcoma is a very rare clinical entity, recognizing the existence of this disease is necessary. Surgical resection should be the favored method for definitive diagnosis and curative treatment.

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Table 1. Additional reported cases of endobronchial leiomyosarcoma in adults (>20 years old)

Author, year	Age/sex	Symptoms	Location	Diagnosis	Therapy	Outcome
Gale et al., 1967[1]	22/F	Cough, chest pain	LUL bronchus	Surgery	Tumor excision	Alive 30 months
Annamalai et al., 1971[2]	21/F	Cough, pain, fever dyspnea, chest	LUL bronchus	Surgery	Pneumonectomy	Alive after discharge
Guccion et al., 1972[3]	20/M	Hemoptysis, intractable pneumonia	Right main bronchus	No mention	Pneumonectomy	Died of surgical complications
Carlson et al., 1978[4]	One endobronchial leiomyosarcoma in three thoracic leiomyosarcomas					Died within one year
Yellin et al., 1984[5]	42/F	Cough, dyspnea	Left main bronchus	Surgery	Intrapericardial pneumonectomy	Alive 7.5 years
Tanaka et al., 1992[6]	55/M	Hemoptysis, chest pain	LLL bronchus		Lobectomy	Alive after discharge
Koizumi et al., 1995[7]	20/M	No symptoms	RML bronchus	Surgery	Lobectomy	Alive 5 months
Sugiyama et al., 1998[8]	59/M	Dyspnea	Left main bronchus	Surgery	Pnuemonectomy	Alive after discharge
Muscolino et al., 2000[9]	69/M	Cough	RUL bronchus	Bronchoscopic biopsy	RUL sleeve lobectomy	Alive 7 years
	62/F	Hemoptysis	Carina and right main bronchus	Bronchoscopic biopsy	Carinal resection with RUL lobectomy	Alive 6.5 years
Lee et al., 2001[10]	59/M	Fever, cough, expectoration of tumor fragment	RLL bronchus	Tumor fragment pathology	Lobectomy	Alive after discharge
Ferri et al., 2003[11]	61/F	Cough, body weight loss	LUL bronchus	Bronchoscopic biopsy	Pneumonectomy	Alive 24 months
Takakura, et al., 2004[12]	35/F	Fever, cough	RIMB	Bronchoscopic biopsy	RML and RLL bilobectomy	Alive after discharge
Present case	51/M	Cough, chest pain, hemoptysis	RML bronchus	Surgery	RML and RLL bilobectomy	Alive 7 months

F, female; M, male; LUL, left upper lobe; LLL, left lower lobe; RML, right middle lobe; RUL, right upper lobe; RLL, right lower lobe; RIMB, right intermediate bronchus.

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