

A Long-Term Survivor with Late-Onset-Repeated Pulmonary Metastasis of a PEComa

Ryuichi Suemitsu, MD,¹ Sadanori Takeo, MD,¹ Noriko Uesugi, MD,² Yuzuru Inoue, MD,³ Motoharu Hamatake, MD,¹ and Masao Ichiki, MD³

A 59-year-old male underwent a surgical resection of a retroperitoneum tumor in 1990 that was diagnosed as leiomyoblastoma. Thereafter he demonstrated pulmonary metastases followed by a total of 3 pulmonary resections from 1995 to 1997. In 2008, he was incidentally found to have a tumor in the left lower lobe, which was diagnosed as a metastatic perivascular epithelioid cell (PEComa) neoplasm. Retrospectively, a primary tumor and pulmonary metastases were also diagnosed as PEComa. We experienced late-onset repeated pulmonary metastasis of a PEComa. (Ann Thorac Cardiovasc Surg 2010; 16: 429–431)

Key words: perivascular epithelioid cell neoplasm, pulmonary metastases, long-term survivor

Introduction

A perivascular epithelioid cell (PEComa) neoplasm is a rare soft tissue tumor that is morphologically characterized by epithelioid cells with clear or eosinophilic cytoplasm, and a perivascular distribution.¹ First described in 1992 by Bonetti et al.,² PEComas have since been reported to originate in many organs, especially in the uterus.³ At present, the details of the clinicopathological features, including survival, remain to be elucidated. We experienced a long-term survivor with pulmonary metastasis of a PEComa.

Case Report

A retroperitoneum tumor that compressed the stomach was incidentally identified in a 59-year-old male. There-

after the patient underwent a surgical resection of the tumor, which in 1990 weighed about 5 kg, and the tumor was diagnosed to be leiomyoblastoma. Thereafter he demonstrated pulmonary metastases followed by a total of 3 pulmonary resections from 1995 to 1997: a partial resection of the left upper lobe (S4 and S5) and the left lower lobe (S6 and S9s) to remove a total of 5 tumors in December 1995, a partial resection of the right lower lobe (S8) for 1 tumor in May 1997, and a partial resection of the left lower lobe (S6) for 1 tumor in December 1997. All resected lesions were diagnosed to be epidermoid leiomyosarcoma and thus were considered to be metastases from the retroperitoneum tumor. He was followed up without further metastasis until 2002. In April 2008 he experienced a sudden onset of chest pains, and an electrocardiogram was taken by his family physician; he was found to have acute myocardial infarction (AMI). He was admitted to our hospital, and chest computed tomography (CT) incidentally revealed an endobronchial tumor in S6 of the left lower lobe (Fig. 1A). After treatment for AMI, he was introduced to our department. Fiberoptic bronchoscopy revealed a polypoid lesion attached to the left lower bronchus originating from B6 (Fig. 1B), and a biopsy of the tumor revealed a PEComa metastasizing to the lung. He underwent a left lower lobectomy in July 2008, made an uneventful recovery, and was discharged on postoperative day 7.

From ¹Department of Thoracic Surgery, ²Department of Pathology, and ³Department of Respiratory Medicine, Clinical Research Institute, National Hospital Organization, Kyushu Medical Center, Fukuoka, Japan

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Address reprint requests to Ryuichi Suemitsu, MD, PhD: Department of Thoracic Surgery, National Hospital Organization, Kyushu Medical Center, Jigyohama 1–8–1, Chuo-ku, Fukuoka 810–8563, Japan.

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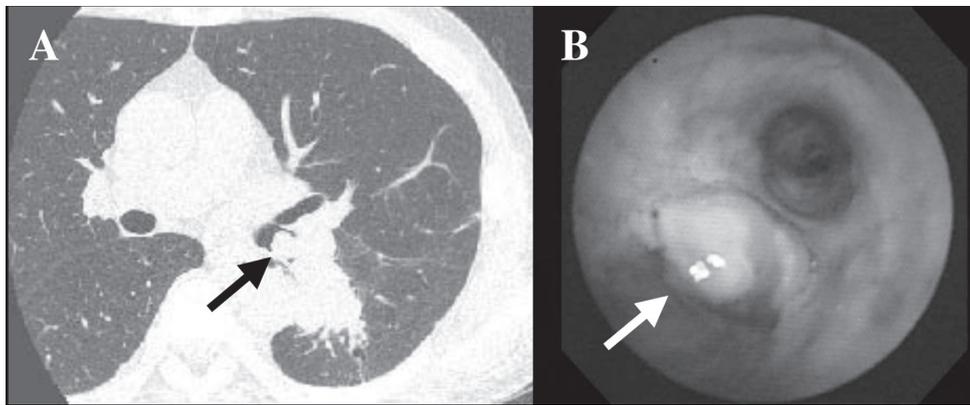


Fig. 1. (A) A chest computed tomography taken after treatment for acute myocardial infarction. An endobronchial tumor was identified in the left lower bronchus of the left S6 region. (B) The fiberoptic bronchoscopic findings show the left main bronchial cavity. A polypoid lesion was revealed in the B6 region, which almost completely obstructed the left lower bronchus cavity.

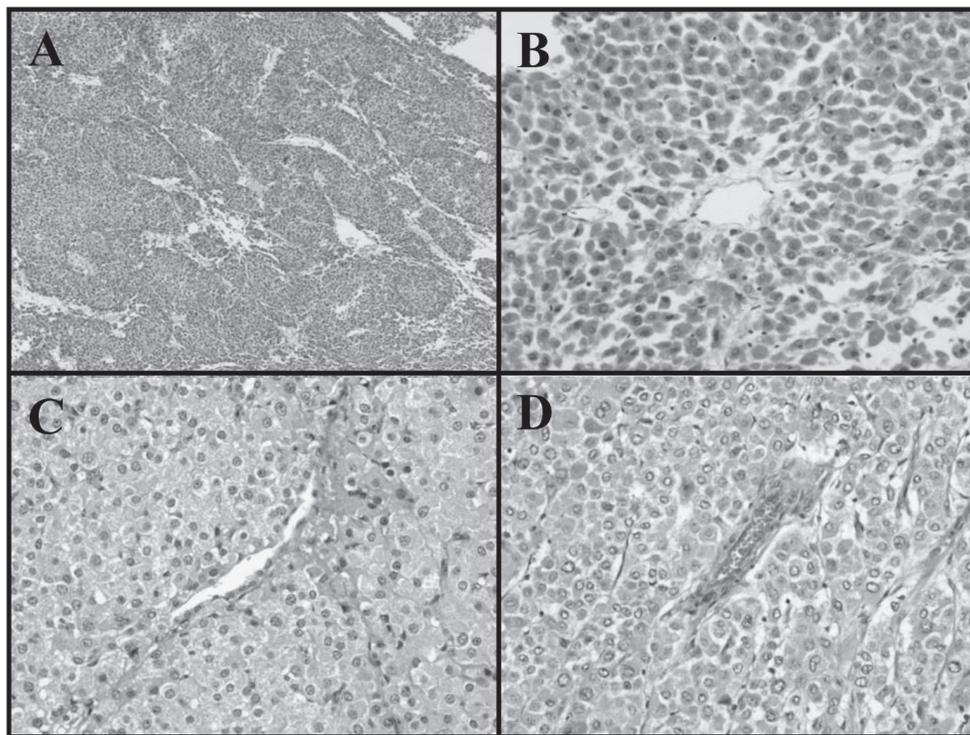


Fig. 2. Microscopic findings. All resected tumors revealed almost the same histological morphology as that of a PEComa, namely, perivascular epithelioid cells arranged with a blood vessel, eosinophilic cytoplasm, and a low mitotic rate. (A) Low-powered field and (B) high-power field of a primary retroperitoneum tumor. (C) The findings of one of the pulmonary metastases obtained in 1997. (D) High-powered field of the pulmonary metastasis in 2008. (A–D) Hematoxylin and eosin stain.

Discussion

PEComa was first defined in the WHO classification of soft tissue neoplasms in 2002, and it has been characterized as a mesenchymal tumor composed histologically and immunohistochemically of distinctive perivascular epithelioid cells, including angiomyolipoma, lymphangiomyomatosis, and clear-cell sugar tumors.¹⁾ However, the detailed characteristics of this tumor remain to be elucidated; furthermore there have been few reports of any long-term survivors with a PEComa. This case had a re-relapse of the lung 11 years after the last pulmonary metastasectomy and 18 years from the initial surgery of the primary tumor.

In this case we obtained past specimens from the previous hospital, and we reconfirmed these findings. Although the primary lesion of the retroperitoneum had been initially diagnosed in 1990 to be leiomyoblastoma initially based on the criteria of soft-tissue tumors, a retrospective microscopic reevaluation showed characteristics that were compatible with a PEComa (Fig. 2A and B). This tumor consisted of clear to lightly eosinophilic cells that were arranged into nests with a radial arrangement around the blood vessels. On the other hand, the diagnoses of resected pulmonary tumors from 1995 to 1997 were epidermoid leiomyosarcoma and were therefore considered to be metastases from the primary lesion that had

been treated at the previous hospital. Our retrospective microscopic findings were also compatible with all pulmonary metastases from the primary tumor (Fig. 2C). Although there was no clear concept regarding PEComas until 2002, a histological and immunohistochemical analysis of the pulmonary lesion, besides a reevaluation of the primary tumor and the previously treated pulmonary lesions, led to a final diagnosis of a metastatic retroperitoneum PEComa (Fig. 2D).

There have been no reports on the recurrence of PEComas in the late-onset period and especially regarding long-term survivors with aggressive treatment of metastasectomies. PEComas are low-grade malignant tumors that tend to recur over a long time, and our patient had previously undergone a total of 4 metastasectomies of the lung during an 18-year period. It has been reported that malignant factors of PEComas are large size, high degree of infiltration, high nuclear grade and cellularity, coagulative necrosis, and high mitotic activity.^{1,4)} However, salvage surgery such as repeated pulmonary resections

might make it possible for PEComa patients to have a long survival even if these tumors demonstrate malignancy.

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