



Progressive Massive Fibrosis Mimicking Lung Cancer: Two Case Reports with Potentially Useful CT Features for Differential Diagnosis

폐암으로 오인된 진행성 거대종괴성 섬유화:
두 건의 증례 보고 및 감별에 유용한 컴퓨터단층촬영 소견

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Progressive massive fibrosis (PMF) with atypical findings is often misdiagnosed as lung cancer. Atypical features of PMF have been described in some reports; however, these reports only introduced their cases with a short literature review. We report two cases of solitary PMFs with no underlying simple pneumoconiosis or rapid growth at atypical location that were mistaken for lung cancer. We also suggest the useful CT findings to aid in the differential diagnosis.

Index terms Computed Tomography, X-Ray; Pneumoconiosis; Lung Diseases, Interstitial; Silicosis

INTRODUCTION

Progressive massive fibrosis (PMF) is characterized by the coalescence of lung nodules to a mass larger than 1 cm in diameter (1-4), and usually appears as irregularly marginated, calcified, bilateral symmetric masses associated with paracicatricial emphysema in the posterior aspect of the upper lung (2, 3). Therefore, it is not difficult to diagnose PMF when typical radiological findings of PMF are shown. Meanwhile, PMF that shows atypical findings, such as when the mass does not show the typical location and findings of PMF or rapidly increases

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in size, is often misdiagnosed as lung cancer (5-7). Here, we report two cases that showed atypical solitary PMF, which were misdiagnosed as lung cancer and underwent surgery. In addition, we would like to suggest helpful CT findings for the diagnosis of PMF.

CASE REPORT

CASE 1

A 73-year-old male was admitted for evaluation of a lung nodule on chest radiography. He was asymptomatic and had no history of smoking. On admission, laboratory tests, physical examination, and pulmonary function tests were normal. Chest CT (Aquilion ONE; Canon Medical Systems, Otawara, Japan) showed a 19-mm-sized, irregularly marginated nodule in the periphery of the apicoposterior segment of the left upper lobe (Fig. 1A). The nodule was distributed in a row in the longitudinal direction and connected to the nodular bronchovascular bundle thickening on the coronal and sagittal reconstructed images (Fig. 1A). It contained no gross calcification and showed heterogeneous enhancement (20–40 Hounsfield units [HU] on pre-contrast images and 30–90 HU on post-contrast images) (Fig. 1A). In addition, multiple small nodules thought to be postinflammatory nodules were scattered in both upper lobes. There were no significantly enlarged lymph nodes, except for small calcified right mediastinal and hilar lymph nodes. The nodule showed hypermetabolism on ¹⁸F-fluorodeoxyglucose (¹⁸FDG) PET/CT (maximum standardized uptake [SUVmax] value, 5.4) (Fig. 1B). Left upper lobe thoracoscopic segmentectomy was performed for both diagnosis and treatment. The gross specimen demonstrated a black-colored 2 cm × 1.5 cm mass. In addition, histopathology showed concentrically arranged collagen bundles, consistent with the diagnosis of complicated silicosis with PMF (Fig. 1C). Later, the patient informed us that he had previously worked as a stonemason. At follow-up 1 month, the patient was asymptomatic.

CASE 2

A 74-year-old male with a history of working as a coal worker was admitted with complaints of cough and sputum for 1 year. He had a smoking history of 10 pack-years and had quit 40 years ago. Pulmonary function test revealed a moderate obstructive lung defect (forced expiratory volume in the first second/forced vital capacity [FEV1/FVC] = 52%), with normal lung volume and a mild decrease in diffusing capacity. Laboratory tests and physical examination results were normal. Chest CT (Aquilion PRIME; Canon Medical Systems) showed an irregularly marginated nodule in the subpleural zone of the right middle lobe with contiguous nodular bronchovascular bundle thickening (Fig. 2A). It showed an increase in size from 10 mm to 20 mm, compared to the previous CT 1 month ago (Fig. 2A). The nodule was distributed in a row in the axial direction on the coronal and sagittal reconstructed images (Fig. 2A), such as the nodule in Case 1. Associated fissural retraction was also visible. It contained focal microcalcifications and showed heterogeneous enhancement (20–80 HU on pre-contrast images and 20–140 HU on post-contrast images) (Fig. 2A). In addition, perilymphatic distributed micronodules suggesting pneumoconiosis were observed in both lungs, predominantly in both upper lobes (Fig. 2A). There were also multiple small to borderline-sized calcified lymph nodes in both the mediastinum and hilum. The nodule exhibited minimal hy-

Fig. 1. Case 1. Atypical, solitary progressive massive fibrosis mimicking lung cancer.

A. Axial thin-section (upper left panel) and pre- and post-contrast mediastinal setting CT images (upper middle and upper right panels) show a 19-mm, irregularly marginated nodule in the periphery of the apico-posterior segment of the left upper lobe. The nodule presents heterogeneous enhancement (20–40 HU on pre-contrast images and 30–90 HU on post-contrast images). Coronal (lower left panel) and sagittal (lower right panel) reconstructed images show the nodule distributed in a row in the longitudinal direction (arrowheads). Contiguous nodular bronchovascular bundle thickening is also noted, giving a radiating appearance on the sagittal image.

HU = Hounsfield units

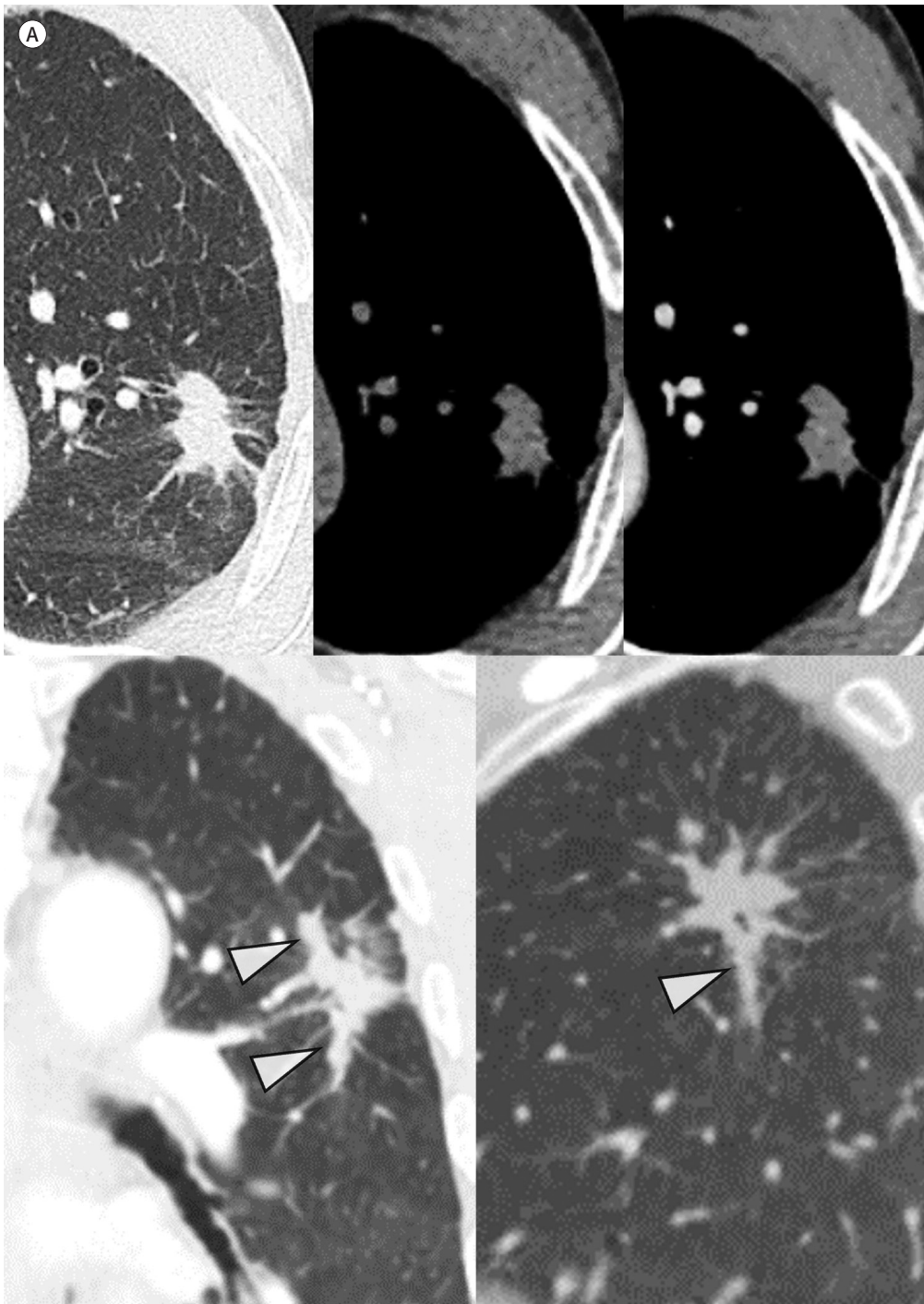
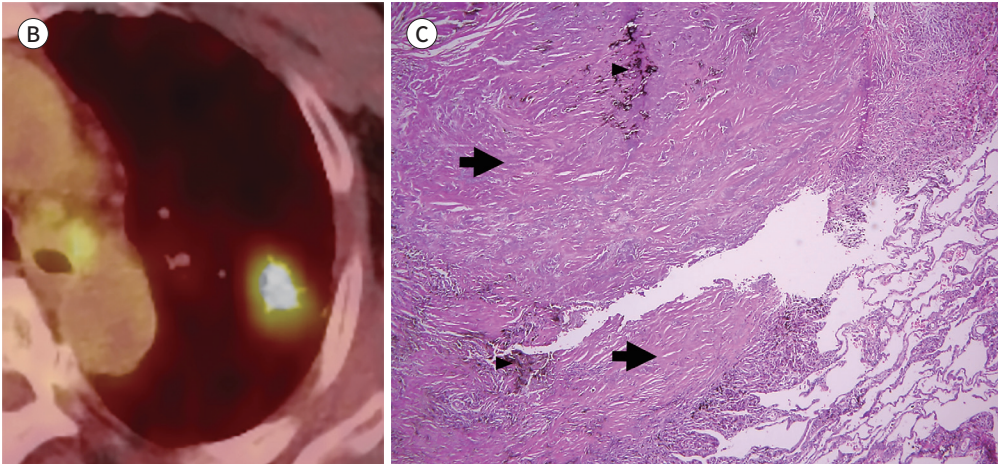


Fig. 1. Case 1. Atypical, solitary progressive massive fibrosis mimicking lung cancer.

B. ^{18}F -fluorodeoxyglucose PET/CT shows hypermetabolism in the nodule (maximum standardized uptake value, 5.4).

C. Histopathological examination shows that the hyalinized collagen bundles (arrows) are arranged concentrically with anthracotic pigmentation (arrowheads), consistent with complicated silicosis.



permetabolism on ^{18}F FDG PET/CT (SUVmax, 2.0) (Fig. 2B). Thoracoscopic right middle lobectomy was performed. The gross specimen described a 2 cm \times 3 cm mass with black pigmentation, and its histopathologic findings also suggested complicated silicosis with PMF (Fig. 2C). At 1 month of follow-up, the patient still had cough and sputum without any other new symptoms.

This retrospective study was approved by the Institutional Review Board of our hospital and the requirement for informed consent was waived (IRB No. WKUH 2021-06-031-003).

DISCUSSION

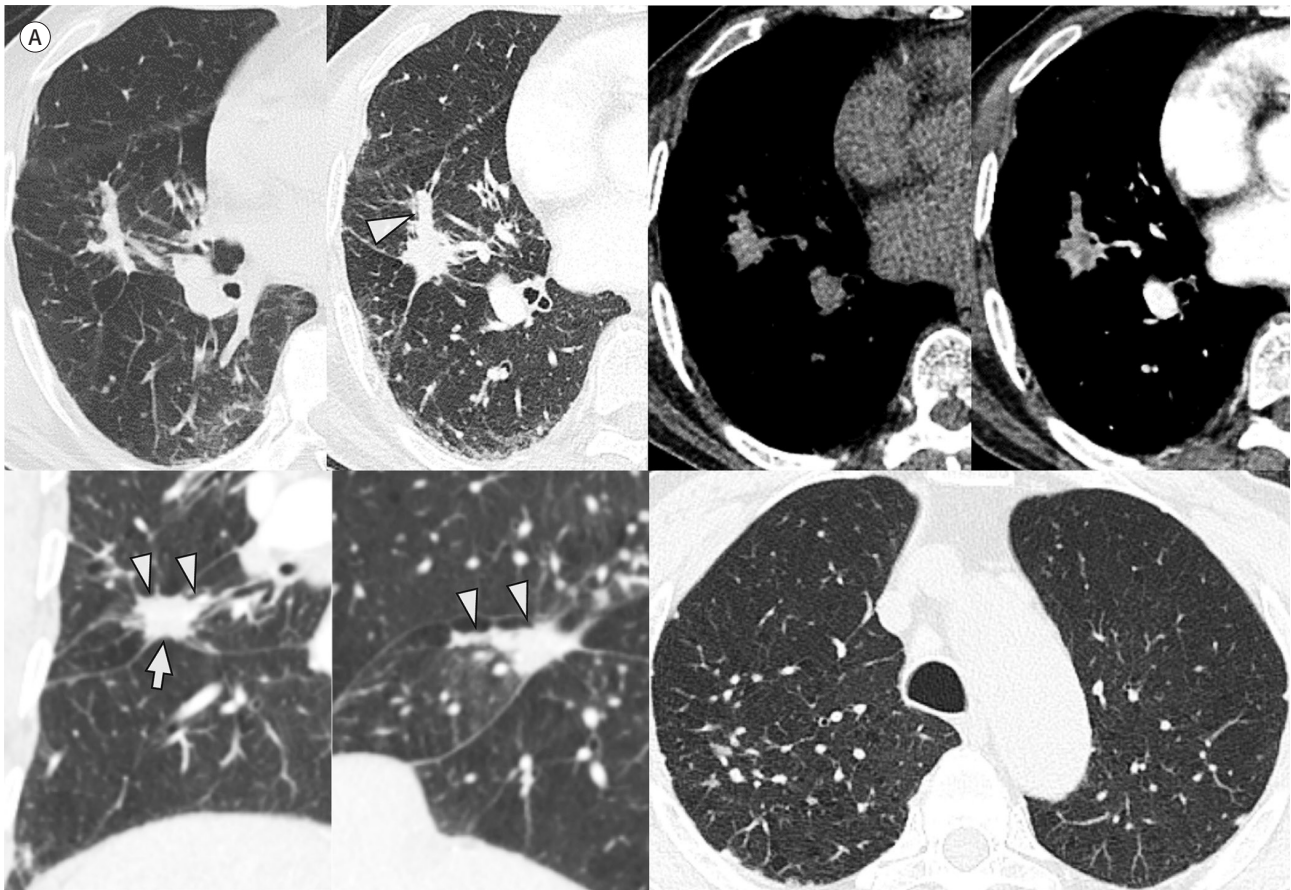
PMF is typically indicated by slow-growing, irregularly marginated and calcified symmetric bilateral opacities in the upper lung zone (2, 3). However, masses presenting atypical features of PMF often mimic lung cancer and subsequently require lung biopsy or surgical resection. These invasive procedures can induce hazardous conditions in patients with pneumoconiosis with impaired lung function. Therefore, noninvasive imaging diagnosis is important for differential diagnosis.

Atypical features of PMF have been described in some reports (5-7): an irregularly marginated, peripherally calcified, heterogeneously enhanced solitary PMF in the right upper lobe without underlying simple pneumoconiosis and occupational history; an irregularly marginated, heterogeneously enhanced solitary PMF in the left lower lobe without internal calcification, underlying simple pneumoconiosis, and associated history; a rapidly growing PMF with ischemic necrosis in the right lung of a patient with known pneumoconiosis. However, they only introduced their cases with a short literature review. Meanwhile, we would like to suggest useful CT findings to aid in differential diagnosis by reviewing two cases.

In the present cases, a solitary PMF from Case 1 showed no evidence of underlying simple

Fig. 2. Case 2. Atypical, solitary progressive massive fibrosis mimicking lung cancer.

A. Axial thin-section CT image (upper middle left panel) and pre- and post-contrast mediastinal setting CT images (upper middle right and upper right panel) show an irregularly margined nodule in the subpleural zone of the right middle lobe with contiguous nodular bronchovascular bundle thickening (arrowhead). The nodule presents heterogeneous enhancement (20–80 HU on pre-contrast images and 20–140 HU on post-contrast images) and an increase in size compared to that in the previous CT performed one month ago (upper left panel). Coronal (lower left panel) and sagittal (lower middle panel) reconstructed images show the nodule distributed in a row in the axial direction (arrowheads). Associated fissural retraction is noted on coronal image (arrow). In addition, distributed perilymphatic micronodules, indicative of pneumoconiosis, are observed on axial thin-section image at the aortic arch level (lower right panel). HU = Hounsfield units



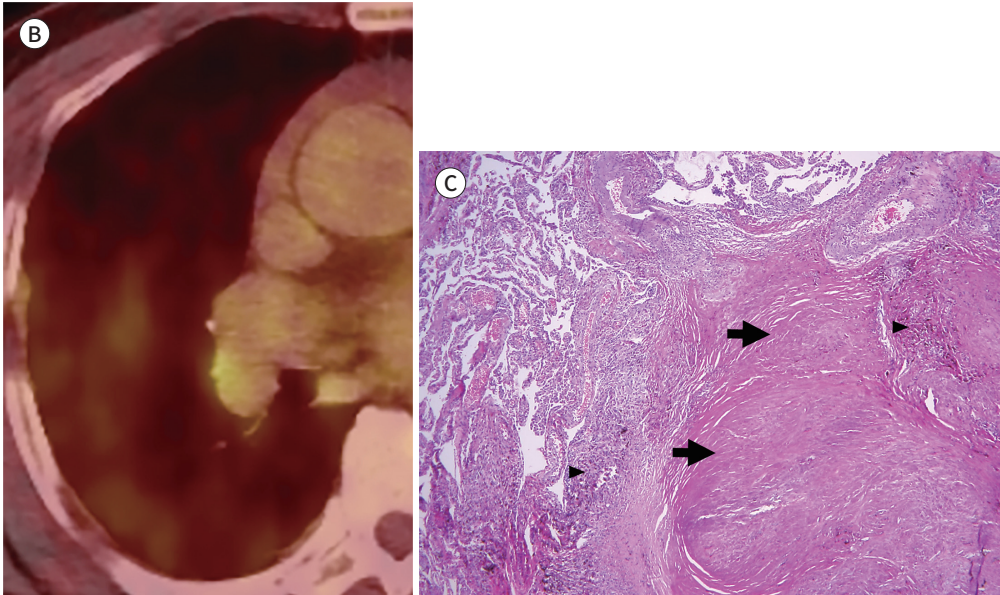
pneumoconiosis on CT, and that from Case 2 showed rapid growth and an atypical location in the right middle lobe with simple pneumoconiosis. PMF usually shows an increase in size with accompanying a decrease in profusion of surrounding smaller nodules (2). Occasionally, these large opacities rapidly grow with ischemic necrosis or combined tuberculosis (2, 7). On the other hand, in Case 2 showing rapid growth of PMF, there was no decrease of adjacent smaller nodules as well as no radiologic and pathologic evidence of combined necrosis, which made the correct diagnosis even more difficult. However, given that a PMF from Case 2 doubled in size during 1 month which is much shorter than the volume doubling time (529 days) of pulmonary adenocarcinoma (8), which might be helpful for differential diagnosis.

Since pneumoconiosis is caused by the disturbance of the lymphatic clearance of dust, it presents with nodular or irregular thickening of the axial and peripheral interstitium, reflecting the impairment of the central and peripheral lymphatic systems (1, 9). Both present cases

Fig. 2. Case 2. Atypical, solitary progressive massive fibrosis mimicking lung cancer.

B. ^{18}F -fluorodeoxyglucose PET/CT shows minimal hypermetabolism in the nodule (maximum standardized uptake value, 2.0).

C. Histopathological examination shows that the hyalinized collagen bundles (arrows) are arranged concentrically with anthracotic pigmentation (arrowheads), consistent with complicated silicosis.



showed that the masses were contiguous with nodular bronchovascular bundle thickening and distributed in a row in the axial or longitudinal direction. We believe that these ancillary CT findings are attributed to the thickening of the axial and peripheral interstitium, consistent with the finding of pneumoconiosis, and may be useful for differential diagnosis. Moreover, these features have been well demonstrated on coronal and sagittal reconstructed images, highlighting the importance of integrated interpretation of the mass using three-dimensional CT images.

In addition, we performed ^{18}F FDG PET/CT for preoperative staging or differential diagnosis. The nodule in Case 1 showed hypermetabolism, whereas Case 2 showed minimal hypermetabolism. Matthew et al. reported that PET imaging has a limited role in the diagnosis of malignancy in patients with pneumoconiosis due to its high rate of false positives (10), which is consistent with the findings in Case 1 that showed hypermetabolism on PET/CT but confirmed with PMF.

In conclusion, our cases suggest that radiologists should be familiar with both typical and atypical PMF findings. Integrated assessment of the mass distribution along the axial and peripheral interstitium using three-dimensional reconstructed CT images may help diagnose PMF mimicking lung cancer.

Author Contributions

Conceptualization, all authors; investigation, K.S.R.; supervision, R.J.Y.; writing—original draft, K.S.R.; and writing—review & editing, R.J.Y.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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REFERENCES

1. Kim KI, Kim CW, Lee MK, Lee KS, Park CK, Choi SJ, et al. Imaging of occupational lung disease. *Radiographics* 2001;21:1371-1391
2. Sirajuddin A, Kanne JP. Occupational lung disease. *J Thorac Imaging* 2009;24:310-320
3. Chong S, Lee KS, Chung MJ, Han J, Kwon OJ, Kim TS. Pneumoconiosis: comparison of imaging and pathologic findings. *Radiographics* 2006;26:59-77
4. Cox CW, Rose CS, Lynch DA. State of the art: imaging of occupational lung disease. *Radiology* 2014;270:681-696
5. Lee GB, Park HC, Hong KW, Lee WJ, Kim KI, Choi KM, et al. Localized pneumoconiosis manifested by solitary pulmonary nodule mimicking lung cancer: one case report. *Korean J Thorac Cardiovasc Surg* 2002;35:611-615
6. Jain AK, Nandy P. A solitary anthracotic lower lobe lung mass mimicking lung malignancy: an unusual presentation. *Egypt J Bronchol* 2016;10:82-84
7. Ahn BY, Baak YM, Chang HS, Kim JH, Kim KA, Lim Y. An unusual case of large opacity in pneumoconiosis. *J Occup Health* 2001;43:50-52
8. Park S, Lee SM, Kim S, Lee JG, Choi S, Do KH, et al. Volume doubling times of lung adenocarcinomas: correlation with predominant histologic subtypes and prognosis. *Radiology* 2020;295:703-712
9. Webb WR, Muller NL, Naidich DP. *High-resolution CT of the lung*. 5th ed. Philadelphia: Wolters Kluwer Health 2015
10. Reichert M, Bensadoun ES. PET imaging in patients with coal workers pneumoconiosis and suspected malignancy. *J Thorac Oncol* 2009;4:649-651

폐암으로 오인된 진행성 거대종괴성 섬유화: 두 건의 증례 보고 및 감별에 유용한 컴퓨터단층촬영 소견

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진행성 거대종괴성 섬유화는 비정형적인 소견을 나타낼 경우 종종 폐암으로 오인된다. 현재까지 비정형적인 소견을 나타내는 진행성 거대종괴성 섬유화에 대해 몇 개의 보고가 있으나 짧은 문헌 고찰과 함께 사례를 소개했을 뿐이었다. 이에 저자들은 컴퓨터단층촬영에서 기저에 단순 진폐증이 없거나, 빠르게 커지는 비정형적 위치의 고립성 종괴로 보여 원발성 폐암으로 오인되었던 진행성 거대종괴성 섬유화 2예를 보고하고 감별에 도움이 되는 컴퓨터단층촬영 소견을 제시하고자 한다.

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