

Intracardiac Extension of a Pulmonary Pleomorphic Carcinoma

Wei-Syun Hu,¹ Huey-Ming Lo,¹ Yu-Wung Yeh,² Ming-Jen Lu,³ Yei-San Hsieh⁴ and Tong-Jong Chen⁵

Pleomorphic carcinoma is an extremely rare histological type of lung malignancy. Although it carries an aggressive clinical course, intracardiac extension has never been reported before. We describe a 53-year-old lady presented to our emergency department with progressive shortness of breath and intermittent fever for six weeks. After detailed work-up, a lung tumor with intracardiac extension via pulmonary vein to the left atrium was disclosed. Pathologic examination of a computerized tomography-guided lung biopsy showed pleomorphic carcinoma. The patient underwent wide excision of the left atrial tumor and subsequent right pneumonectomy smoothly. Both the left atrial and lung tumors showed the same histologic picture of pleomorphic carcinoma. However, brain metastasis was disclosed three months after operation. The patient eventually died of severe sepsis six months later.

Key Words: Intracardiac extension • Left atrial tumor • Lung malignancy • Pleomorphic carcinoma

INTRODUCTION

Pleomorphic carcinoma is an extremely rare histological type of lung malignancy, mainly occurs in males who smoke heavily, and also pursues an aggressive clinical course.¹⁻⁴ However, direct intracardiac extension via pulmonary vein was never reported. We describe the first case of pleomorphic carcinoma of lung with direct intracardiac extension via pulmonary vein.

CASE REPORT

A 53-year-old housewife, a non-smoker, visited our

emergency department due to progressive shortness of breath, dizziness, and dry cough for about six weeks. Intermittent low-grade fever was also noted. Physical examination showed a body temperature of 37.8 °C, a regular pulse rate of 80 bpm, a respiratory rate of 18 breaths per minute, and a blood pressure of 118/55 mmHg. The other physical findings were unremarkable.

Hemogram revealed leukocytosis with left shift. Marked elevated C-reactive protein (24.7 mg/dl) was also noted. Chest radiography revealed a huge mass lesion located in the right upper lung field (Figure 1A). A computerized tomography (CT) scan of the chest showed a huge mass about 6 cm in size with lobulated border and necrotic cavities over the posterior segment of the right upper lung with direct extension to the left atrium via the right pulmonary vein (Figure 1B). Transesophageal echocardiography revealed a huge mass which almost completely filled the left atrium (Figure 2), protruding across the mitral valve into the left ventricle during diastolic phase.

Empirical antimicrobial agent was prescribed after a thorough septic work-up. Positron emission tomography scan revealed no evidence of distant metastasis. CT-guided biopsy of the right upper lung tumor was at-

Received: March 10, 2009 Accepted: May 26, 2009

¹Divisions of Cardiology; ²Chest Medicine, Department of Internal Medicine; ³Divisions of Cardiovascular Surgery; ⁴Chest Surgery, Department of Surgery; ⁵Department of Pathology, Shin Kong Wu Ho-Su Memorial Hospital, Taipei, Taiwan.

Address correspondence and reprint requests to: Dr. Huey-Ming Lo, Division of Cardiology, Department of Internal Medicine, Shin Kong Wu Ho-Su Memorial Hospital, No. 95, Wen Chang Road, Shih Lin District, Taipei, Taiwan. Tel: 886-2-2833-2211 ext. 2072; E-mail: m006450@ms.skh.org.tw

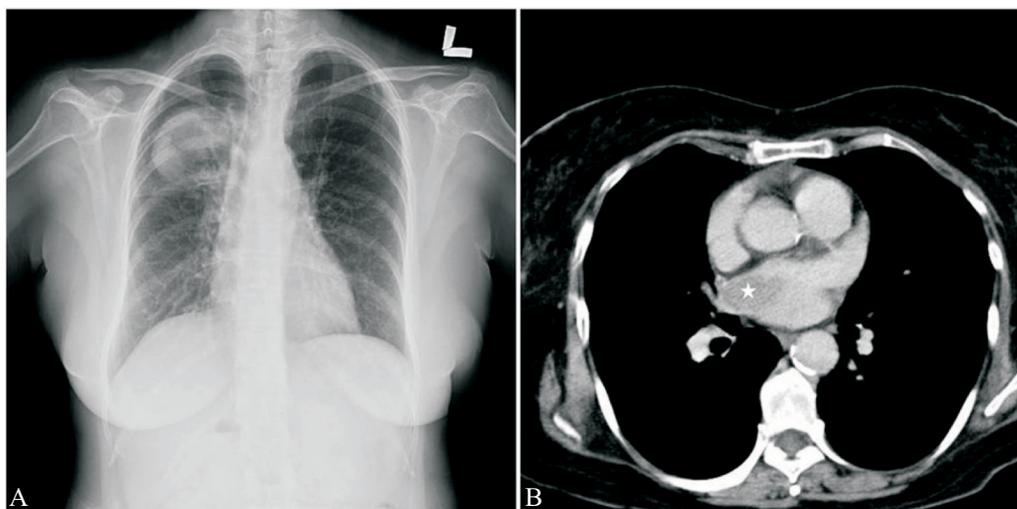


Figure 1. (A) Chest radiography shows a huge mass lesion located in the right upper lung field. (B) Chest computerized tomography scan shows a huge mass about 6 cm in size with lobulated border and necrotic cavities in the posterior segment of the right upper lung field extending through the right pulmonary vein into the left atrium as an intracardiac mass (asterisk).



Figure 2. Transesophageal echocardiography reveals a huge mass (asterisk), which almost completely filled the left atrium.

tempted, and pathologic examination was reported as pulmonary pleomorphic carcinoma. Surgical treatment was planned based upon the potential for life-threatening condition. On gross examination after longitudinal left atriotomy, one huge, yellowish and fragile tumor mass about 8×4 cm in size extending from the right superior pulmonary vein to the left atrium was noted. Wide excision of the left atrial tumor and subsequent right pneumonectomy was performed. Microscopically, the tumor was composed of neoplastic spindle-shaped and multinucleated giant cells bearing bizarre hyperchromatic and

pleomorphic nuclei, prominent nucleoli and abundant eosinophilic cytoplasm, favoring pleomorphic carcinoma. On immunohistochemistry, the tumor cells revealed positive reactivity for both cytokeratin and vimentin, with negative reactivity for desmin.

Progressive normalization of leukocyte count followed by improvement of fever was noted post-operatively. Tumor fever was impressed, and therefore antimicrobial agent was discontinued after three days of afebrile status. The patient tolerated surgery well and received adjuvant chemotherapy thereafter. Three months later, brain metastasis was disclosed via image study and she died six months later of severe sepsis.

DISCUSSION

According to the most recent World Health Organization histology classification of lung tumors, pleomorphic carcinoma is one subtype of sarcomatoid carcinoma. Pulmonary sarcomatoid carcinoma is rare, accounting for approximately 0.1-0.4% of all pulmonary malignancies.¹⁻⁵ It mainly occurs in male heavy-smokers, usually diagnosed in the fifth or sixth decade, and the prognosis is poor due to clinical aggressiveness.¹⁻⁴ Other subtypes of sarcomatoid tumor include giant cell carcinoma, spindle cell carcinoma, pulmonary blastoma and

carcinosarcoma based on their different histological characteristics.¹⁻⁵

Local invasion of regional lymph node, pleura, chest wall, or mediastinum as well as distant metastasis of bone, brain, lung, liver, or adrenal gland, have been reported in pulmonary pleomorphic carcinoma.^{1-4,6} To our knowledge, there was no previously published report about pulmonary pleomorphic carcinoma with direct intracardiac extension. Ng et al. reported a similar case of pulmonary sarcomatoid carcinoma with direct intracardiac extension.⁷ However, the pathologic examination of that case was carcinosarcoma which contained both spindle cell and adenocarcinoma component with atypical mitotic figures and necrosis. In contrast, our case showed a pleomorphic carcinoma which consisted of neoplastic spindle cells and multinucleated giant cells without adenocarcinoma component. Furthermore, on immunohistochemistry studies, the spindle cell component revealed positive reactivity for vimentin and the adenocarcinoma component for AE 1/3, Cam 5.2 and focally thyroid transcription factor-1 (TTF-1) in that case, whereas the present case had dual expression of cytokeratin and vimentin, showing the distinction between pleomorphic cell carcinoma and carcinosarcoma.

Lung cancer is one of the most common causes of cardiac metastasis; however, very few reports about direct intracardiac extension are documented. Pulmonary squamous cell carcinoma, adenocarcinoma, large cell carcinoma, adenosquamous cell carcinoma, mucoepidermoid carcinoma, atypical carcinoid tumor, leiomyosarcoma, carcinosarcoma and chondrosarcoma with direct cardiac involvement have been reported.⁷ This patient represents the first case of pulmonary pleomorphic carcinoma extending through pulmonary vein into the left atrium as an intracardiac mass.

Although the prognosis of pleomorphic carcinoma is usually disappointing once the diagnosis has been established, extensive surgical resection and post-operative adjuvant therapy may provide a means of extended survival. In cases with direct intracardiac extension, sudden

cardiac death due to systemic embolization or left ventricular inflow obstruction may occur if left untreated;^{8,9} surgical intervention may be helpful in the amelioration of patients' symptoms.

In summary, this is the first described case of pulmonary pleomorphic carcinoma with intracardiac extension to the left atrium. We would like to suggest that physicians should evaluate cases with pulmonary pleomorphic carcinoma carefully if there is direct intracardiac extension. Once direct intracardiac extension occurs, surgical intervention may be helpful in the amelioration of patients' symptoms.

REFERENCES

1. Mochizuki T, Ishii G, Nagai K, et al. Pleomorphic carcinoma of the lung: clinicopathologic characteristics of 70 cases. *Am J Surg Pathol* 2008;32:1727-35.
2. Yamamoto S, Hamatake D, Ueno T, et al. Clinicopathological investigation of pulmonary pleomorphic carcinoma. *Eur J Cardiothorac Surg* 2007;32:873-6.
3. Rossi G, Cavazza A, Sturm N, et al. Pulmonary carcinomas with pleomorphic, sarcomatoid, or sarcomatous elements: a clinicopathologic and immunohistochemical study of 75 cases. *Am J Surg Pathol* 2003;27:311-24.
4. Chang YL, Lee YC, Shih JY, Wu CT. Pulmonary pleomorphic (spindle) cell carcinoma: peculiar clinicopathologic manifestations different from ordinary non-small cell carcinoma. *Lung Cancer* 2001;34:91-7.
5. Travis WD. Pathology of lung cancer. *Clin Chest Med* 2002; 23:65-81.
6. Kim TH, Kim SJ, Ryu YH, et al. Pleomorphic carcinoma of lung: comparison of CT features and pathologic findings. *Radiology* 2004;232:554-9.
7. Ng SW, Lim CH, Wang CW, Chua YL. Primary pulmonary sarcomatoid carcinoma with intracardiac extension. *Jpn J Thorac Cardiovasc Surg* 2006;54:416-9.
8. Brandt RR, Rubin J, Reeder GS. Intracardiac extension of a lung tumor causing left ventricular inflow obstruction. *J Am Soc Echocardiogr* 1995;8:930-3.
9. Kodama K, Doi O, Tatsuta M. Unusual extension of lung cancer into the left atrium via the pulmonary vein. *Int Surg* 1990;75:22-6.