

Solitary Fibrous Tumor of the Pleura Manifesting as an Air-Containing Cystic Mass: Radiologic and Histopathologic Correlation

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Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm that typically presents as a well-defined lobular soft tissue mass commonly arising from the pleura. We report an extremely rare case of an SFT containing air arising from the right major fissure in a 58-year-old woman. Chest CT showed an ovoid air-containing cystic mass with an internal, homogeneously enhancing solid nodule. To our knowledge, this is the first case in the literature. The histopathologic findings were correlated with the radiologic findings, and the mechanism of air retention within the tumor is discussed.

Index terms: Solitary fibrous tumor of the pleura; Air-containing mass; CT; Histopathology

INTRODUCTION

Solitary fibrous tumor (SFT) is a rare neoplasm that typically presents as a well-defined lobulated soft tissue mass commonly arising from the visceral pleura. On CT, small SFTs typically appear as well-defined, homogeneous, lobular, soft-tissue masses. Large SFTs are frequently appear heterogeneous before and after contrast injection due to myxoid change, hemorrhage, necrosis, or cystic degeneration (1-3).

However, to the best of our knowledge, no cases of SFTs containing air have been reported in the literature. Herein, we report an extremely rare case of an SFT manifesting as an air-containing cystic mass arising from the posterior visceral pleura enveloping the right upper lobe within the

right major fissure.

CASE REPORT

A 58-year-old woman was transferred to our hospital due to an incidentally identified lung mass at an outside hospital. She was a never smoker and was asymptomatic. Chest radiography showed an ovoid, thick-walled, air-containing cystic mass in the right upper lung zone where the mass was located eccentrically and surrounded by crescent-shaped air on the medial aspect (Fig. 1A). Chest CT demonstrated a 4.5 cm-sized, spindle-shaped cystic mass containing air within the superior portion of the right major fissure (Fig. 1B, C). This mass had an irregular thin wall with a thickness of several millimeters. A well-defined soft tissue nodule was eccentrically located within the lesion and measured approximately 3 cm. The nodule had a homogeneous density of 25 Hounsfield units (HU) on pre-contrast CT and was relatively homogeneously enhanced after intravenous contrast administration, with a density of 50 HU (Fig. 1D, E). Her previous chest CT images, performed at the outside institution approximately 2 years prior, revealed a same-sized cystic mass containing a much smaller internal soft tissue nodule compared to the lesion on the present CT images (Fig. 1F). F18-fluorodeoxyglucose (FDG) PET/CT

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demonstrated mild FDG uptake with a maximum standard uptake value (SUVmax) of 2.3 (Fig. 1G, H).

Video-assisted thoracoscopic surgery was performed to resect the tumor. An encapsulated ovoid cystic mass was shown in the pleural space, with a broad short pedicle and prominent blood vessels over the thin serosal lining of the tumor (Fig. 1I). The stalk arose from the posterior portion of the visceral pleura enveloping the right upper lobe within the upper portion of the right major fissure. Cut sections of a gross specimen revealed an air-containing cystic mass

with an eccentrically yellow-gray-colored solid nodule (Fig. 1J). Histopathologic examination with hematoxylin-eosin (HE) staining revealed spindle-shaped tumor cells mixed with collagen fibers in the tumor and its cystic wall without evidence of mitosis or nuclear pleomorphism, representing a benign SFT (Fig. 1K, L). There was cleft-like cystic air space surrounding the solid nodule, which was partially lined by cuboidal cells (Fig. 1L). Immunohistochemical studies of the tumor showed diffuse staining for CD34 and vimentin, and the cells lining the cystic portion of the tumor stained

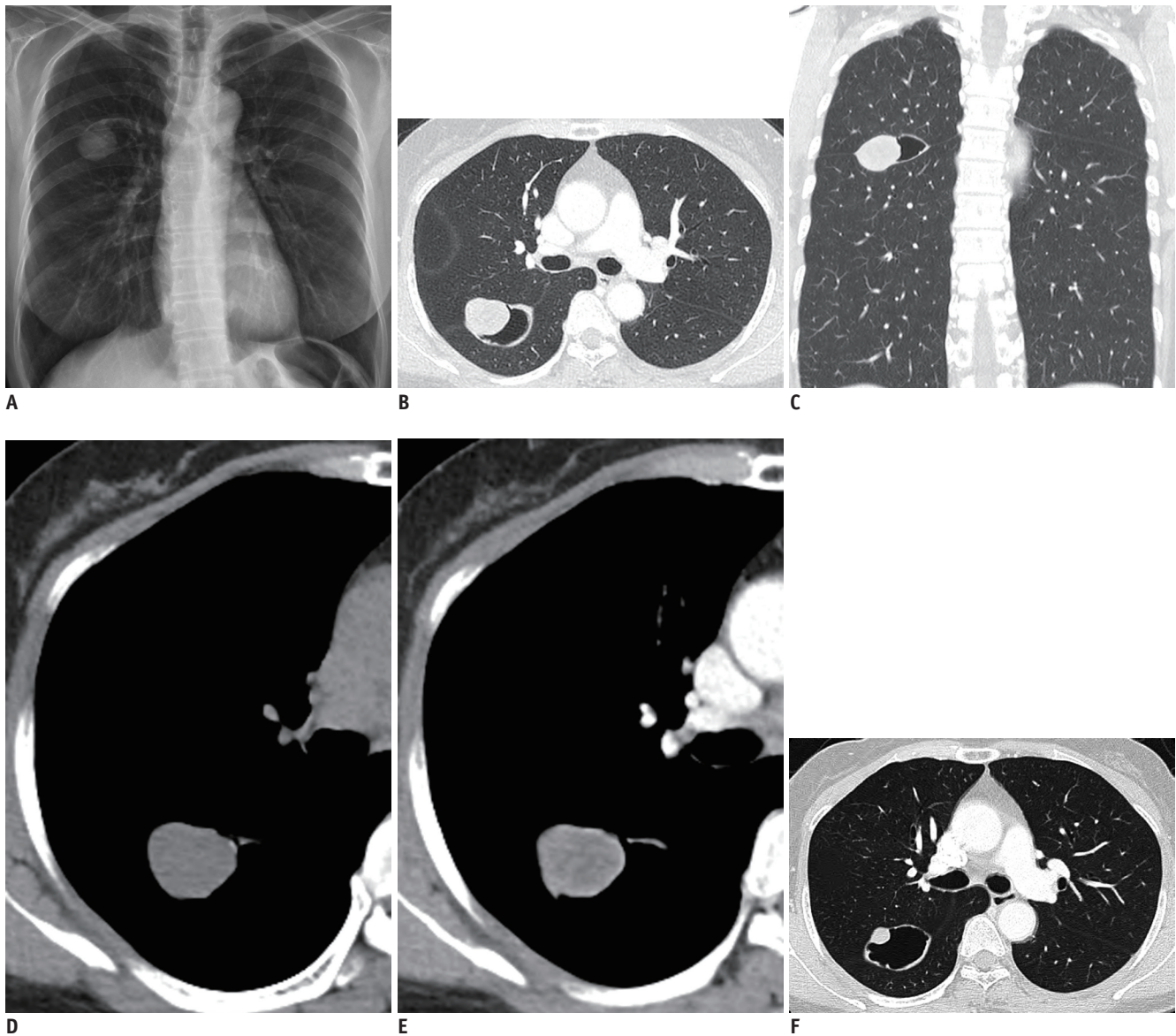


Fig. 1. Solitary fibrous tumor of pleura containing air in 58-year-old woman.

A. Chest radiograph on admission shows air-containing cystic mass in right upper lung field. **B-E.** Chest CT images demonstrate 4.5 cm ovoid or spindle-shaped cystic mass along upper portion of right major fissure, which contains eccentrically located soft tissue nodule surrounded by large amount of air (**B, C**). Mass has irregular thin wall. Internal soft tissue nodule enhanced relatively homogeneously after contrast injection (**D, E**). **F.** Chest CT image obtained two years prior to current presentation reveals same-sized cystic mass, with much smaller internal solid nodule compared to recent CT images (**B**), suggesting nodule growth.

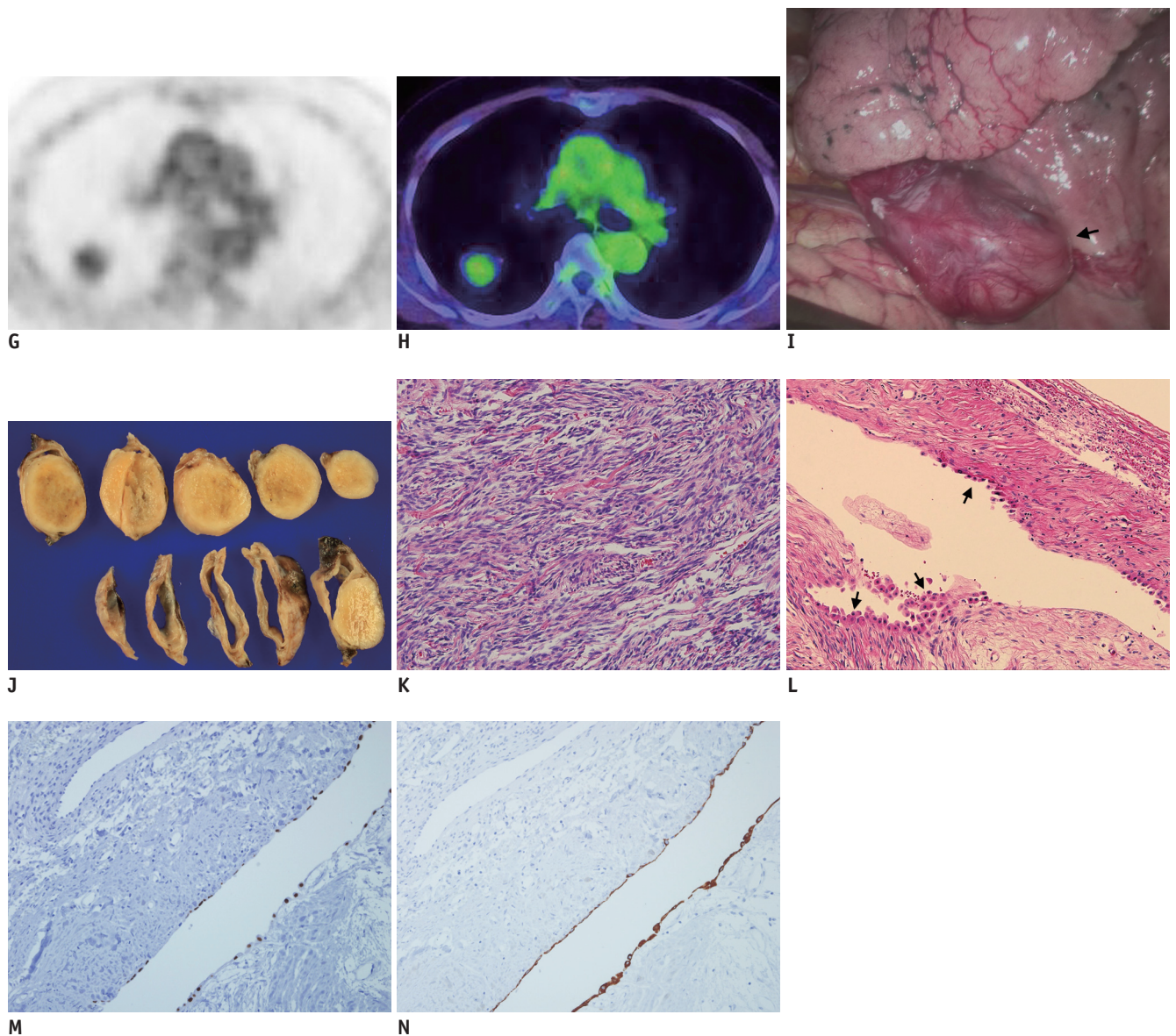


Fig. 1. Solitary fibrous tumor of pleura containing air in 58-year-old woman.

G, H. F18-fluorodeoxyglucose (FDG) PET/CT scans of soft tissue nodule show mild FDG uptake with SUVmax of 2.3. **I.** Intraoperative photograph shows well-encapsulated pedunculated pleural tumor (arrow) arising from visceral pleura of right major fissure covering right upper lobe. **J.** Photograph of cut sections of resected specimen reveals well-encapsulated air-containing cystic mass with eccentrically located yellow-gray-colored solid nodule. **K-N.** Photomicrographs of specimen demonstrate variable components of spindle-shaped tumor cells and collagen fibers arranged in haphazard pattern, consistent with SFT in both internal nodular (**K**) and cystic (**L**) portions of tumor (hematoxylin-eosin stain, x 200). Cystic spaces are lined by cuboidal cells (arrows, **L**). Stains for TTF1 (**M**) and CK7 (**N**) show immunoreactivity in cystic lining epithelium, suggesting that they were pneumocytes.

positively for TTF1 and CK7, suggesting that they were pneumocytes originating from alveoli or terminal respiratory epithelium (Fig. 1M, N). Antibodies for actin, desmin, S100, and CD117 (c-kit) were nonreactive in the tumor cells.

DISCUSSION

Solitary fibrous tumor is a rare mesenchymal neoplasm that can be either benign or malignant. The tumors typically

affect the pleura but can develop in a number of other locations including the mediastinum, lung, tracheobronchial tree, abdomen, head, neck, and central nervous system (1, 4). The tumors most commonly present during the fifth and sixth decades of life and occur equally in men and women (5). Although SFTs of the pleura are asymptomatic in up to 50% of patients, they can be associated with chest pain, dyspnea, cough, hypoglycemia, and clubbed finger (6). SFT of the pleura is suspected to be derived from the

submesothelial connective tissues that originate from the differentiation of a primitive multipotent mesenchymal cell. Tumors are typically highly vascular with vessels of varying sizes, and approximately 50% of SFTs of the pleura have a pleura-based pedicle, approximately 1 cm in length, and contain hypertrophic arteries and veins (6).

Chest radiographs of a patient with SFT of the pleura characteristically demonstrate a peripheral, well-defined, solitary nodule or mass, occupying the lower hemithorax, which may mimic diaphragmatic elevation or eventration (1). The tumor may also be located within a fissure, simulating a parenchymal mass. CT images of small SFTs typically show homogeneous, well-defined, lobular, noninvasive, soft-tissue nodules or masses. SFTs usually enhance strongly after contrast injection, which can be heterogeneous, particularly for large tumors. Heterogeneous attenuation of the tumor after contrast enhancement is correlated with hemorrhage, necrosis, or cystic change, and tumoral calcification occurs in 7% to 25% of patients, typically in those with larger tumors, and is often related to necrosis (1-3, 6).

An SFT containing a large amount of air has not previously been reported. However, several cases of pulmonary sclerosing hemangioma surrounded by slit-like air space have been described (7, 8). In these reports, the investigators suggested several possible mechanisms of air-space development in the tumors. Proliferation and hyalinization of alveolar mesenchymal cells around the bronchus can lead to airway narrowing and distention of distal air space (7). Air meniscus may also be induced by disparate rates of contraction between the tumor and capsule, possibly after tumor hemorrhage (7) or by airway communication after peritumoral hemorrhage (8). In rare cases, benign metastasizing leiomyomas can also present as fluid- or air-filled cysts. Cleft formation within the tumor caused by entrapment of the surrounding alveolar lining epithelial cells may eventually progress to cysts or bullae (9, 10). In the present case, immunohistochemically stained pneumocytes were present in the cystic portion of the tumor, representing the entrapped lung tissue, which can serve as the best explanation of air collection within the tumor. Peripheral lung tissue entrapped by the tumor may have resulted in air trapping within the tumor through

the communicated bronchioles, and eventually, the air space may have enlarged through both the check-valve mechanism and tumor growth.

To the best of our knowledge, this is the first case of an SFT containing a large amount of air within the tumor, and we speculate that entrapment of peripheral lung tissue by the growing tumor with eventual air-trapping over time may have resulted in the large air space within the tumor. Although this finding is extremely uncommon, radiologists should not exclude the possibility of SFT in the diagnosis of a pleural or subpleural nodule containing intra-tumoral air.

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