Pictorial Essay

J Korean Soc Radiol 2020;81(5):1109-1120
https://doi.org/10.3348/jksr.2019.0165
pISSN 1738-2637 / eISSN 2288-2928

대한영상의학회지

Imaging Features of Various Benign and Malignant Tumors and Tumorlike Conditions of the Pleura: A Pictorial Review

흉막의 여러 가지 양성 및 악성 종양 혹은 종양 같은 질환들의 영상 소견: 임상 화보

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Received October 2, 2019
Revised November 12, 2019
Accepted November 21, 2019

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Pleural masses may be caused by various conditions, including benign and malignant neoplasms and non-neoplastic tumorlike conditions. Primary pleural neoplasms include solitary fibrous tumor, malignant mesothelioma, and primary pleural non-Hodgkin's lymphoma. Metastatic disease is the most common neoplasm of the pleura and may uncommonly occur in patients with hematologic malignancy, including lymphoma, leukemia, and multiple myeloma. Pleural effusion is usually associated with pleural malignancy. Rarely, pleural malignancy may arise from chronic empyema, and the most common cell type is non-Hodgkin's lymphoma (pyothorax-associated lymphoma). Non-neoplastic pleural masses may be observed in several benign conditions, including tuberculosis, pleural plaques caused by asbestos exposure, and pleural loose body. Herein, we present a review of benign and malignant pleural neoplasms and tumorlike conditions with illustrations of their computed tomographic images.

Index terms Pleura; Pleural Neoplasm; Computed Tomography, X-Ray

INTRODUCTION

Pleural tumors are not uncommon diseases but they can be quite challenging for some radiologists to differential diagnosis. Although there are various modalities for pleural evaluation, simple chest radiography and computed tomography is most commonly the first imaging technique. Therefore in this article, the authors would like to present common pleural diseases with typical radiologic findings.

BENIGN NEOPLASM

SOLITARY FIBROUS TUMOR OF THE PLEURA

Solitary fibrous tumor of the pleura (SFTP) is an uncommon, slow-growing mesenchymal tumor and may occur in benign and malignant forms (1). About 80% of SFTPs originate from visceral pleura, and 20% from the parietal pleura (2). Most patients are asymptomatic or have chest pain, cough or dyspnea. Patient may show extrathoracic manifestations including hypertrophic osteoarthropathy (4–35%) and hypoglycemia which is usually associated with very large tumors and is noted in less than 5% of patients (3).

Typical CT findings of SFTP are a well-defined or lobulated soft-tissue mass adjacent to the chest wall or within a fissure, frequently showing an obtuse angle with the pleural surface (1, 4). On contrast enhancement, SFTPs enhance intensely due to rich vascularization (5). Large tumors demonstrate heterogenous attenuation due to calcification, myxoid changes and areas of hemorrhage, necrosis, or cystic degeneration and form an acute angle with the pleura, mimicking a peripheral pulmonary mass (1, 4). CT findings that suggest a malignant form include a diameter larger than 10 cm, central necrosis, and ipsilateral pleural effusion (4). Differential diagnosis of SFTP can include nerve sheath tumors, pleural malignant mesothelioma and pulmonary neoplasms which show pleura based well-defined masses (Fig. 1).

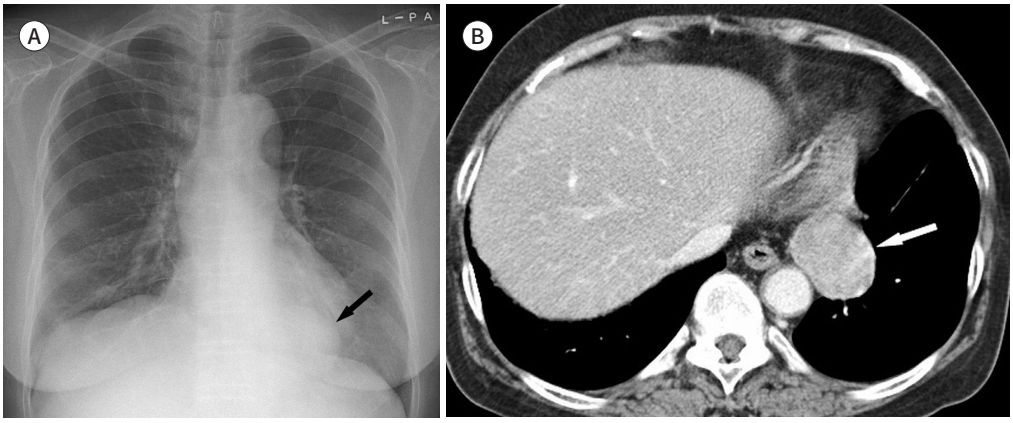
PRIMARY MALIGNANT NEOPLASMS

MALIGNANT MESOTHELIOMA

Malignant pleural mesothelioma (MPM) is neoplasm arising from the mesothelial cells of the pleura, peritoneum, or rarely, the pericardium, and is associated with exposure to asbestos. Patients commonly show symptoms of chest pain, dyspnea, cough, and weight loss and the prognosis is poor, with a median survival time of 12 months after diagnosis (6).

Most typical CT findings of MPM are unilateral pleural effusion, pleural thickening or mass, and interlobar fissure thickening (Figs. 2, 3) (7). As the disease progresses, tumor en-

Fig. 1. Solitary fibrous tumor of the pleura in a 65-year-old woman.
A. Chest radiography shows a mass with smooth margin in the left retrocardiac area (arrow).
B. Axial contrast-enhanced CT shows a well-defined homogeneously enhancing soft tissue mass adjacent to the mediastinal pleura (arrow).



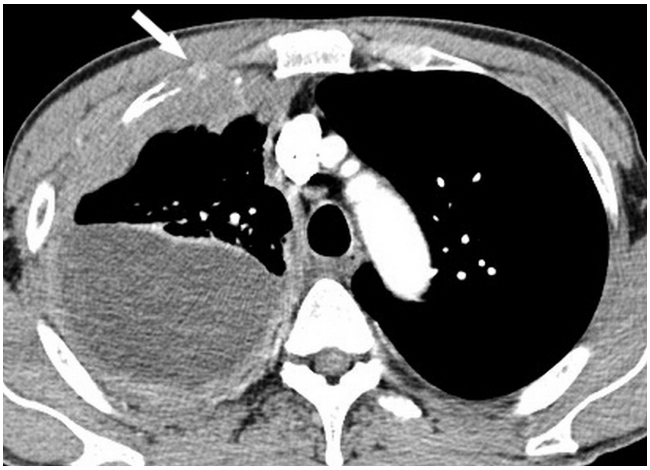


Fig. 2. Malignant mesothelioma in a 60-year-old man.
Diffuse irregular pleural thickening enveloping the right lung is noted with chest wall invasion and rib destruction (arrow).

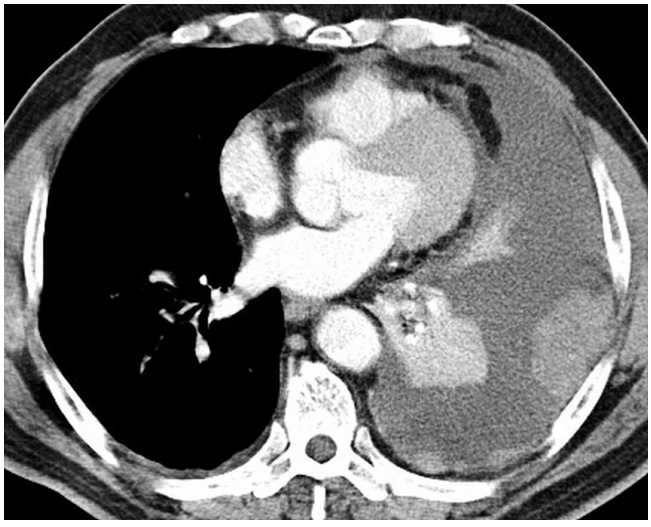


Fig. 3. Malignant mesothelioma in a 71-year-old man.
An enhancing pleural mass and nodules are noted in the left hemithorax with pleural effusion.

cases the lung and forms a pleural rind which leads to decrease of lung volume (6, 8). Also, as a result, contraction of the affected hemithorax with ipsilateral mediastinal shift, elevated ipsilateral hemidiaphragm, and narrowed intercostal space may be presented (6, 9). In some cases, calcified pleural plaques that represent asbestos-related disease are noted (9). MPM may extend to the chest wall, mediastinum, and diaphragm showing loss of fat and tissue planes. In rare cases, MPM may present with a localized pleural mass (Fig. 4). When advanced, it may metastasize to the hilar and mediastinal lymph nodes, and the lung manifesting as nodules and masses (6, 9). Differential diagnosis of malignant mesothelioma includes pleural metastasis from lung cancer or extrathoracic primary tumor. Although there are no differential CT features in pleural thickening, metastasis shows more frequent metastatic lymph nodes or metastatic lung nodules than malignant mesothelioma (10).

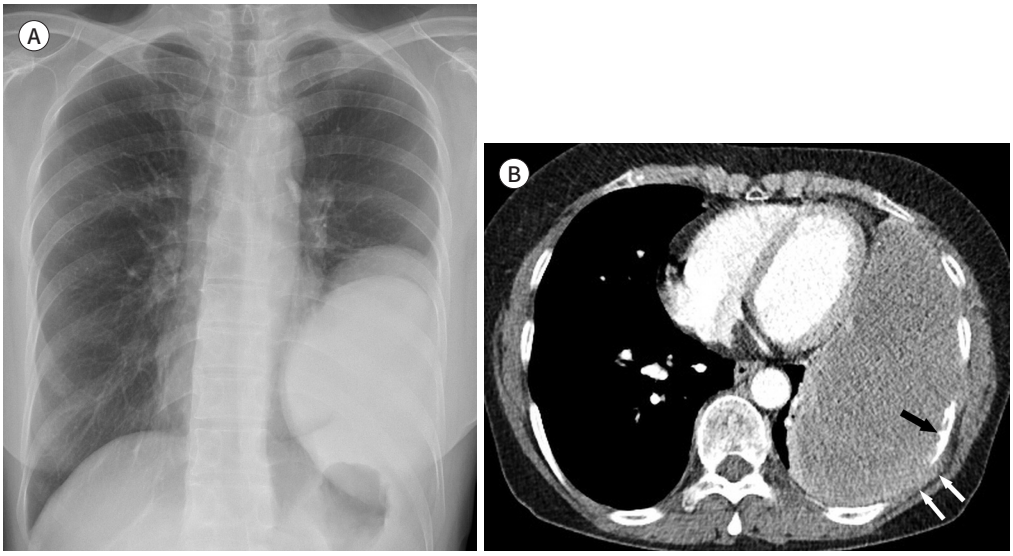
PRIMARY PLEURAL NON-HODGKIN'S LYMPHOMA

Primary pleural lymphoma is rare, although the pleural involvement is common manifestations of secondary lymphoma. Primary pleural lymphoma has a male predominance and

typically presents in middle-aged patients infected with Human Immunodeficiency Virus or harboring other immunocompromised states, such as recipients of solid-organ transplants, and in the elderly, often in human herpesvirus type 8 endemic areas. Epstein Bar virus co-infection is commonly found (11).

On CT scan, they usually show a lenticular or crescentic soft-tissue mass with diffuse nodular pleural thickening and have a tendency to invade adjacent structures. Also it can depict as isolated pleural effusion without any definite mass. A characteristic CT finding which helps differentiate primary pleural lymphoma from other pleural tumors is the "pleural sandwich sign" representing that the intercostal arteries are sandwiched in between pleural and extrapleural masses on axial contrast-enhanced CT images (Fig. 5) (12).

Fig. 4. Malignant mesothelioma in a 63-year-old woman.
A. Chest radiography shows a huge well-defined mass in left lower hemithorax.
B. Contrast-enhanced CT shows a large enhancing mass in the left lower hemithorax, with chest wall invasion (white arrows) and destruction of the adjacent rib (black arrow). Malignant mesothelioma of the sarcomatoid type was confirmed on percutaneous needle biopsy.



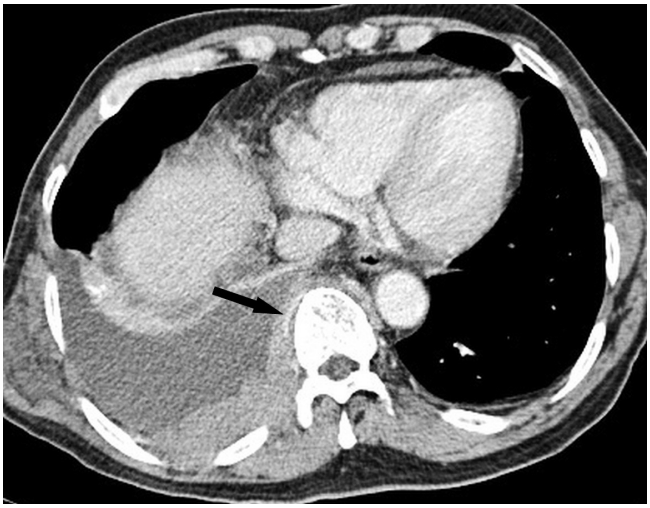


Fig. 5. Primary pleural lymphoma in a 68-year-old man.
The "pleural sandwich sign" is noted as enhancing pleural and extrapleural masses enveloping the intercostal artery (arrow) associated with pleural effusion.

SECONDARY MALIGNANT NEOPLASMS

METASTASIS

Pleural metastasis accounts for the vast majority of pleural malignant lesions. About 40% of pleural metastasis is from lung cancer, 20% from breast carcinoma, 10% from lymphoma, and 30% from other primary carcinomas (13).

CT findings of pleural metastasis of tumor show pleural effusion most commonly, pleural nodularity (Fig. 6), irregular pleural thickening (Fig. 7), and plaque which are rather rare (13, 14). In case of a lung cancer, ipsilateral pleural effusion to the primary lesion is common (Fig. 7). For other primary carcinomas however, it usually appears as bilateral effusion (15).

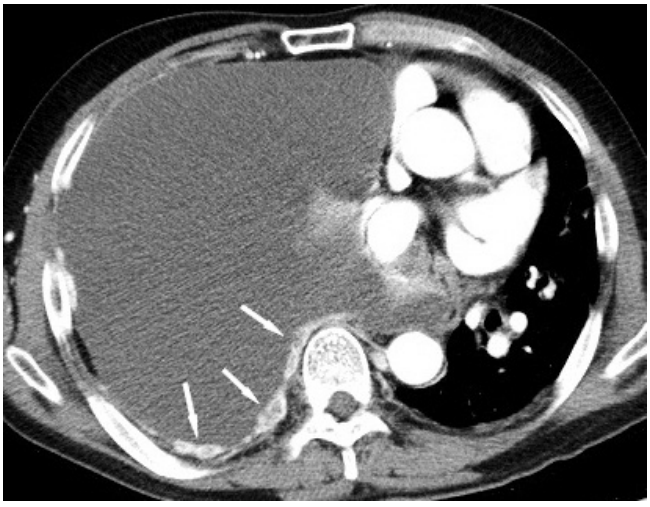
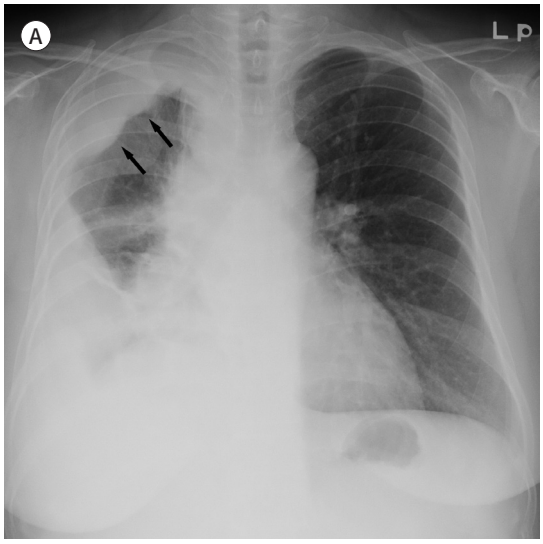
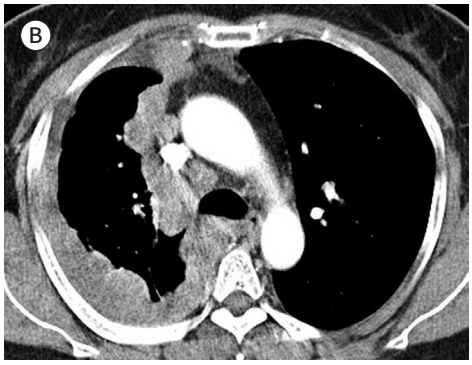


Fig. 6. A 55-year-old man with renal cell carcinoma and malignant pleural effusion.
Contrast-enhanced CT shows large right pleural effusion with multiple highly-enhancing pleural nodules (arrows) predominantly in the dependent area of the hemithorax.

Fig. 7. A 41-year-old woman with lung cancer and diffuse pleural metastasis.
A. Chest radiography shows right pleural effusion with an irregular pleural mass at the apex (arrows).
B. CT demonstrates a diffuse irregular enhancing pleural mass involving the mediastinal and costal pleuras.





LEUKEMIA

Leukemia is a hematologic malignancy which produces abnormal white blood cell in the bone marrow, and is classified as myeloid or lymphoid depending on the abnormal cell type. Most common leukemic involvement in the thorax is manifested as lymphadenopathy (Fig. 8A), but it can also involve the lungs, pleura, heart, and soft tissue. Pleural involvement most commonly occurs in acute lymphocytic leukemia (16).

Leukemic infiltration of the pleura can be depicted as pleural effusion, pleural masses or thickening, or both (Fig. 8B). Most frequent cause of pleural effusion in patients with leukemia is infection, followed by malignancy, and volume overload (16, 17).

MULTIPLE MYELOMA

Multiple myeloma is a hematologic malignancy which is characterized by proliferation of monoclonal plasma cells, and consequently producing a large amount of monoclonal immunoglobulins. Due to the overproduction of plasma cells and accumulation of abnormal immunoglobulins in the organs, patients may have low blood cell counts causing anemia and bleeding tendency, and show symptoms of bone pain, renal failure, neurologic symptoms and etc.

Fig. 8. A 36-year-old man with acute lymphocytic leukemia.
A, B. Contrast-enhanced CT shows bilateral axillary lymphadenopathies (arrows) (A) and bilateral pleural effusions with multiple enhancing pleural masses (arrows) (B).

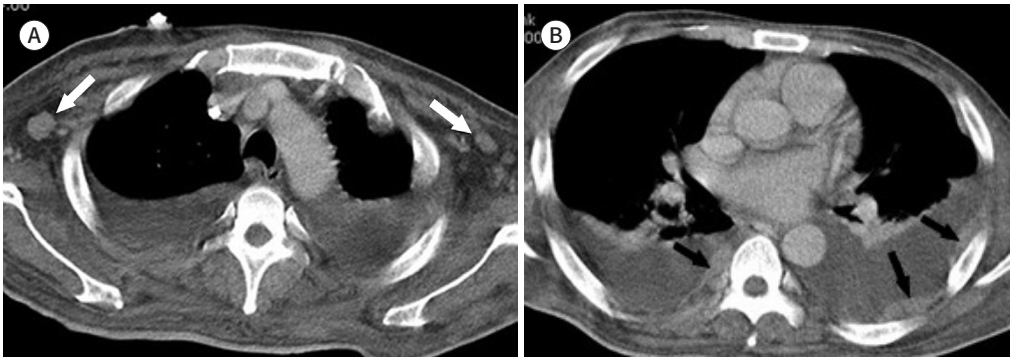
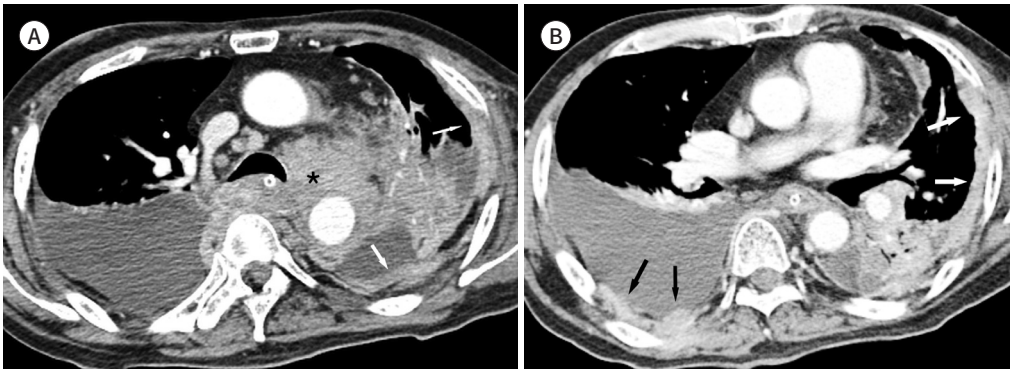


Fig. 9. A 49-year old man with multiple myeloma.
A, B. Contrast-enhanced CT reveals diffuse irregular pleural thickening in the left hemithorax (white arrows) and enhancing soft tissue masses in the mediastinum enveloping the aorta (asterisk) and extrapleural space (black arrows).



When the thorax is involved in multiple myeloma, it usually involves the bone or the lung parenchyme. In rare cases, pleural effusion is noted with heterogeneously enhancing irregular pleural thickening (18) or multifocal pleural thickening with mass-like lesions (Fig. 9) (19).

PYOTHORAX ASSOCIATED MALIGNANT NEOPLASMS

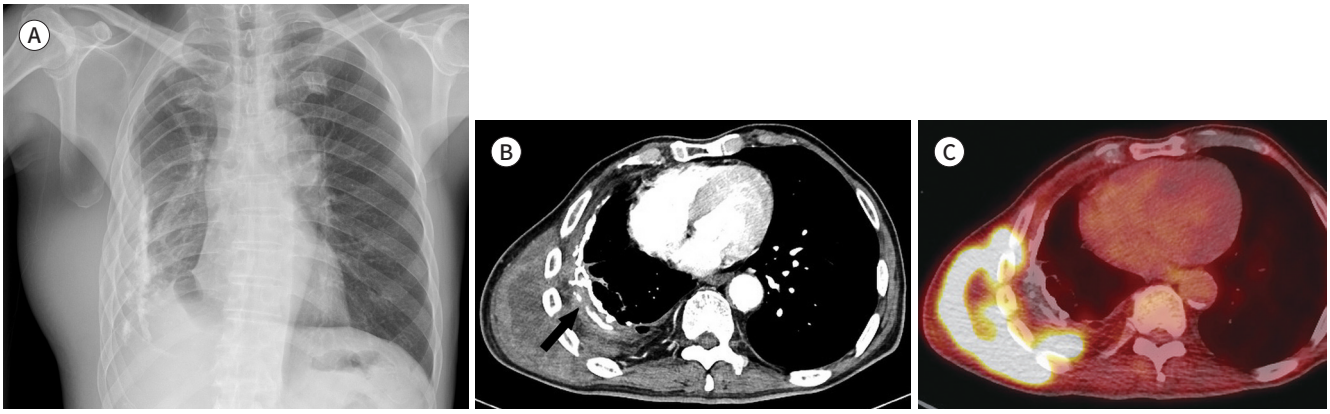
Malignant neoplasm is a rare complication of chronic empyema, and is likely to occur after more than 5 years of chronic empyema. Various malignant cell types can be associated with chronic empyema including malignant lymphoma (Fig. 10), squamous cell carcinoma (Fig. 11), malignant mesothelioma, and various sarcoma, etc (20). Malignant lymphoma is the most common cell type (pyothorax associated lymphoma; PAL) and most are non-Hodgkin's lymphoma. Radiologic findings include soft tissue mass with necrosis, rib destruction (Fig. 11), medial deviation of the calcified parietal pleura. Extensive necrosis, simulating empyema necessitatis, is common in PAL (Fig. 11) (21).

NON-NEOPLASTIC TUMORLIKE CONDITIONS

TUBERCULOSIS

Pleural tuberculosis may present with irregular or nodular pleural thickening simulating malignant pleural effusion (Fig. 12). Tuberculous pleurisy is usually caused by the rupture of subpleural caseous focus into the pleural space and as the infection progresses, chronic tuberculosis empyema takes place in the pleura (22). On CT images, pleural tuberculosis usually shows pleural effusion with uniform mild pleural thickening with enhancement, but sometimes demonstrates nodular pleural thickening, circumferential or mediastinal pleural involvement, or pleural thickening of more than 1 cm thickness (23) which is CT findings more suggestive of pleural malignancy rather than benign disease. Rarely, a layer of low attenuation

Fig. 10. A 76-year-old man with pyothorax associated lymphoma.
A. Chest radiography shows a soft tissue mass in the right lateral chest wall in association with chronic empyema.
B. CT reveals a chest wall mass showing central necrosis and peripheral enhancement, invading the extrapleural space. Focal breakdown of pleural calcification is noted in the pleural empyema (arrow).
C. ^{18}F-FDG PET/CT shows strong FDG uptake (maximum standardized uptake value: 28.3) in the enhancing portion of the mass on CT. Non-Hodgkin's lymphoma (diffuse large B cell) was confirmed on biopsy.
FDG = fluorodeoxyglucose

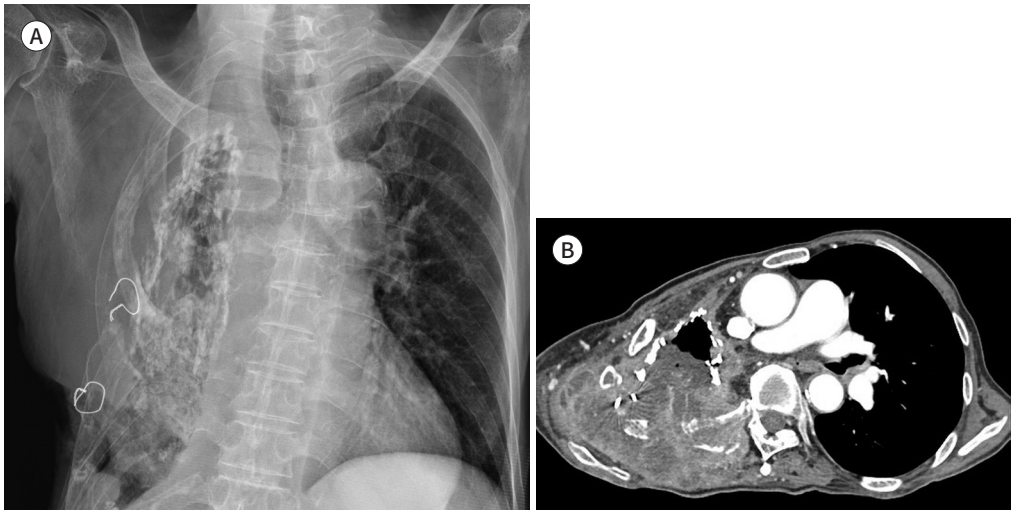


due to caseation necrosis is observed in the thickened parietal pleural peel (Fig. 12) (24).

ASBESTOS-RELATED PLEURAL PLAQUE

A pleural plaque is the most common form of the pleuropulmonary abnormality associated with asbestos exposure. It is considered to be a marker of asbestos exposure, indicating an increased risk of mesothelioma, lung cancer and asbestosis (25). Usually patients have no symptoms and may show slight decrease of lung function when a large extent of pleural is affected. On CT scan, pleural plaque is a localized pleural thickening of variable size and thickness and variable degree of calcification (Fig. 13). Patients usually have bilateral and multiple pleural plaques located along the chest wall, diaphragm and mediastinum, but rarely show a single plaque or unilateral hemithorax involvement. In terms of distribution, the upper ventral and lower dorsal chest walls, right diaphragm, and left mediastinum are more involved and costophrenic angles are generally spared (26).

Fig. 11. A 68-year-old man with pyothorax-associated squamous cell carcinoma.
A, B. Chest radiography (A) and contrast-enhanced CT (B) show a necrotic soft tissue mass in the right chest wall invading the extrapleural space and chronic empyema, with destruction of the adjacent ribs and pleural calcification. Histopathologic examination of the biopsy specimen revealed squamous cell carcinoma.



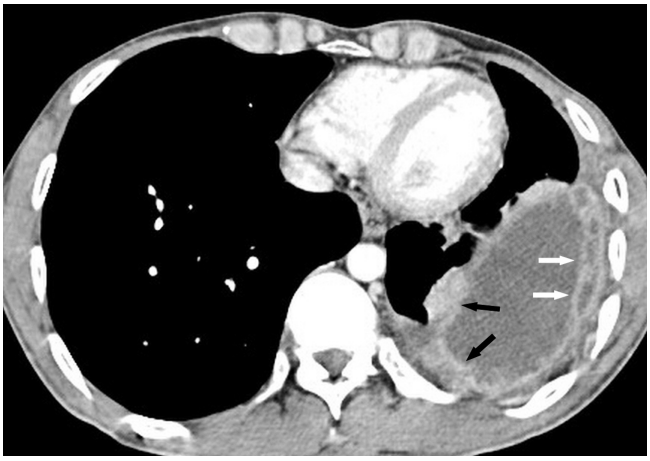


Fig. 12. A 33-year-old man with pleural tuberculosis.
Contrast-enhanced CT demonstrates left pleural effusion with nodular pleural thickening (black arrows) and increased attenuation in the adjacent extrapleural fat. A low-attenuation layer is noted in the thickened parietal pleura (white arrows).

Fig.13. A 63-year-old man with asbestos-related pleural plaques and malignant mesothelioma of the peritoneum.
A, B. Chest radiography and CT show bilateral multiple calcified pleural plaques along the chest wall and diaphragm.
C. CT of the upper abdomen shows ascites and peritoneal enhancement (arrow). Malignant mesothelioma was confirmed on peritoneal biopsy.

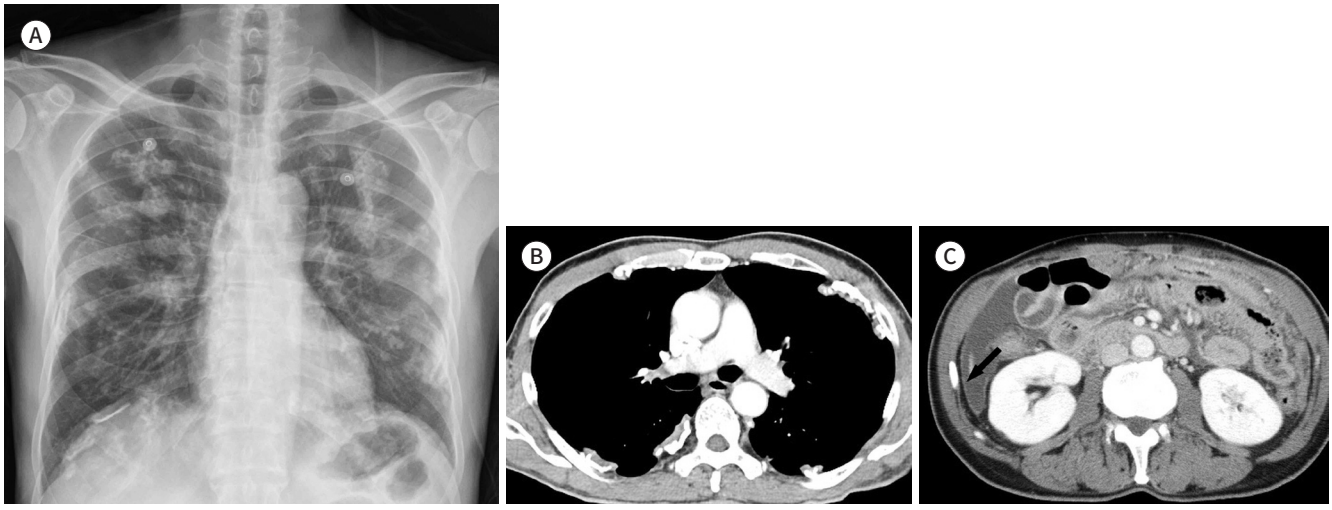
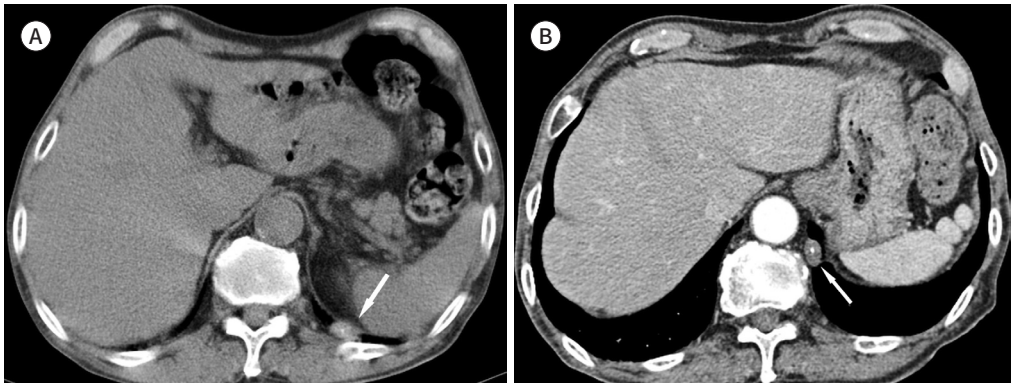


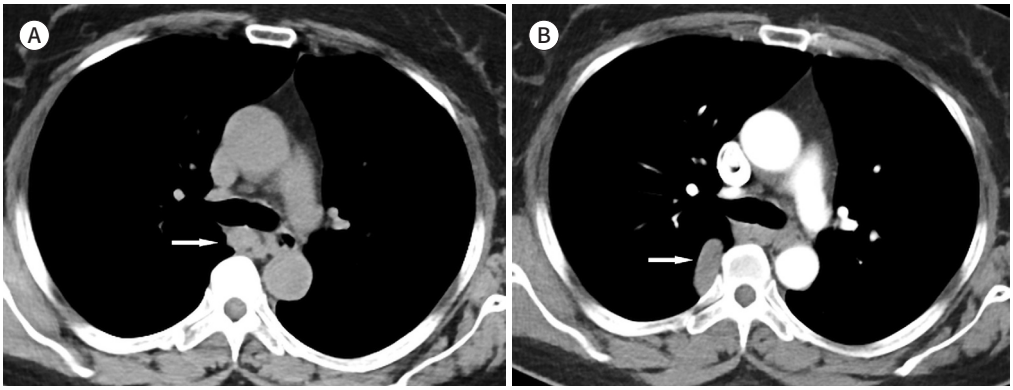
Fig. 14. A 76-year-old man with a pleural loose body.
A, B. Initial (A) and 5-year follow-up (B) chest CT scans show a migrating small soft tissue nodule with a tiny calcification in the left lower hemithorax (arrows).



PLEURAL LOOSE BODY

A pleural loose body, also known as 'thoracolithiasis' is an uncommon benign nodule, which moves freely in the pleural cavity. The etiology remains unclear and the histology of loose bodies consists of an outer wall of fibrous tissue and a variable central core which is usually fatty tissue with or without calcification. Generally it is found incidentally and is found at different locations in the pleura on serial radiographs or CT scans (27). On CT scans, pleural loose body usually shows a diameter of 5–15 mm, an ovoid nodule with smooth margin, and with or without calcifications (Figs. 14, 15). They tend to be located in the dependent portion of the pleural cavity due to gravity (28). When a small nodule moving freely in the pleural cavity is calcified, it can be easily diagnosed as thoracolithiasis. However, in some cases with little or no calcification, it may be difficult to differentiate with a benign pleural neoplasm. In this case, it might be helpful to diagnose pleural loose body if they show no contrast enhancement with mobile character (Figs. 14, 15) (27).

Fig. 15. A 58-year-old woman with a pleural loose body.
A. Pre-contrast CT shows a soft-tissue density mass in the subcarinal area (arrow).
B. Post-contrast CT shows that the mass migrated to the right paravertebral area (arrow). The attenuation value of the mass was around 40 Hounsfield units on both pre- and post-contrast enhanced images.



Author Contributions

Conceptualization, B.J.Y., K.Y.; data curation, all authors; investigation, all authors; methodology, all authors; project administration, K.Y.; resources, all authors; supervision, K.Y.; visualization, all authors; writing—original draft, B.J.Y., K.Y.; and writing—review & editing, B.J.Y., K.Y.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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대한영상의학회지

흉막의 여러 가지 양성 및 악성 종양 혹은 종양 같은 질환들의 영상 소견: 임상 화보

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흉막의 종괴는 다양한 양성과 악성 종양이 있고 종양은 아니지만 종양 같은 질환들에 의해 발생한다. 일차 흉막 종양에는 고립섬유종양, 악성중피종, 그리고 원발성 흉막 비호지킨 림프종이 있다. 흉막에 발생하는 가장 흔한 종양은 전이성 질환이며, 림프종, 백혈병, 다발성 골수종 등의 혈액 종양을 가진 환자에서 드물게 나타난다. 흉수는 주로 흉막의 악성 질환과 연관이 있다. 드물지만 흉막 종양이 만성 농흉에서 생길 수 있으며, 그중 가장 흔한 것은 비호지킨 림프종이다(농흉과 연관된 림프종). 종양이 아닌 흉막 종괴로는 다양한 양성 질환에서 보일 수 있으며, 여기에는 결핵, 석면 흉막판, 그리고 흉막 유리체가 포함된다. 이 임상화보에서 저자들은 흉막의 다양한 양성과 악성 종양 및 종양성 질환들에 대한 특징적인 전산화단층촬영 소견에 대해 알아보고자 한다.

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