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# Rapid growing pulmonary cavernous lymphangioma after chronic process for ten years



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## ABSTRACT

**INTRODUCTION:** Solitary cavernous lymphangioma is very rare disease characterized by abnormally proliferating lymphatic vessels. We report a 49-year-old woman with a cavernous pulmonary lymphangioma showing rapid growth after remaining indolent for 10 years.

**PRESENTATION OF CASE:** Chest computed tomography revealed a solitary, poorly demarcated mass in the left lower lobe; however, the tumour grew in size over the next 6 months. A left lower lobectomy was performed following suspected lung cancer. Histopathological and immunohistochemical analysis of the resected specimens revealed a pulmonary cavernous lymphangioma.

**DISCUSSION:** It is difficult to make an accurate diagnosis of solitary cavernous lymphangioma by imaging findings, therefore a surgical resection is recommended as the diagnostic and therapeutic modality.

**CONCLUSION:** A pulmonary lymphangioma should be included in the differential diagnosis of a rapidly growing tumour.

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## 1. Introduction

Lymphangioma is a rare and benign disease presenting as a congenital lymphatic malformation that arises because of a defect in bud development [1]. While thoracic lymphangiomas are usually observed in the mediastinum, pulmonary lesions are not very common. Pulmonary lymphangiomas are well known to frequently exhibit a cystic form on computed tomography (CT). Therefore, if a pulmonary tumour has a non-cystic appearance on the CT, it is important to differentiate it from lung cancer. Herein, we report a case of a rapidly growing solitary cavernous pulmonary lymphangioma mimicking a malignant tumour.

## 2. Presentation of case

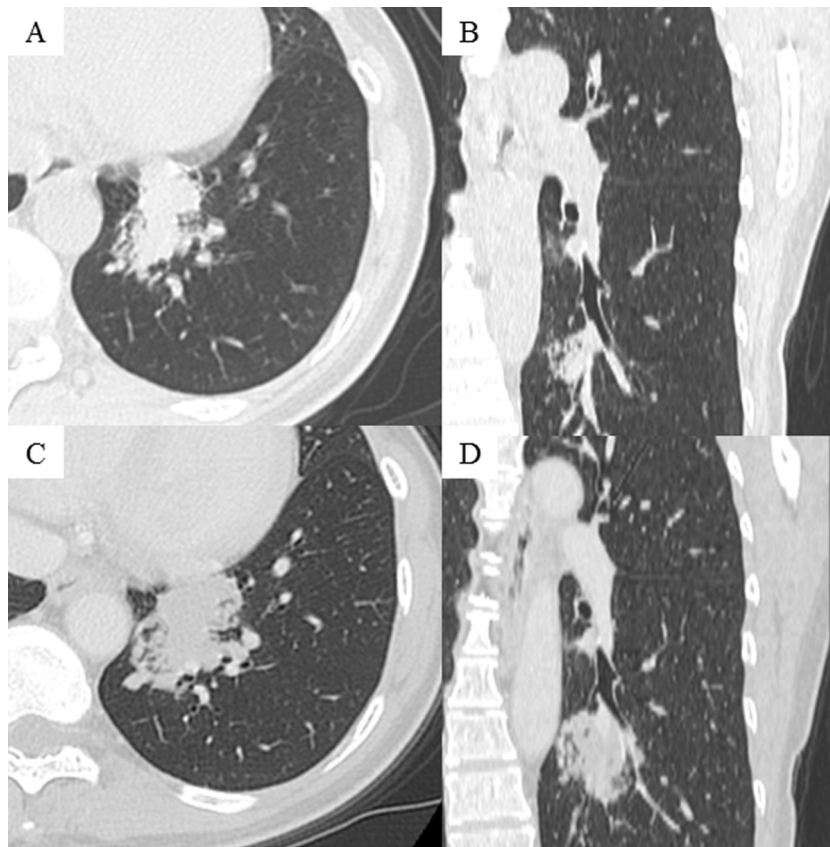
A 49-year-old woman who underwent left mastectomy for breast cancer 10 years ago had a suspicious lesion in her lung that was detected on preoperative chest CT. She did not receive the deep X-ray therapy for the treatment of breast cancer, but received adjuvant chemotherapy consisting of four courses of vinorelbine

(25 mg/m<sup>2</sup>) and docetaxel (60 mg/m<sup>2</sup>), followed by endocrine therapy (20 mg daily tamoxifen administration for five years and 3.6 mg monthly goserelin injection for three years). Although CT examinations performed every year revealed no changes in the abnormal mass during ten year-postoperative follow-up sessions, the lesion recently grew larger over a 6-month period. She was admitted to our outpatient clinic complaining of a dry cough and low-grade fever that manifested 1 month before the growth was detected on CT. She was a never smoker and had no family history of malignancy. Except for anaemia due to hypermenorrhoea, physical examinations and other laboratory tests (including for tumour markers) did not reveal any abnormalities.

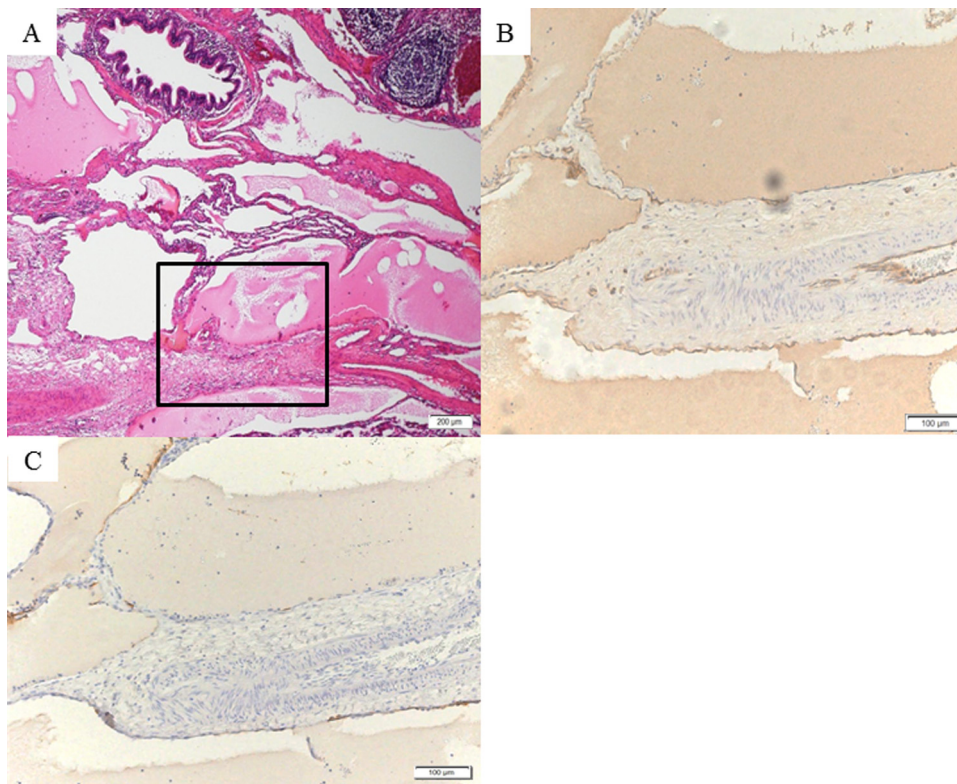
On her first visit of our hospital, the chest CT scan demonstrated a non-cystic poorly demarcated mass, 3.9 × 2.2 cm in size in the lower lobe of the left lung (Figs. 1A and B). Transbronchial lung biopsy did not result in a definite diagnosis. <sup>18</sup>F-fluorodeoxyglucose-positron emission tomography (PET) showed slight accumulation; the maximum standardized uptake value was 2.26. A subsequent CT scan after 6 months showed obvious tumour growth to 4.5 × 3.0 cm (Figs. 1C and D). After surgical resection of this tumour was indicated suspecting malignancy, the patient underwent a video-assisted thoracoscopic left lower lobectomy followed by mediastinal lymph node dissection. The mass was located on the root of the left inferior pulmonary vein, and intraoperative pathologic evaluation did not rule out malignancy.

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**Fig. 1.** Axial (A) and coronal sections (B) of high-resolution computed tomography (CT) scans of the lung demonstrating a poorly demarcated mass, measuring  $3.9 \times 2.2$  cm in diameter, in the lower lobe of the left lung. Axial (C) and coronal sections (D) of the subsequent CT scan 6 months later showing tumour growth, measuring  $4.5 \times 3.0$  cm.



**Fig. 2.** Histologic analysis shows multiple cystic spaces, containing lymph fluid, surrounding the bronchus (A). An immunohistochemical examination revealed that the monolayer epithelial cells lining the wall of the cyst were positive for cluster of differentiation antigen 31 and D2-40 (B and C, respectively). B and C are magnifications of the area indicated by the square in A.

Microscopic observation revealed spread cavernous spaces containing lymph fluid surrounding the bronchus (Fig. 2A). A single layer of endothelium lined the cyst wall. Immunohistochemical staining of these cells showed positivity for cluster of differentiation antigen 31 (CD31) and D2-40, which is a specific lymphatic vessel marker (Figs. 2B and C). The tumour was thus diagnosed as a pulmonary lymphangioma.

The postoperative course was uneventful; the patient was discharged on postoperative day 4 and is currently well with no signs of recurrence 2 years after surgery.

### 3. Discussion

Lymphangioma is a rare, benign vascular malformation of the lymphatic system composed of cystically dilated lymphatic vessels. While lymphangioma is known to affect any part of the body, the most common sites are the neck (75%) and axilla (20%).<sup>1</sup> Although it is not uncommon for thoracic lymphangiomas to be found in the mediastinum, where they account for 0.7% to 4.5% of all mediastinal tumours, pulmonary lymphangiomas are extremely rare [2]. The clinical manifestation of pulmonary lymphangioma is dependent on the age of the patient and the extent of the tumour. In adults, such tumours may remain asymptomatic and are found incidentally on chest radiography or CT, as occurred in the present case. Conversely, symptoms related to compressions of adjacent anatomic structures, such as cough, dyspnoea, vocal cord paralysis, venous compression, stridor, or dysphagia often occur in children.

Lymphangioma subtypes are histologically classified as cystic, cavernous, or capillary, and tumour development generally depends on the tissue density of the surrounding structure. Cystic lymphangiomas most commonly appear around fat tissue where there is little resistance to their growth; these lymphangiomas include cervical, axial, and mediastinal lesions. On the other hand, cavernous or capillary lymphangiomas often appear around muscular tissues, such as the lip, cheek, or tongue, where the resistance of the surrounding tissue is greater [3]. The majority of lung lymphangiomas are characterized by a cystic structure; solitary lung lymphangioma with a non-cystic appearance (as in the present case) appears to be rare. The rapid growth of the tumour may be owing to the softer parenchyma of the lung.

It is extremely rare for pulmonary lymphangiomas to exhibit rapid growth after remaining indolent for 10 years. Lymphangiomas appear to originate from the aberrant development of the lymphatic system, whereas secondary forms develop in adults owing to lymphatic channel obstruction caused by radiation, surgery, or infection [3]. Since histologic obstruction of the lymph vessel was not detected in this patient, it is reasonable to assume that an infection may have induced the tumour to grow, especially as slight accumulation was observed on PET.

Solitary lymphangioma of the lung is difficult to diagnose using imaging; according to a series of published reports, surgical resection is recommended for diagnosis and determination of the therapeutic intervention method [4,5]. The lymphatic properties of such tumours are difficult to clarify histopathologically, and the detection of immunohistochemical markers such as CD31, D2-40, and lymphatic vessel endothelial receptor 1 is useful for diagnosis of lymphangioma. D2-40 in particular is a more specific marker than the other proteins [6,7].

### 4. Conclusion

We experienced a rapidly growing pulmonary cavernous lymphangioma. Our case suggests that pulmonary lymphangioma should be included in the differential diagnosis of a rapidly growing pulmonary tumour, and surgical resection is recommended as the diagnostic and therapeutic modality.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Funding

None.

### Conflicts of interest

All authors have no competing interests.

### Ethical approval

None.

### Author contributions

Shinji Shinohara and Ryoichi Nakanishi conceived and designed the report. Manabu Yasuda contributed to the clinical management of the patients. Ryoichi Nakanishi, Manabu Yasuda and Fumihito Tanaka reviewed and revised the manuscript. All the authors read and approved the final manuscript.

### Guarantor

Shinji Shinohara.

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