

2015 대한흉부외과학회 전공의 연수교육

Surgery for Congenital Anomalies and Benign tumors of The Lung

김현구

고려대학교 구로병원

Congenital Abnormalities of the Lung

CASE 1

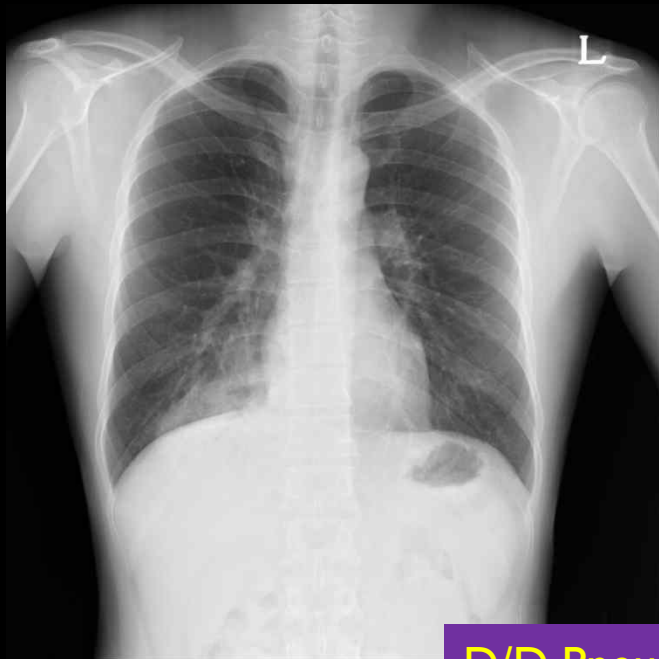
✓ M/33

✓ Symptoms

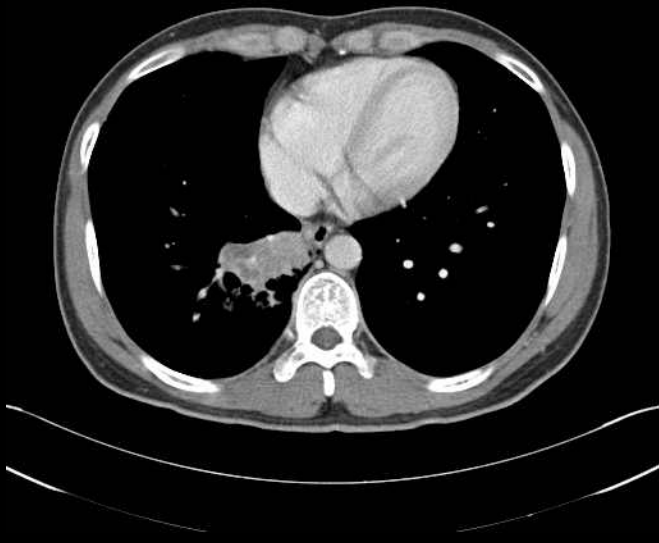
: Cough, sputum (onset: 2~3 weeks ago)

✓ Lab.

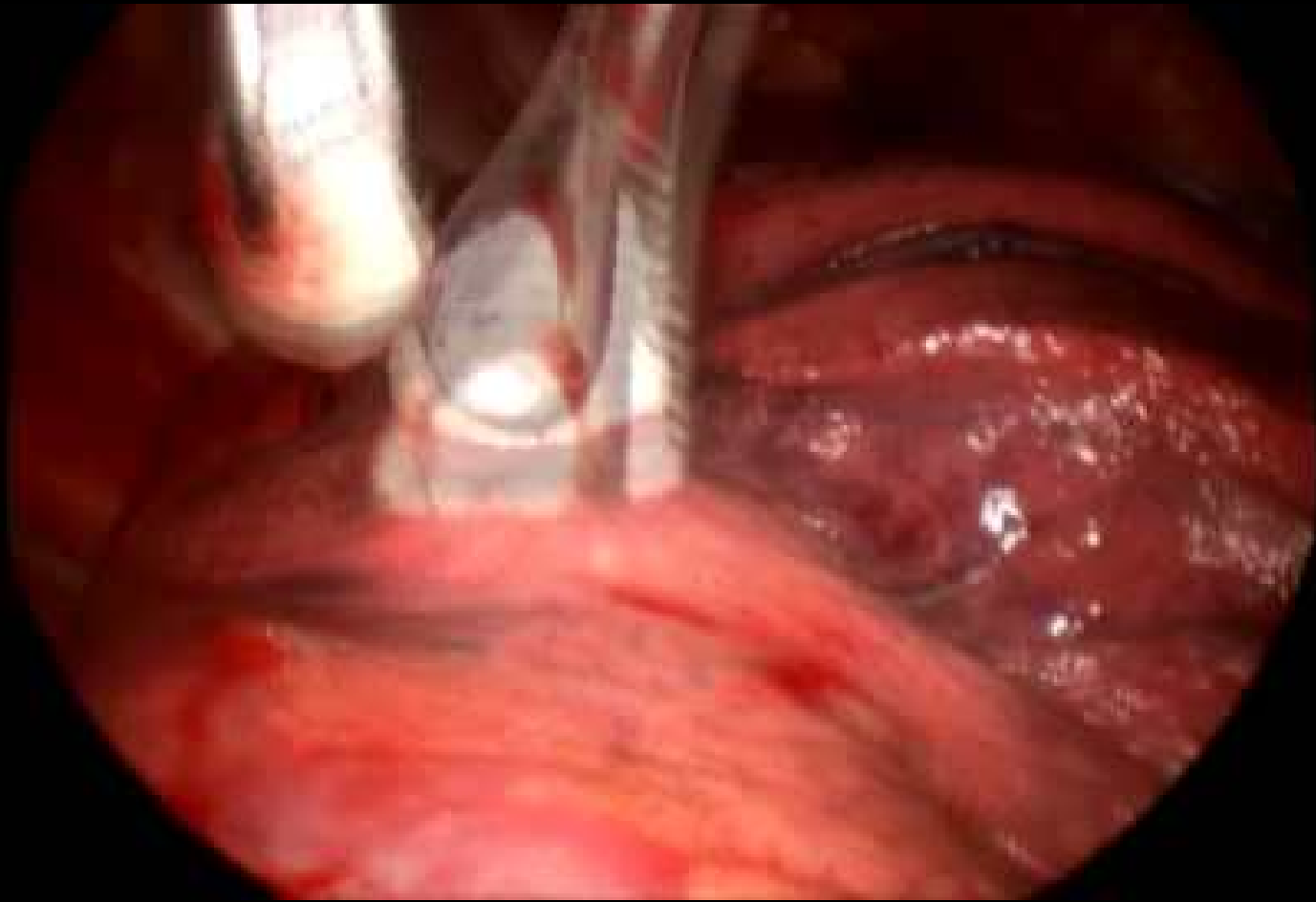
: WBC: 7100, CRP: 9.99, Sputum culture: (-)



D/D Pneumonia with abscess formation in right basal lung, combined congenital anomaly

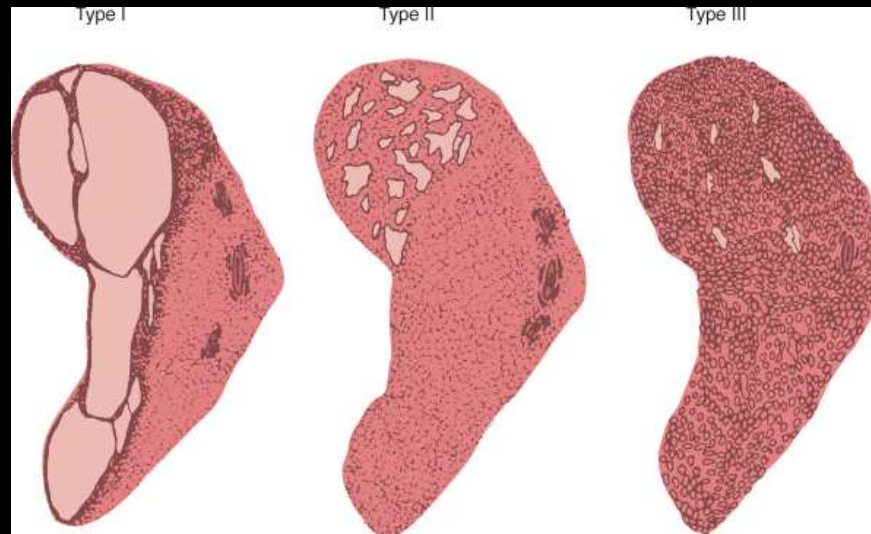


Operation



Congenital Cystic Adenomatoid Malformation (CCAM)

- Segmental bronchial atresia during fetal lung bud development



Stocker's classification of congenital cystic adenomatoid malformation. **Type I** typically has one or more large cysts. (55% of cases). **Type II** has numerous small cysts (<20mm). **Type III** is essentially a solid mass of tissue.

- Equally affected at both lungs, commonly in lower lobes
- Size of lesion: primary determinant of prognosis

➤ Symptoms

1. Cough and recurrent pulmonary infection in older children
2. Asymptomatic: incidentally noted in chest x-ray

➤ Regression: first few months after birth

→ For asymptomatic patients, observation for 1 year is appropriate

➤ Surgical resection

1. At the onset of respiratory symptoms
2. No evidence of regression

➤ Reasons for early resection

1. Malignant potential: pleuropulmonary blastoma, rhabdomyosarcoma, BAC (M/C in type I)
2. Expecting compensatory lung growth after resection

CASE 2

✓ M/25

✓ Symptoms

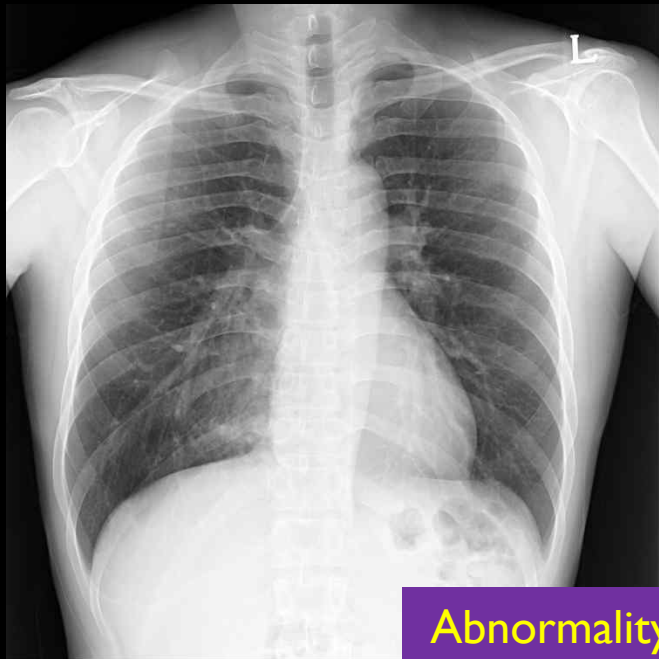
: Dyspnea (onset: 3~4 weeks ago)

✓ Past Hx.

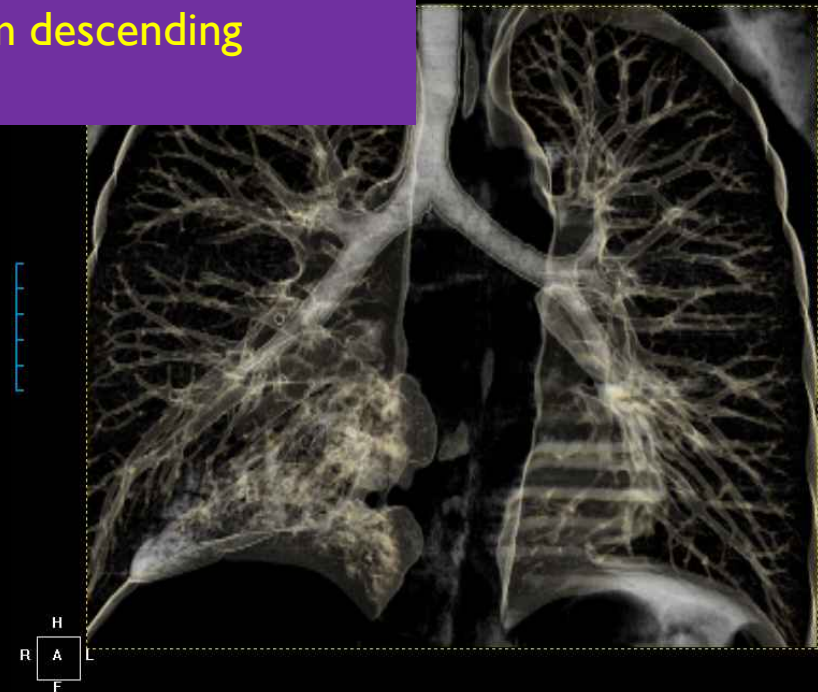
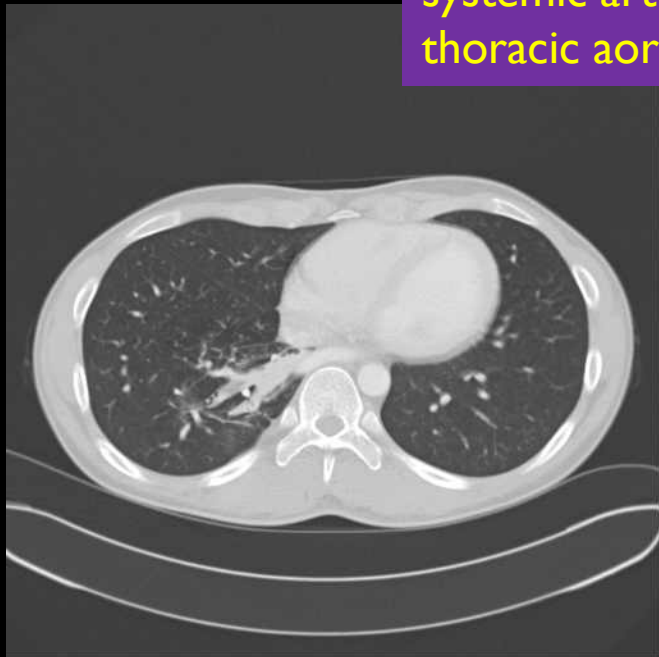
: 2년 전 건강검진에서 흉부 X-ray상 이상 소견 발견

✓ Lab.

: WBC, ABGA: within normal limit



Abnormality in RLL with a large abnormal systemic artery supply from descending thoracic aorta.



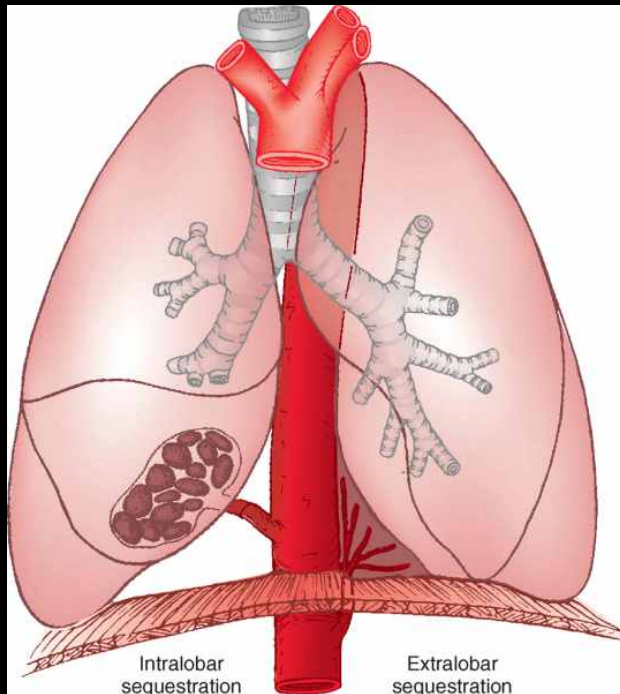
Operation



Pulmonary Sequestration

- Absence of communication with the tracheobronchial tree and pulmonary arteries
- **Blood** supply: systemic artery
drainage: pulmonary or systemic veins
- **Symptoms**
 1. Hemodynamic changes, audible continuous murmur
 2. Respiratory difficulties in an infant
 3. Chronic pulmonary infection in older ages and teenagers
 4. Asymptomatic mass in an adult

Extra / Intralobar type



Extralobar

➤ Characteristics

1. Completely separate from the remaining lung
2. Has its own pleural investment
3. Usually located in the inferior portion of the thoracic cavity but, anywhere from the neck to just below the diaphragm

➤ Blood supply

- Artery: descending aorta, abdominal aorta, subclavian, intercostal
- Vein: azygos, hemiazygos, subclavian, portal vein

➤ May be connected with esophagus

➤ 3-D CT for finding systemic artery

➤ **Associated congenital anomaly:** Diaphragmatic hernia, pericardial defect, anomalous pulmonary venous drainage

➤ Surgery

- Resection soon after making diagnosis
- Resection lesion itself without any accompanying lung tissue

Intralobar

➤ Characteristics

1. Congenital, possibly acquired resulting from chronic infection
2. Surrounded normal lung tissue without separate pleural investment
3. Located in lower lobe in all cases

➤ Blood supply

- Artery: descending thoracic aorta, variable branches of aorta
- Vein: inferior pulmonary vein, systemic vein

➤ Recurrent infection

➤ Surgery

- Resection soon after diagnosis
- Removal of the entire lobe including normal lung tissue

CASE 3

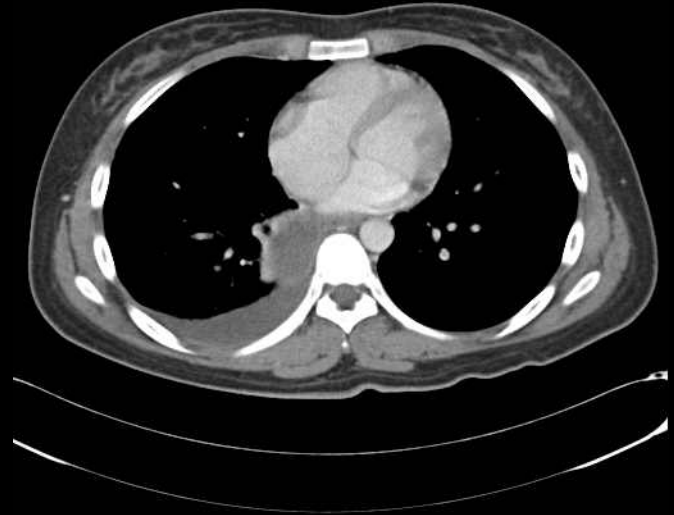
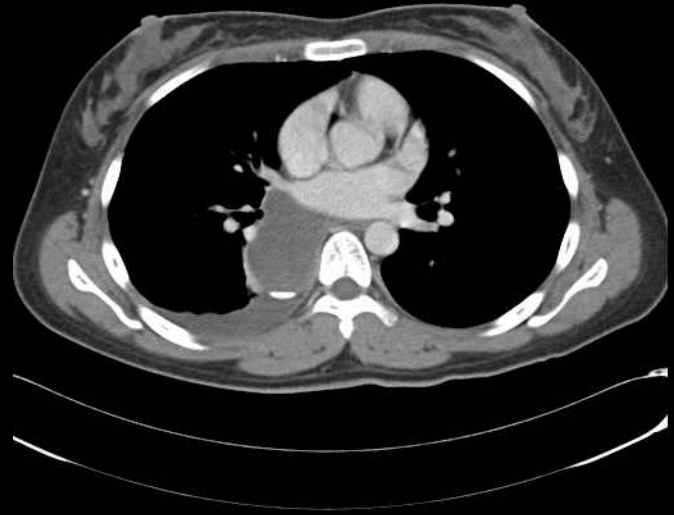
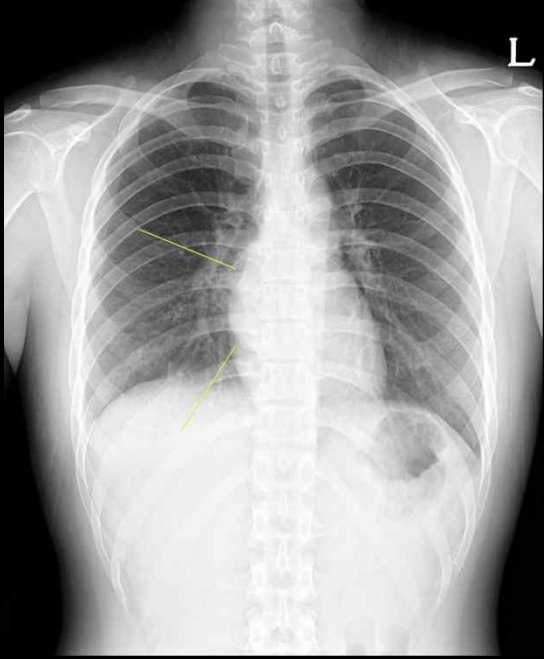
✓ F/27

✓ Symptoms

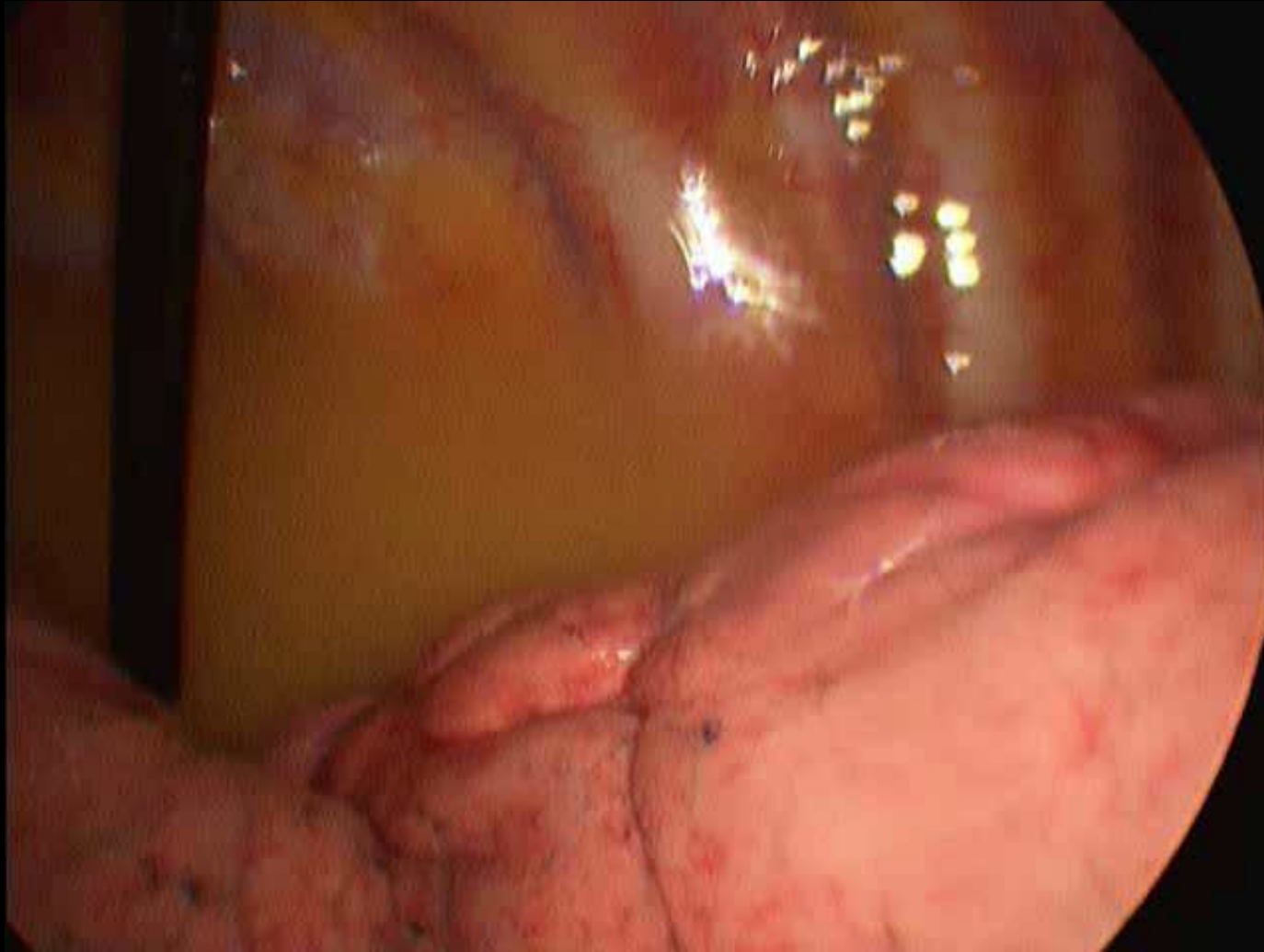
: right chest pain (onset: 5 days ago)

✓ Lab.

: WBC, ABGA, Culture: within normal limit



Operation



Bronchogenic Cyst

➤ Type according to the location

1. Mediastinal

- 2/3 of patients
- Paratracheal, carinal, hilar

2. Parenchymal

- may communicate with bronchus

3. Others: Inferior pulmonary ligament, retroperitoneal, neck

➤ Solitary cyst filled with fluid or mucus

➤ Symptoms

- Compression of air way
 - : wheezing, stridor, shortness of breath, pneumonia, emphysema
- 1/3 of patients
 - : incidental on imaging in asymptomatic child or adult

➤ Surgery

- Soon after diagnosis because of high likelihood of symptoms developing
- Simple resection for mediastinal location
- Extensive resection for parenchymal location

Congenital Abnormalities of Lung

Lesion	Location	Radiologic Findings	M:F	Symptoms	Vasculature
Lobar emphysema	LUL 40%	Hyperlucent lobe	2.5 : 1	Tachypnea, respiratory distress	Normal
	RML 35%	Mediastinal shift			
	RUL 20%				
CCAM	Equal all lobes	Type I—single cyst	1 : 1	Respiratory distress	Normal
		Type II—many small cysts			
		Type III—solid mass			
Sequestration					
Extralobar	Basilar, left 80%	Wedge-shaped mass	2 : 1	Usually none	Arterial—descending aorta
					Venous—hemiazygos
Intralobar	Posterior basal, left 60%	Cyst or solid	1 : 1	None or infection	Arterial—descending aorta
					Venous—Pulmonary vein
Bronchogenic cyst					
Mediastinal	Pericarinal	Round mass	1 : 1	None or wheezing	Normal
Parenchymal	Lower lobes	May contain air	1 : 1	Infection, wheezing	Normal

Benign Tumors of the Lung

CASE 4

✓ M/53

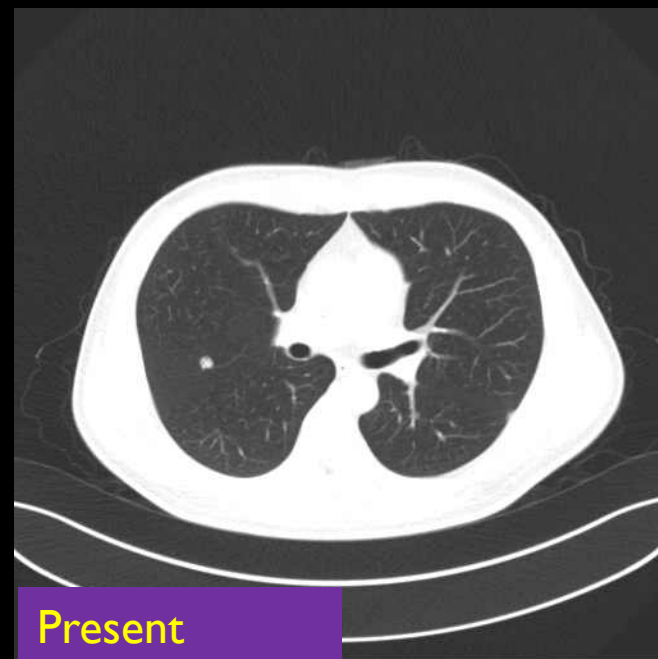
✓ Symptoms

- 1년 전 건강검진상 SPN발견

SPN in RML

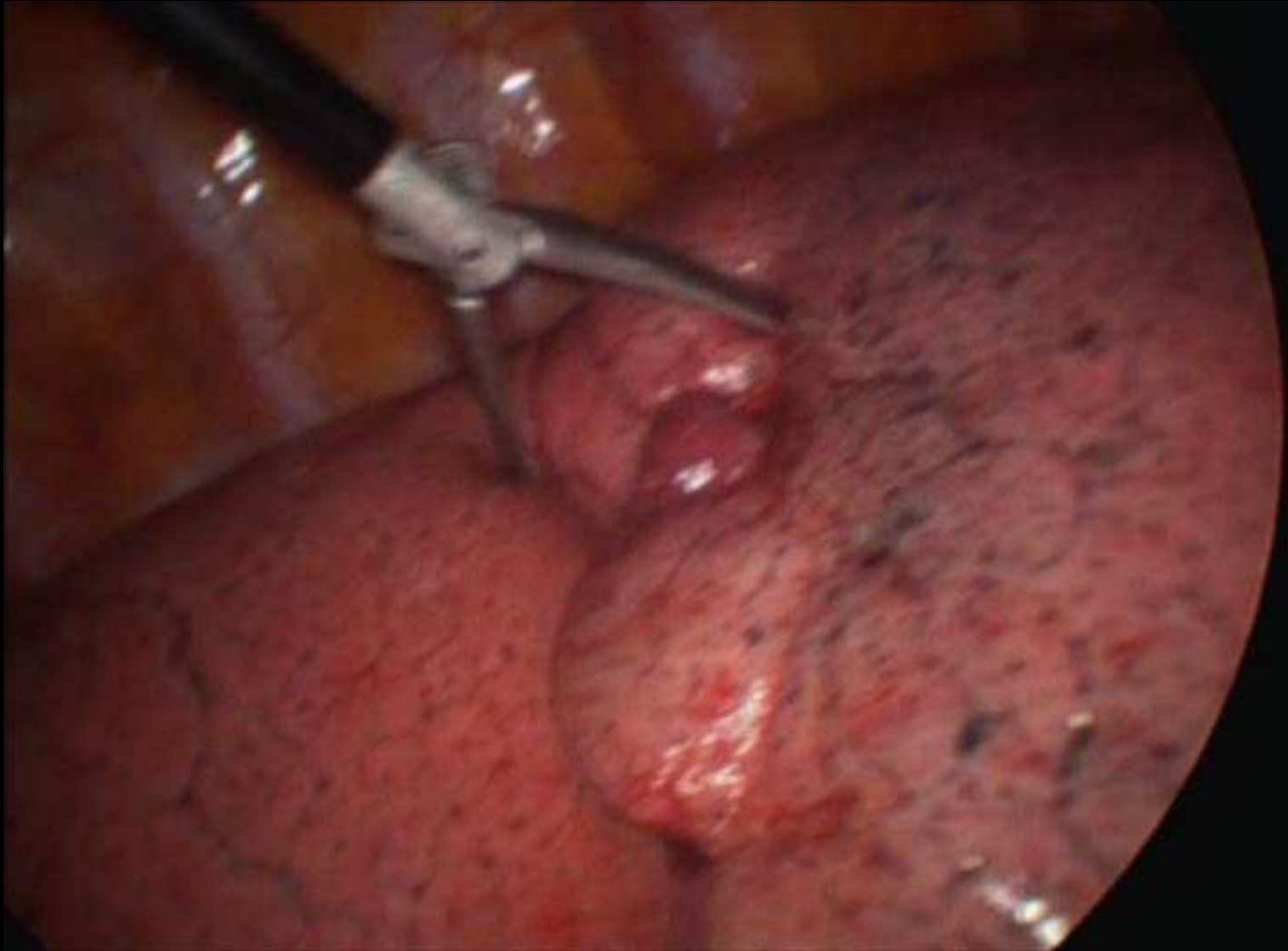


1 year ago



Present

Operation



Harmatoma

➤ Prevalence

- M/C benign tumor: 4~8% of pulmonary neoplasm
- Half: chromosomal abnormality (12q14-15)
- Male dominant (2:1 or 3:1)
- Age: 30~60 years

➤ Radiologic findings

- Composed of mainly of cartilage and gland-like formation with significant amount of fat, connective tissue, smooth muscle
- M/C in peripheral area
- Endobronchial lesion in 3~20%
- Well-circumscribed lesion
- Mean size: 1.8cm in diameter (0.2~5cm)
- Popcorn calcification: presence of fat density (up to 50%)

➤ Symptoms

- Asymptomatic
- Must be differentiated from carcinoma on chest X-ray
- Needle aspiration biopsy: diagnostic yield in 85%

➤ Fate

- Slow growth
- Tumor growth was observed in 48% for 4.1 years (3.2±2.6mm/year)
- Reasonable to watch these lesions
- Malignancy is rare.

➤ Treatment

- If biopsy has not been confirmed, Surgery is an acceptable approach
- If the mass is $>2.5\text{cm}$, Remove it.
- Preoperative marking under CT guidance is recommended.
- Enucleation or wedge resection, segmentectomy, or lobectomy

➤ Malignant associated with Hamartoma

- Adenocarcinoma, or sarcoma might have been developed from hamartoma
- Bronchial carcinoma was identified 6.3~6.6 times more often with a hamatoma.

CASE 5

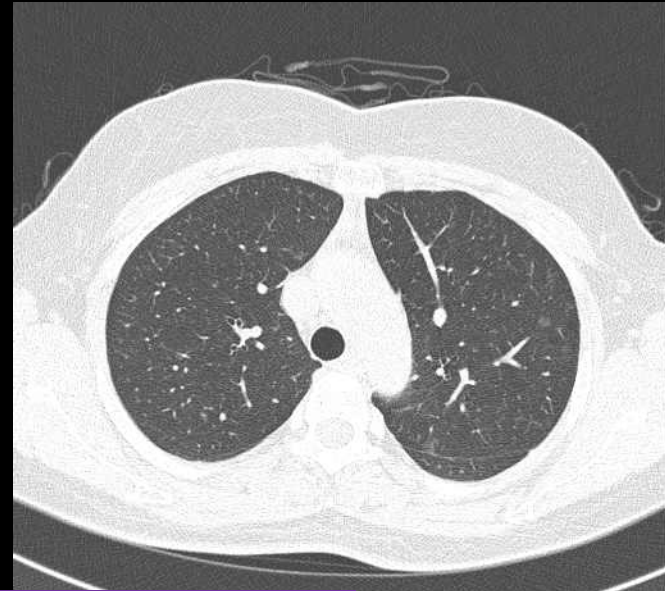
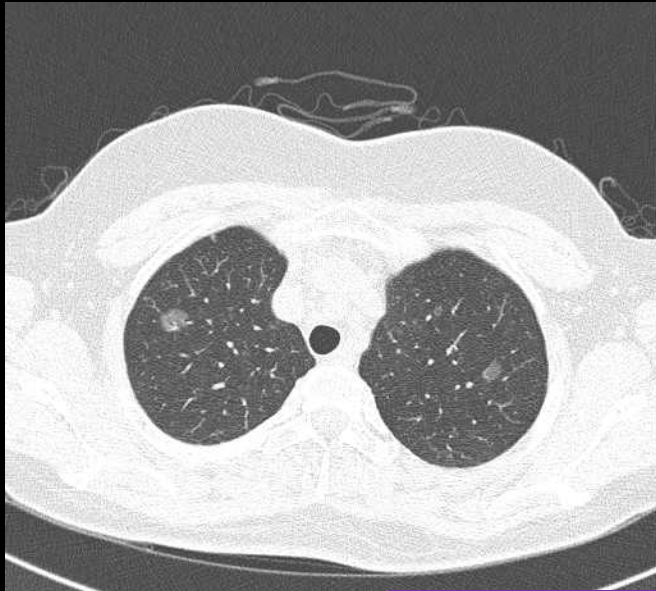
✓ F/54

✓ PHx.

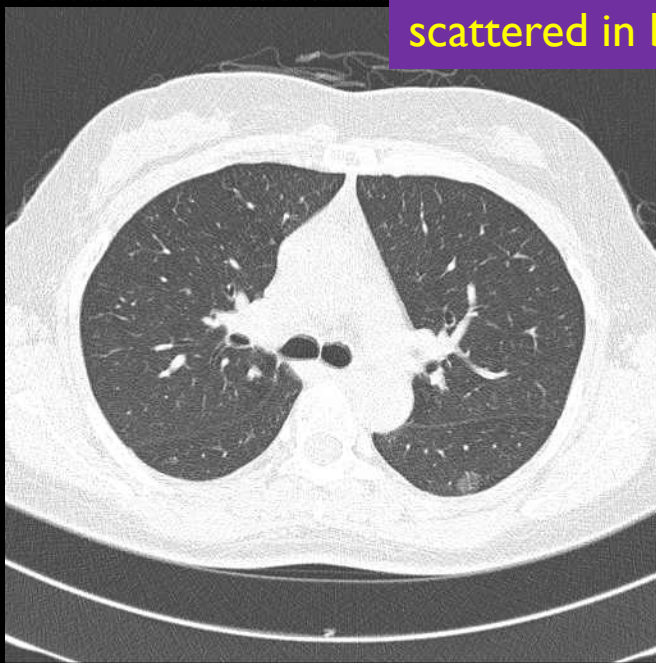
: 3년 전 rectal cancer로 수술 받음

✓ C.C.

: CT F-up중 multiple GGO lesion 발견



Multiple, variable-sized, ground-glass nodules are scattered in both lungs



Operation



AAH

➤ Nomenclature

- *Atypical adenomatous hyperplasia*
- *Aveolar cell hyperplasia*
- *Atypical alveolar hyperplasia, bronchioloalveolar cell adenoma*

➤ Pathology

- One or more small, asymptomatic ground-glass opacity in the peripheral lung
- Proliferation of minimally atypical cuboidal type II pneumocytes
- 1~10mm in size (majority <5mm)
- Any lesion >5mm: probable carcinomatous tumor
- Premalignant lesion to adenocarcinoma

➤ Fate

- Size of tumors grew to 0.2 cm or more, AAH to BAC
 - 0.3 cm or more to minimally invasive adenocarcinoma
 - 1.0 cm or more to overtly invasive adenocarcinoma
 - 2.0 cm or more to overtly invasive adenocarcinoma with LN metastasis

➤ Incidence

- 12 to 23% in resected materials and 3% in autopsy materials.
- Usually found in the upper lobe of the lung and occurs most frequently in patients with adenocarcinoma
- Multiple AAH lesions reportedly show a significantly higher frequency of preceding malignancies

➤ Management

- Clinical differentiation between BAC and AAH by needle aspiration is essentially impossible
- If GGO is resected and found to represent
AAH → nothing more than wedge resection
BAC → wedge resection, segmentectomy, or lobectomy

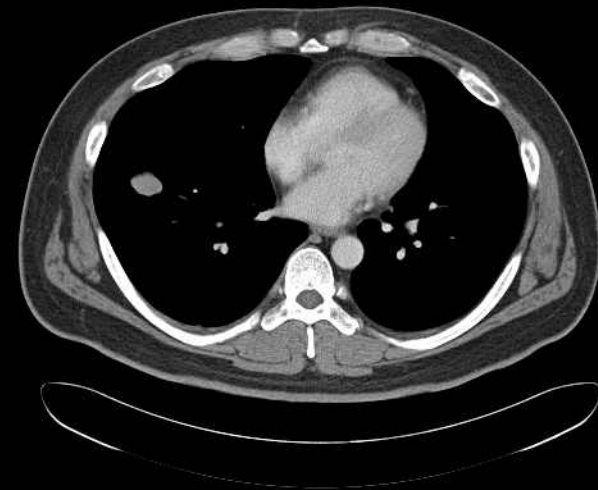
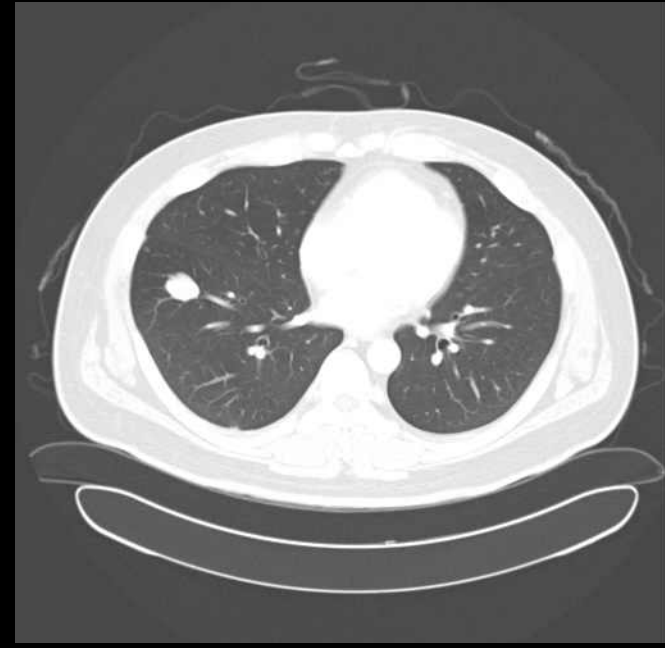
CASE 6

✓ M/41

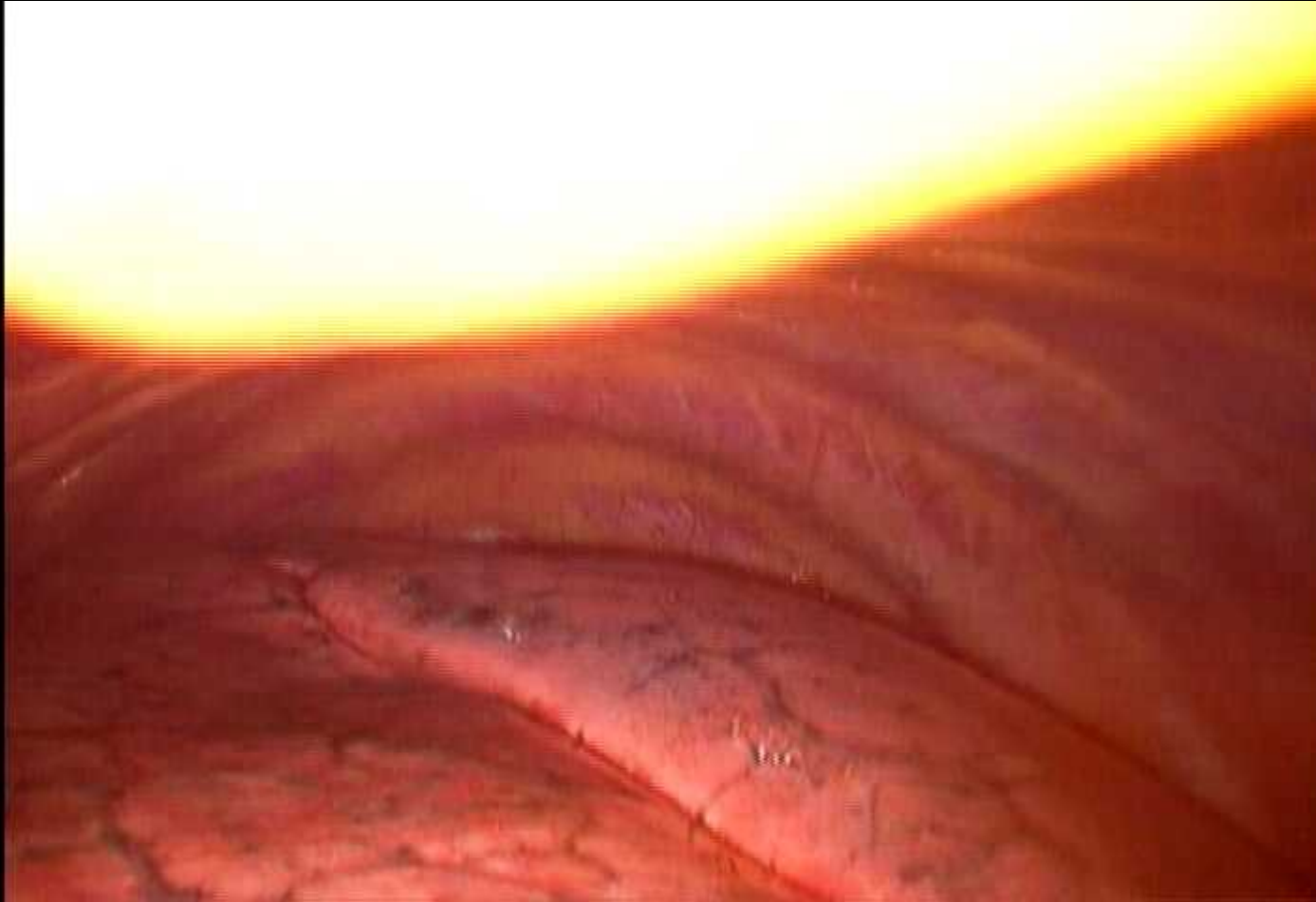
✓ C.C.

: 건강검진상 SPN발견

A 2.5cm sized oval shaped lesion in RLL anterior basal segment, with mild contrast enhancement.



Operation



Leiomyoma

➤ Prevalence

- Rare (2%), but M/C soft tissue tumor of lung
- 2/3 in women
- Mean age: 35 years old

➤ Symptom and Sign

- Asymptomatic
- Found incidentally on chest X-ray or CT
- Endobronchial symptoms in few

➤ Pathology

- Composed of smooth muscle fiber
- Origin
 - Bronchial smooth muscle or wall of bronchial artery
 - Metastatic myomas of uterine origin

➤ Treatment

- Surgical resection is treatment of choice

Benign Tumors of the Lung

Origin Unknown

- Hamartoma
- Clear cell (sugar) tumor
- Teratoma

Epithelial Tumors

- Papilloma
- Polyps
- Atypical adenomatous hyperplasia

Mesodermal Tumors

- Fibroma
- Lipoma
- Leiomyoma
- Chondroma
- Granular cell tumor
- Sclerosing hemangioma (alveolar pneumocytoma)

Other

- Inflammatory myofibroblastic tumor
- Xanthoma
- Amyloid
- Mucosa-associated lymphoid tumor

Occurrence (%)

- Hamartoma (76.9)
- Benign mesothelioma (12.3)
- Xanthomatous and inflammatory pseudotumors (5.4)
- Lipoma (1.5)
- Leiomyoma (1.5)
- Hemangioma (0.8)
- Adenoma of mucous glands (0.8)
- Mixed tumor (0.8)

Thank you !