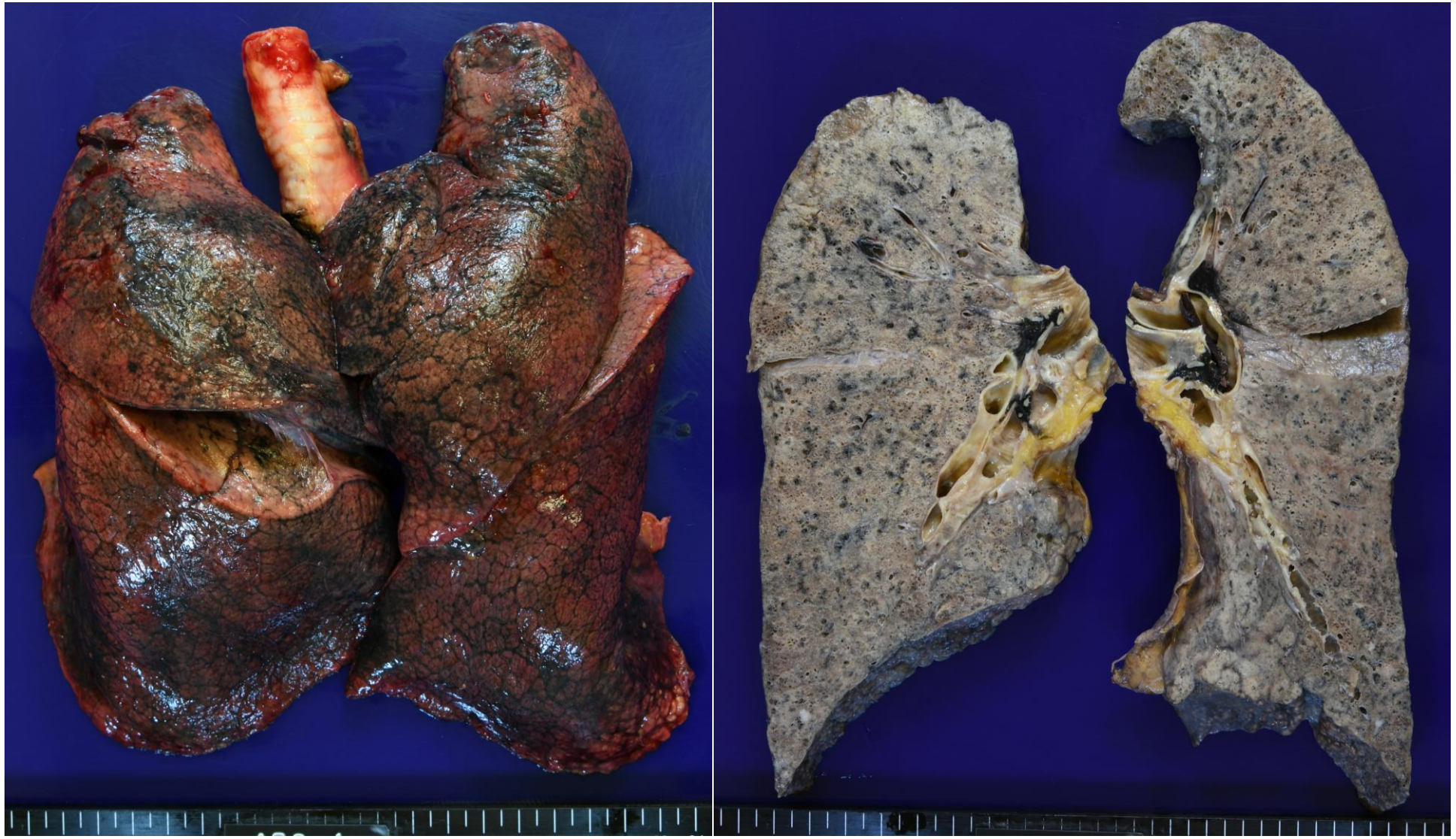
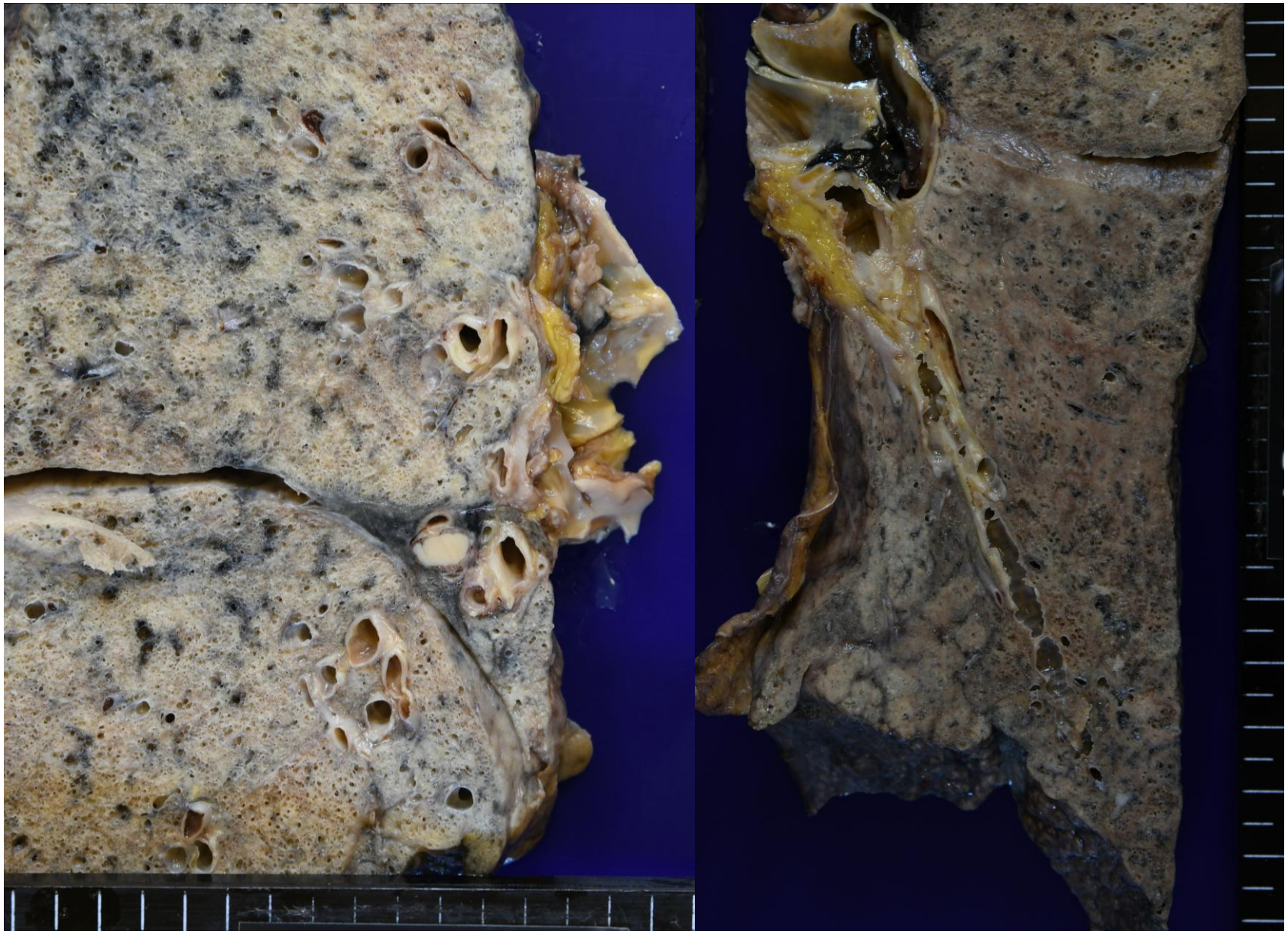
Idiopathic pulmonary fibrosis: an autopsy case

Idiopathic pulmonary fibrosis (IPF) is a chronic, progressive and fibrosing lung disease. IPF gets worse over time. The lung parenchyma is replaced by altered extracellular matrix and alveolar architecture is destroyed. The process leads to decreased lung compliance, disrupted gas exchange and ultimately respiratory failure. The disease usually affects people aged around 70 to 75 years old, and is rare in people under 50. The cause is unknown, hence the term “idiopathic” is used. The exposure to certain types of dust, viral infections, a family history of IPF, autoimmune disorders, acid reflux and smoking may be linked to the etiology.

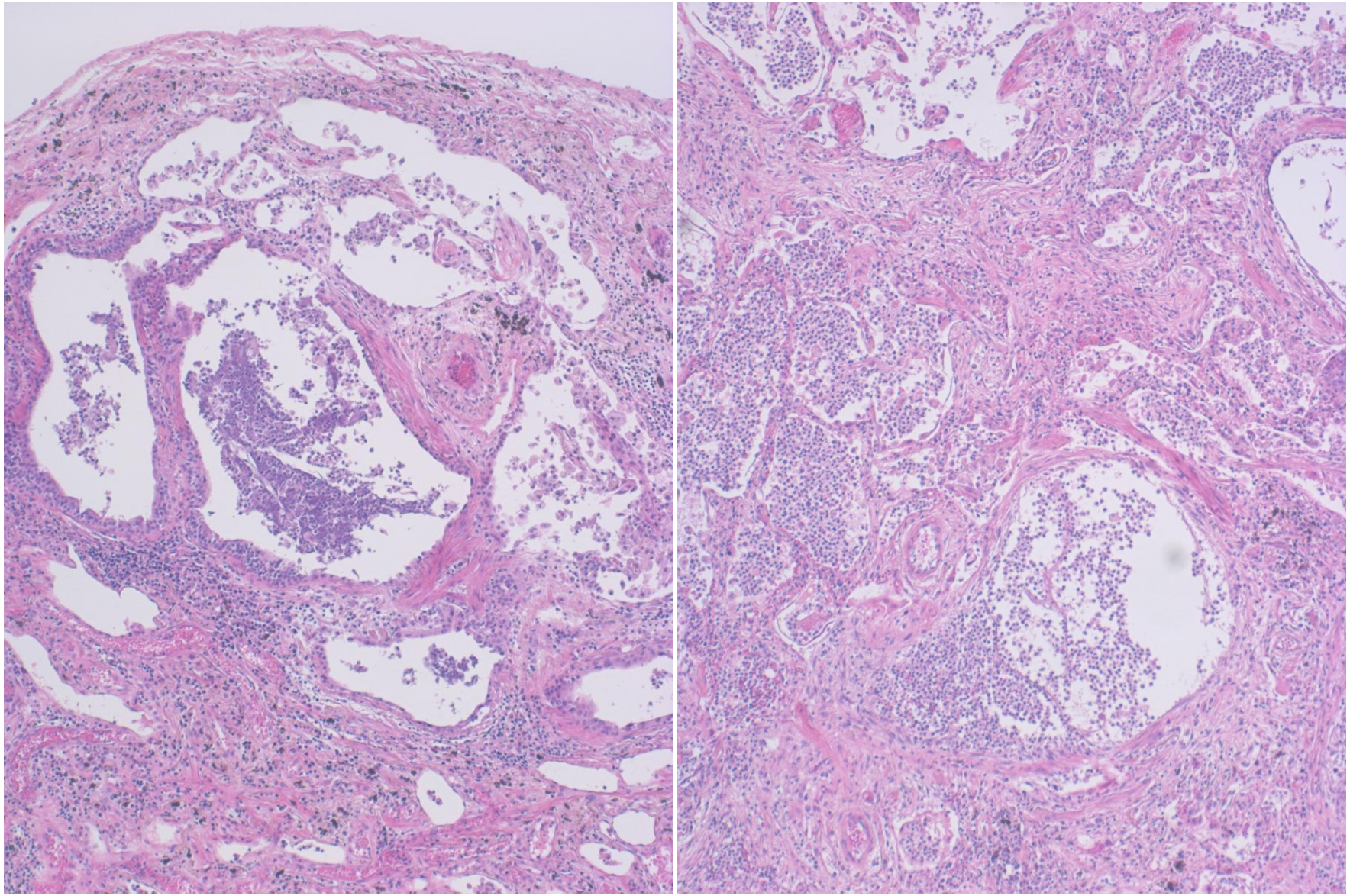
Ref.: Wolters PJ, et al. Pathogenesis of idiopathic pulmonary fibrosis. Annu Rev Pathol 2014; 9: 157-79. doi: 10.1146/annurev-pathol-012513-104706



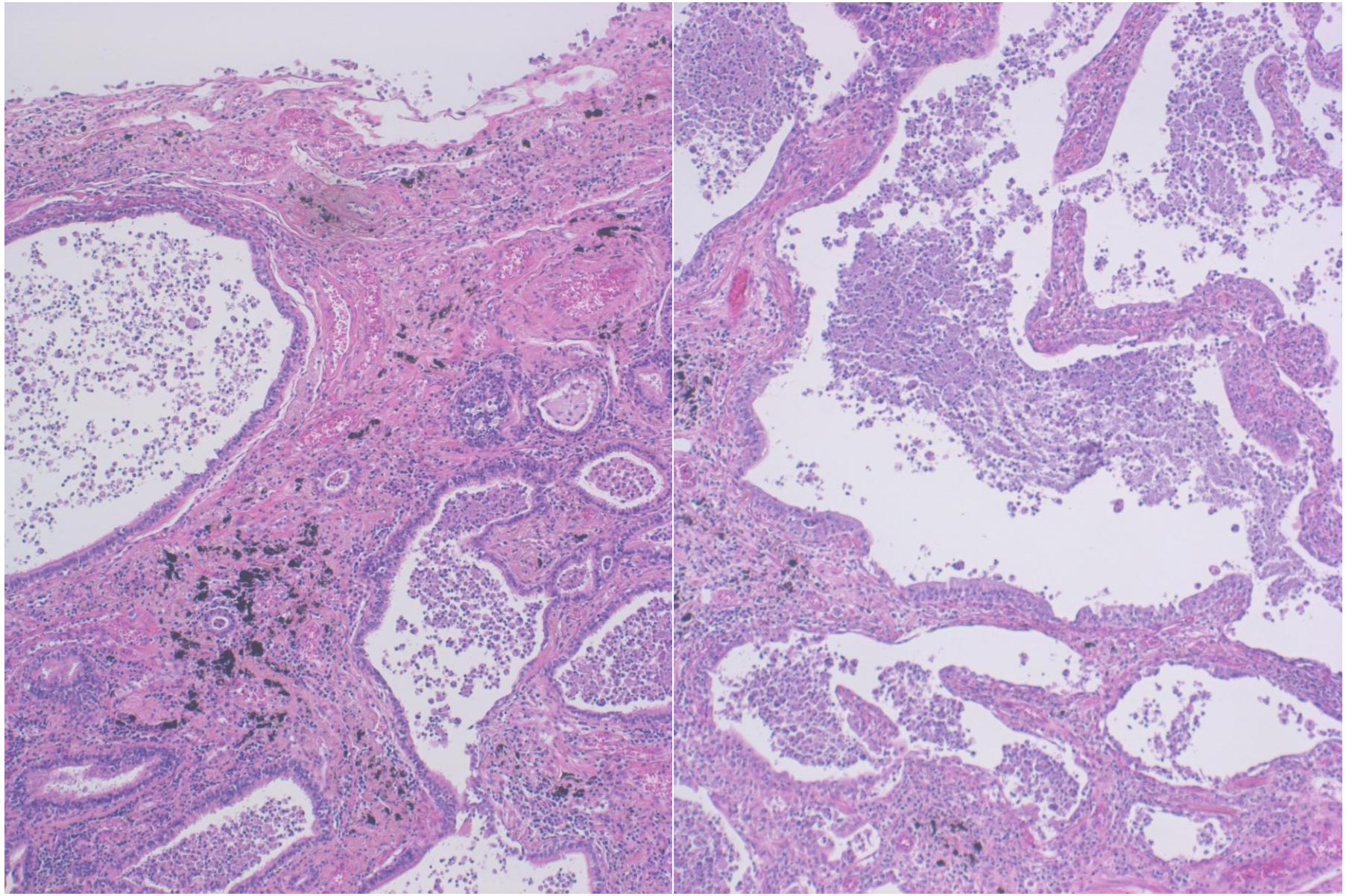
Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. Both lungs are hardened and fibrotic changes are diffusely observed (left: unfixed, right: cut surfaces after formalin fixation). The lung weight: left 350 g, right: 380 g.



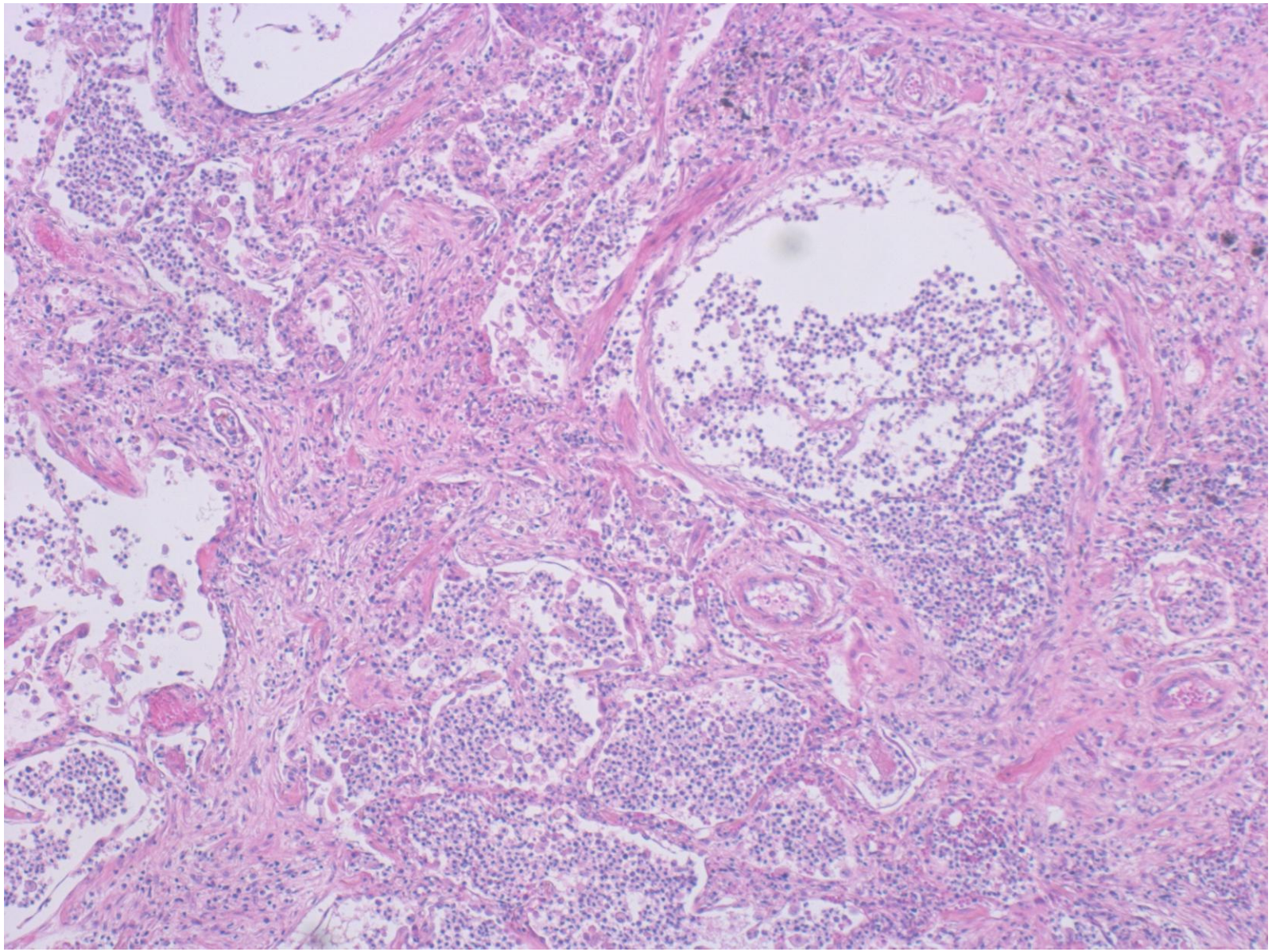
Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. The cut surfaces of the lung after formalin fixation show diffuse consolidation and fibrosis (features of UIP). The right middle lobe is shrunken to show middle lobe syndrome caused by old tuberculosis. Caseous necrosis is seen in the hilar lymph node.



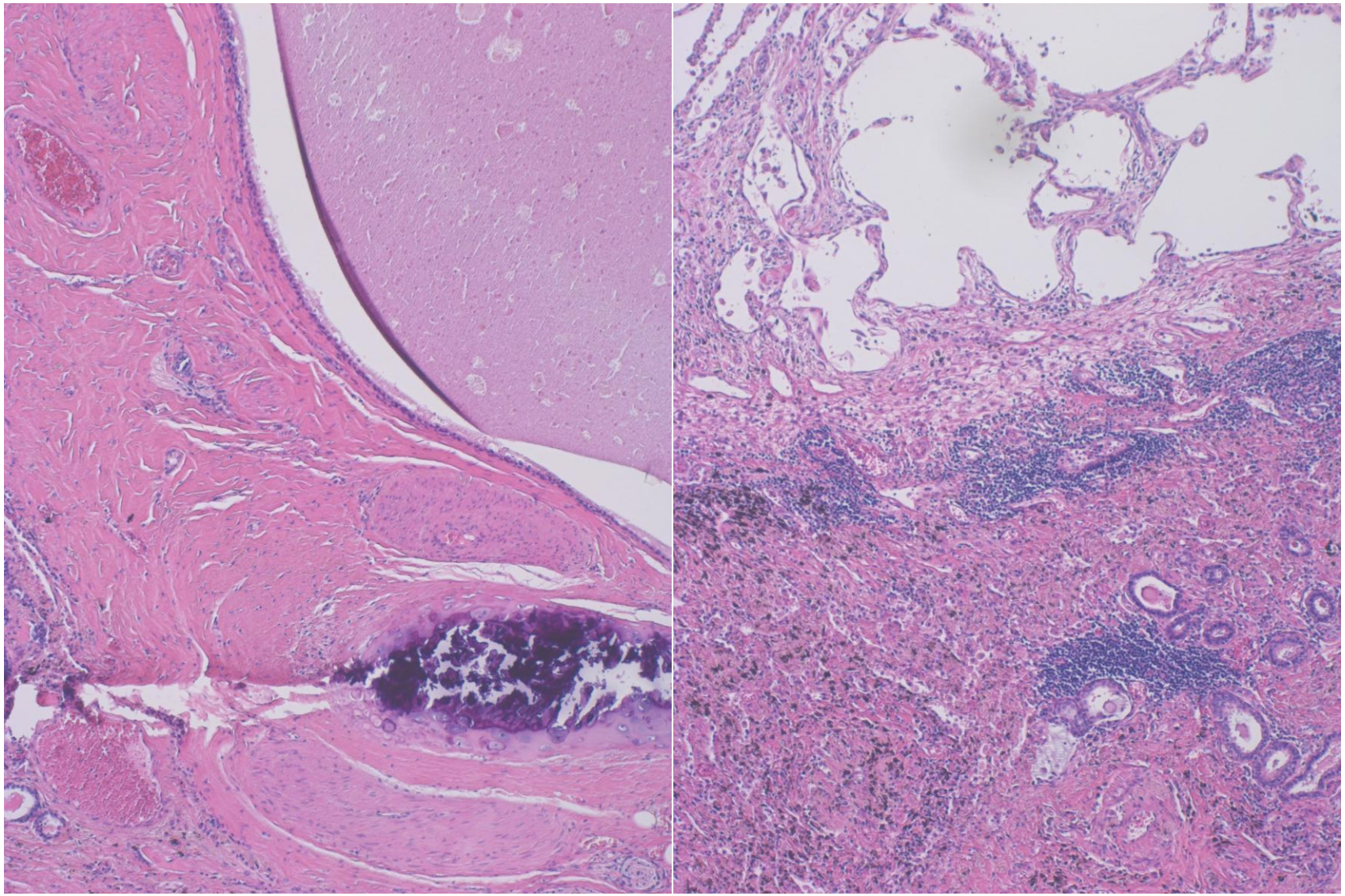
Idiopathic pulmonary fibrosis (UIP) seen in a 90 y-o female patient. Features of honeycomb lung with interstitial fibrosis are observed beneath the pleura (left lung). Secondary aspiration pneumonia with neutrophilic infiltration is associated (H&E-1).



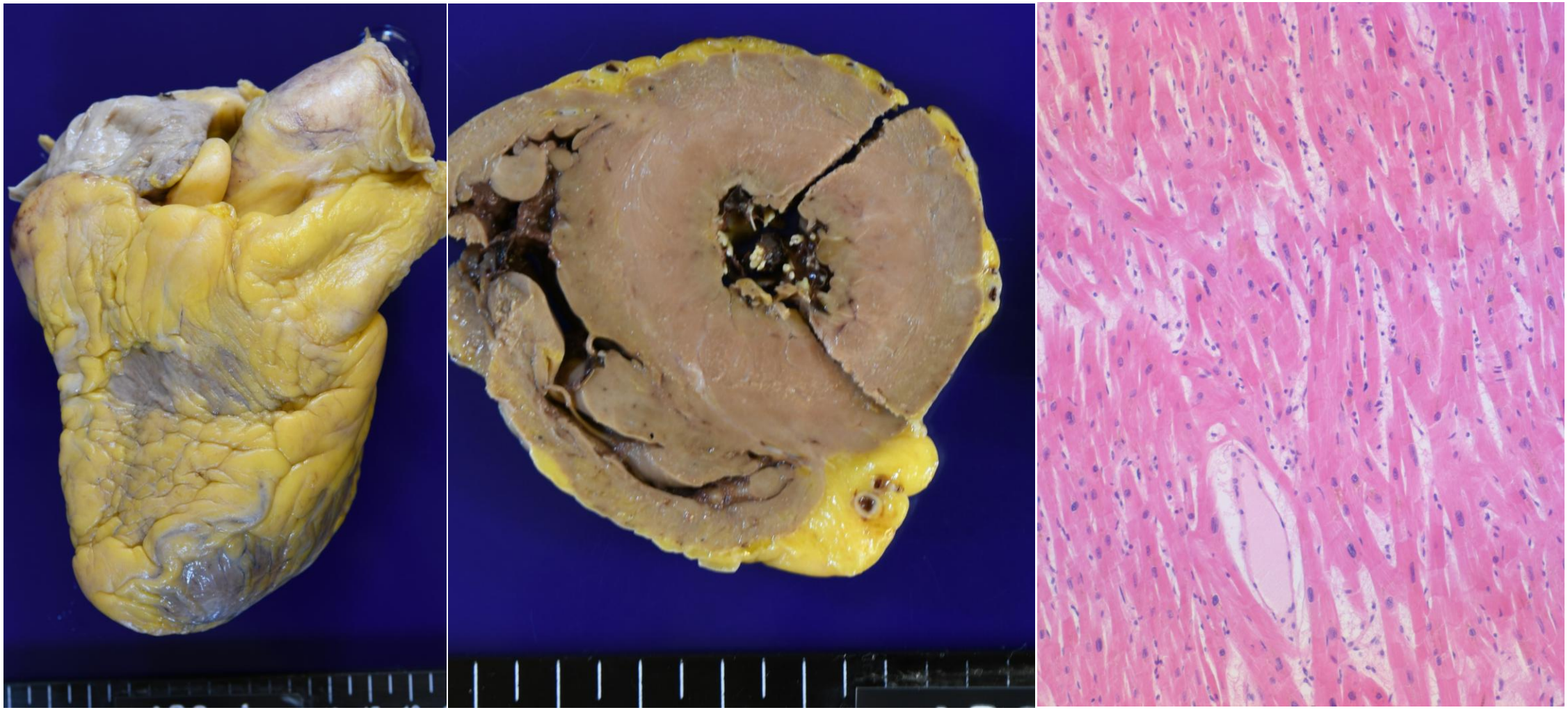
Idiopathic pulmonary fibrosis (UIP) seen in a 90 y-o female patient. Features of honeycomb lung with interstitial fibrosis are observed beneath the pleura (right lung). Secondary aspiration pneumonia with neutrophilic infiltration is associated (H&E-2).



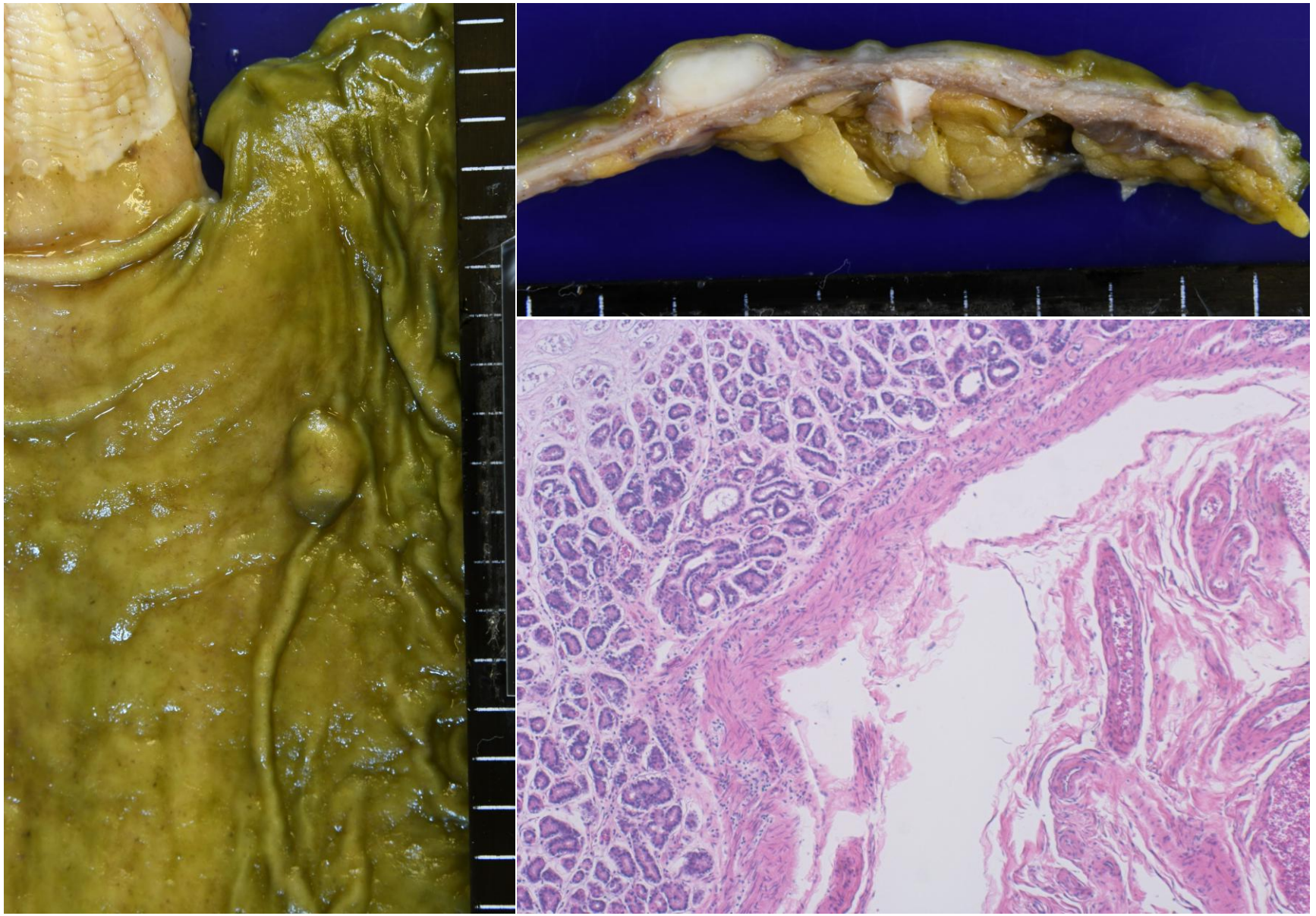
Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. The left lower lobe of the lung shows bronchopneumonia (aspiration pneumonia) with neutrophilic infiltration, causing acute exacerbation. Interstitial fibrosis is discerned (H&E-3).



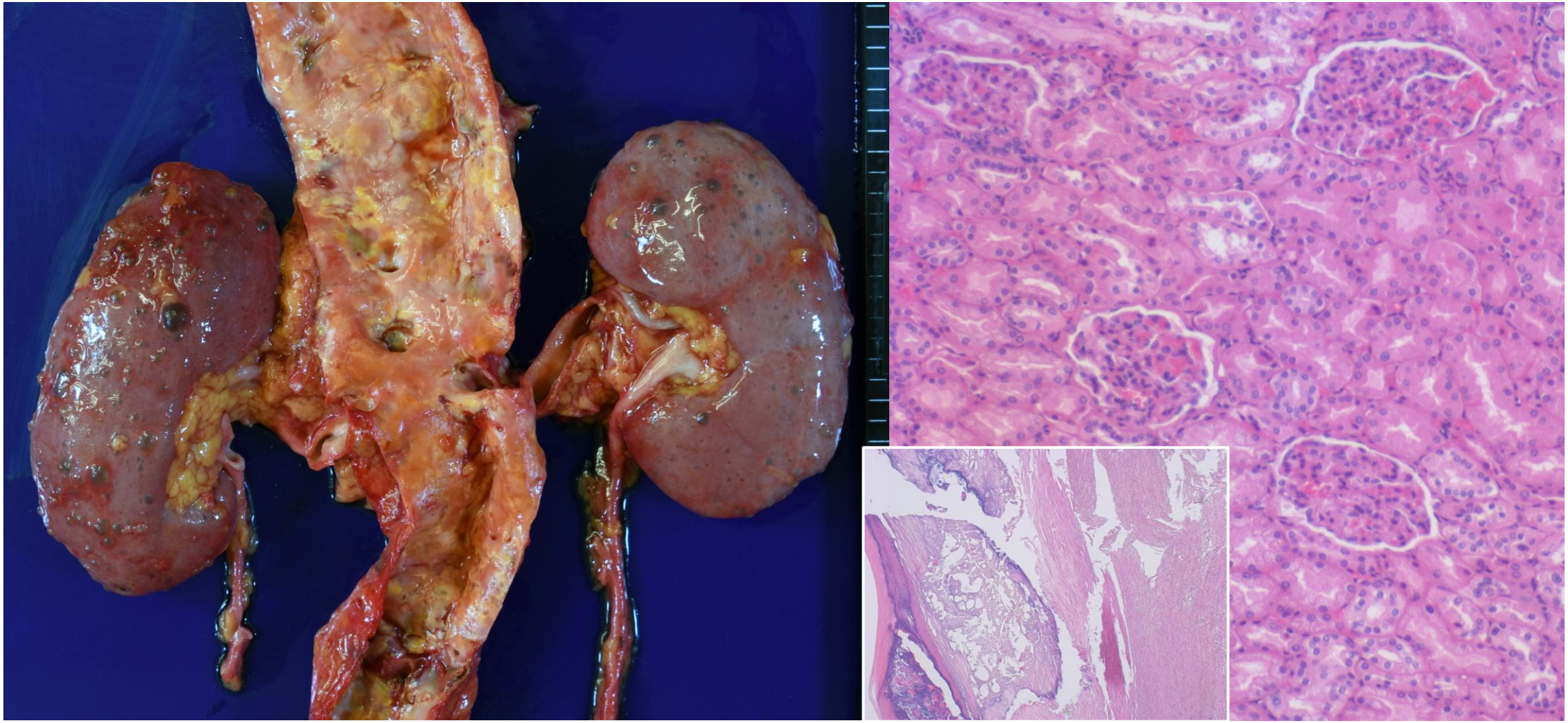
Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. The lesion of middle lobe syndrome shows collapse of the lung parenchyma, dilatation of the bronchus, interstitial calcification and lymphocytic infiltration (H&E-4).



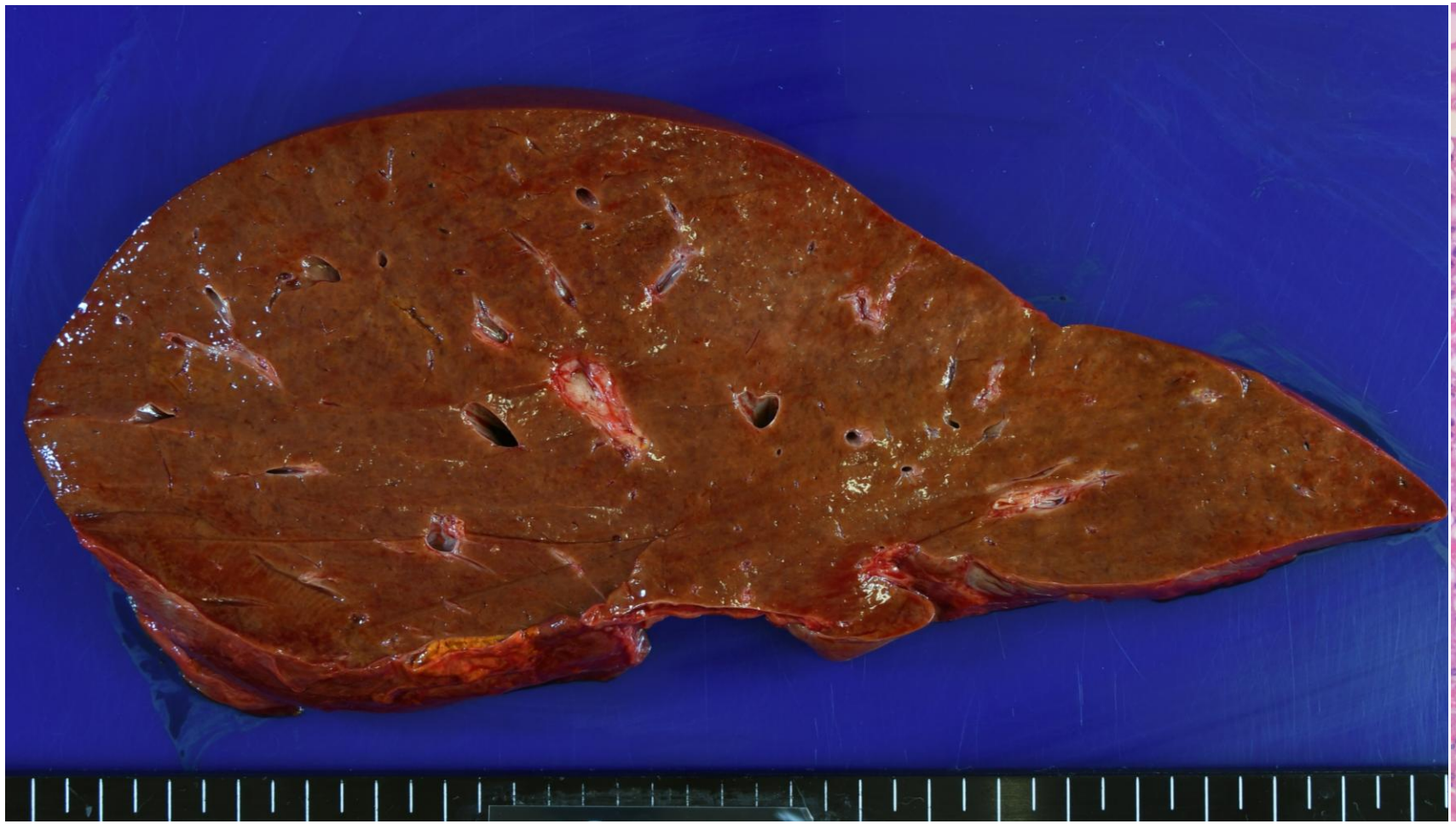
The heart weighing 320 g show mild left ventricular hypertrophy. The coronary artery remains intact. Mild hypertrophy of cardiomyocytes is seen microscopically (right: H&E).

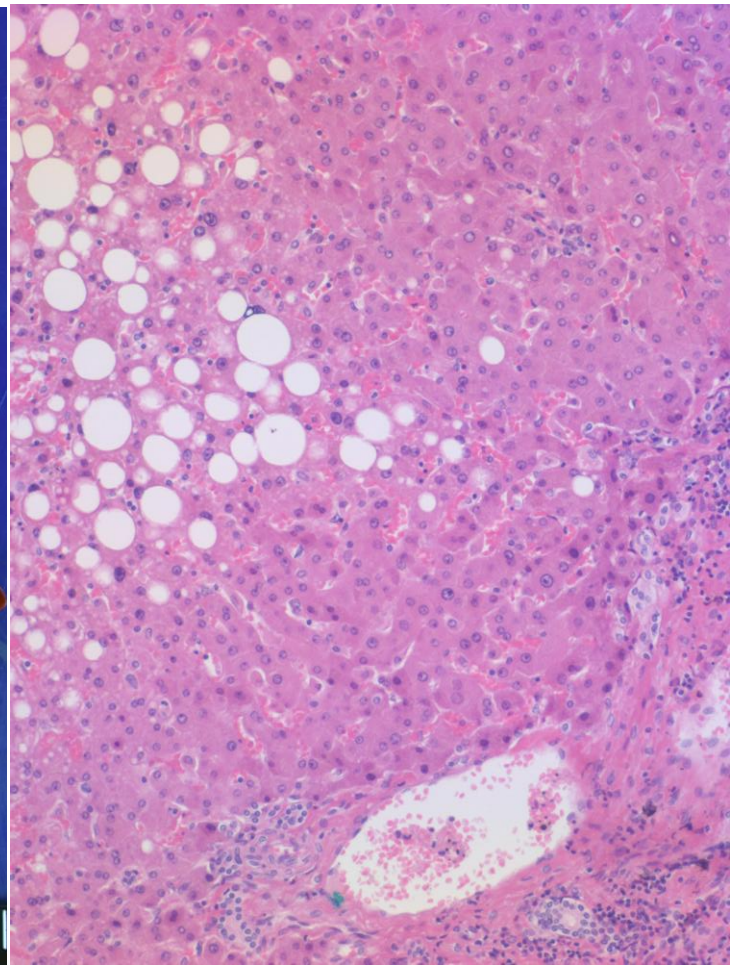


Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. Localized lymphangiectasis in the gastric fundus(gross appearance and H&E).

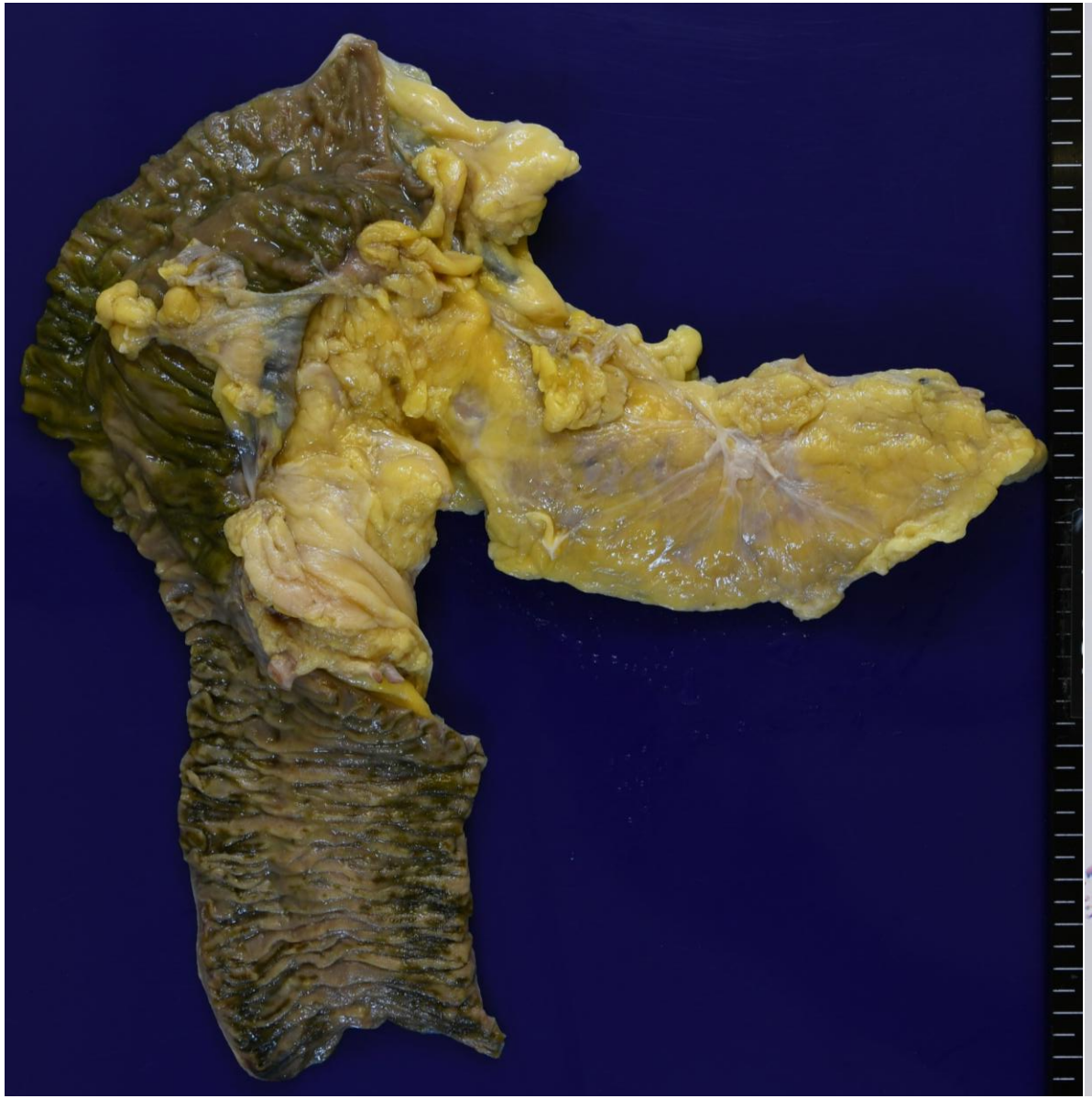


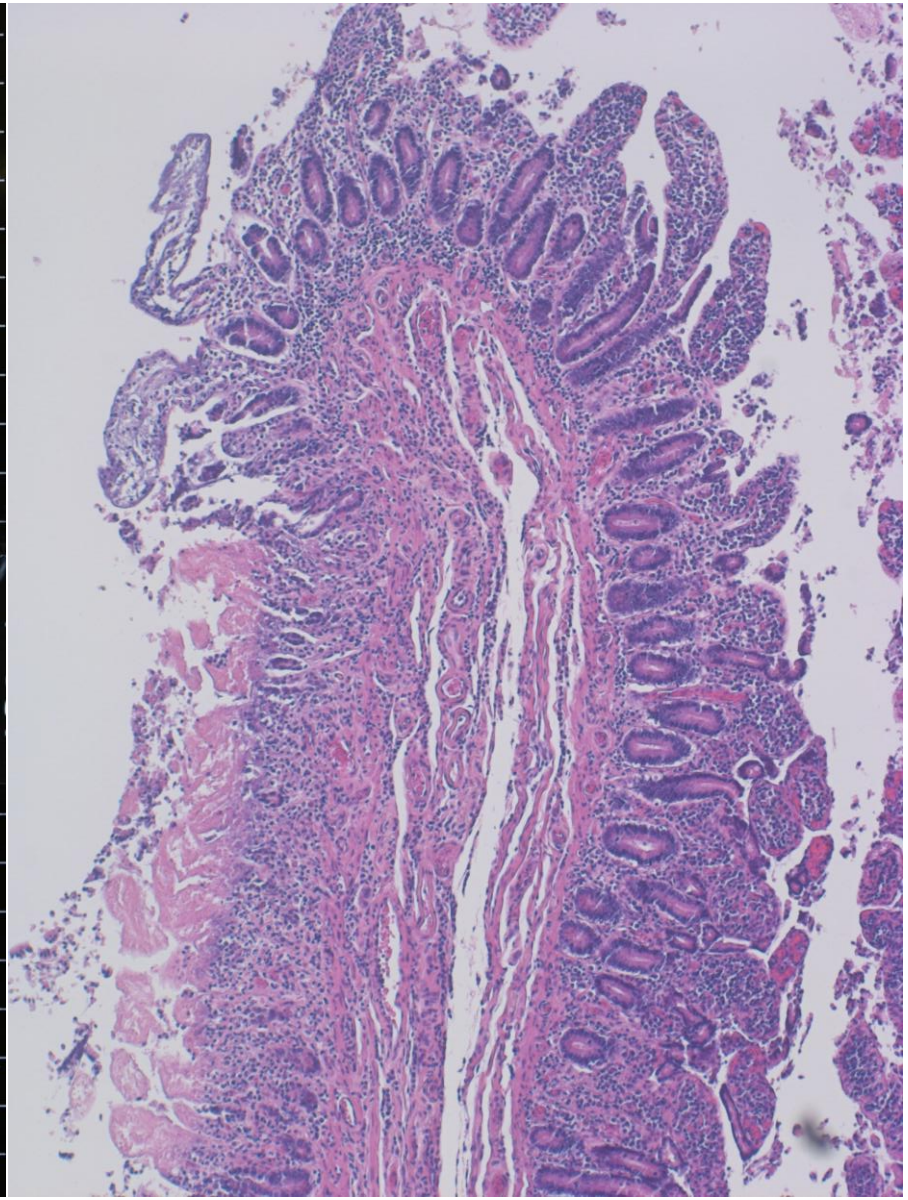
Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. The kidneys are unremarkable both grossly (left) and microscopically (right). The aorta reveals atherosclerosis with calcification (right inset, H&E).



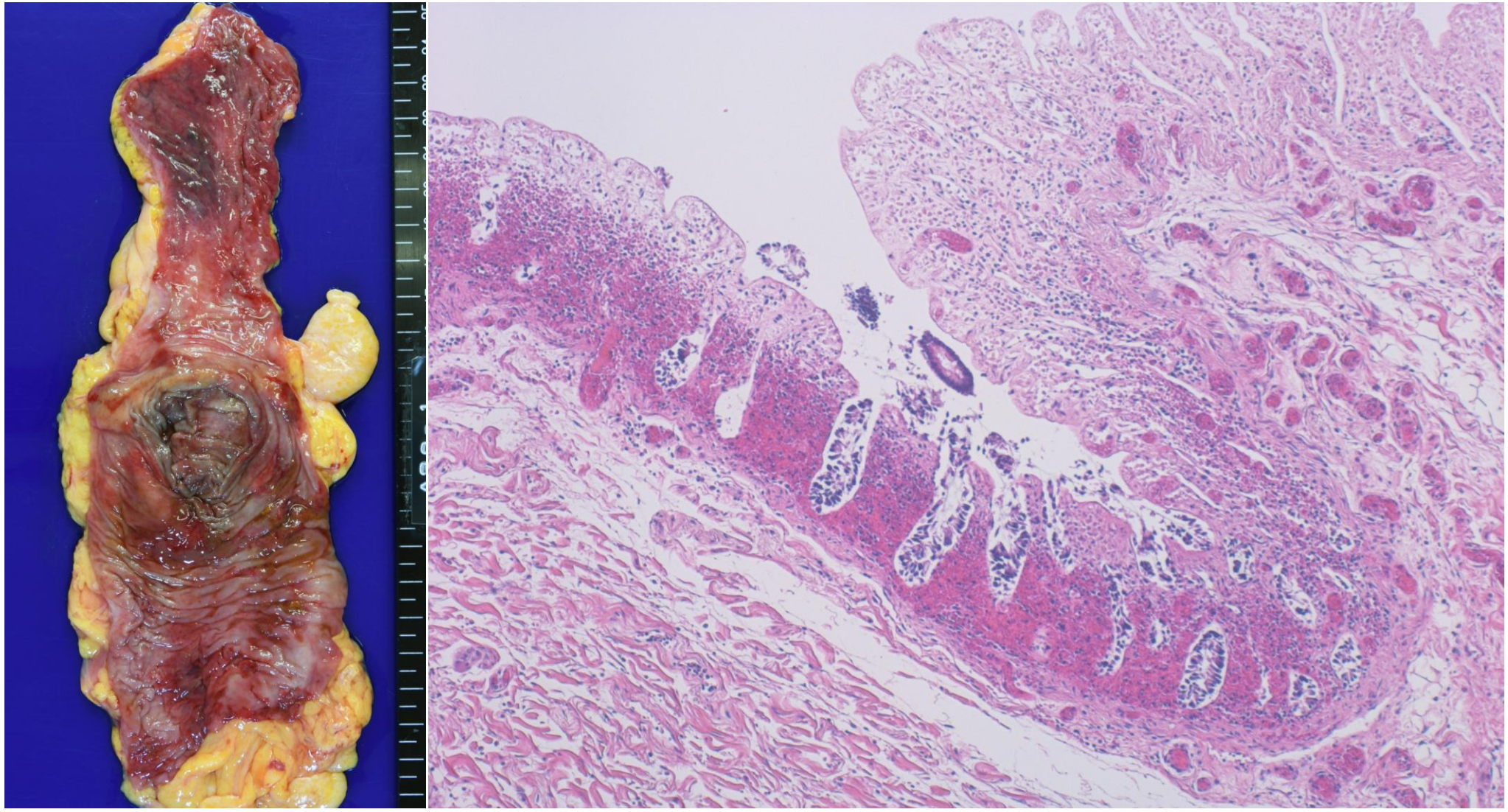


Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. The liver weighs 680 g.
Mild lipid deposition is seen in the centrilobular zone (right, H&E).

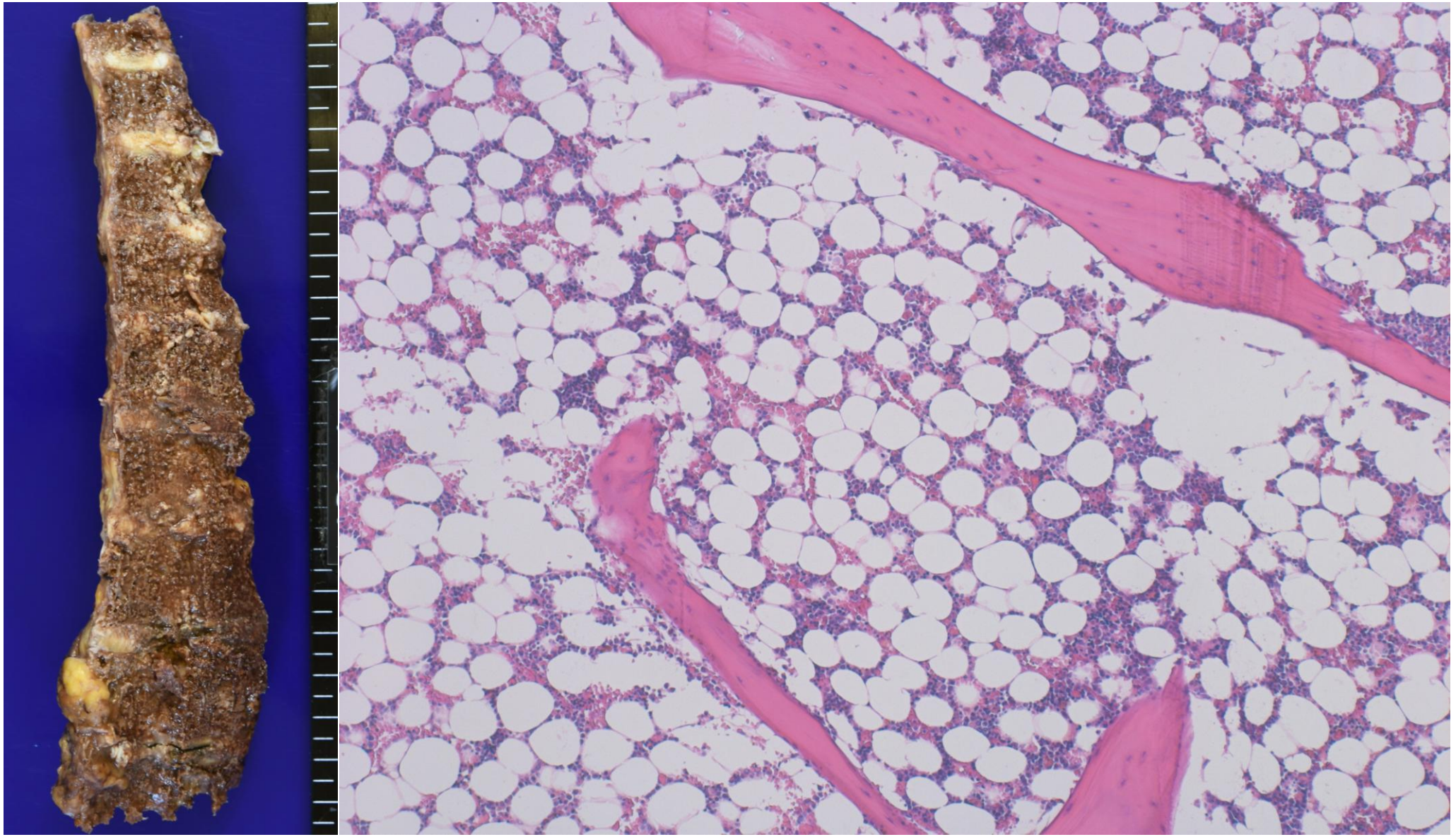




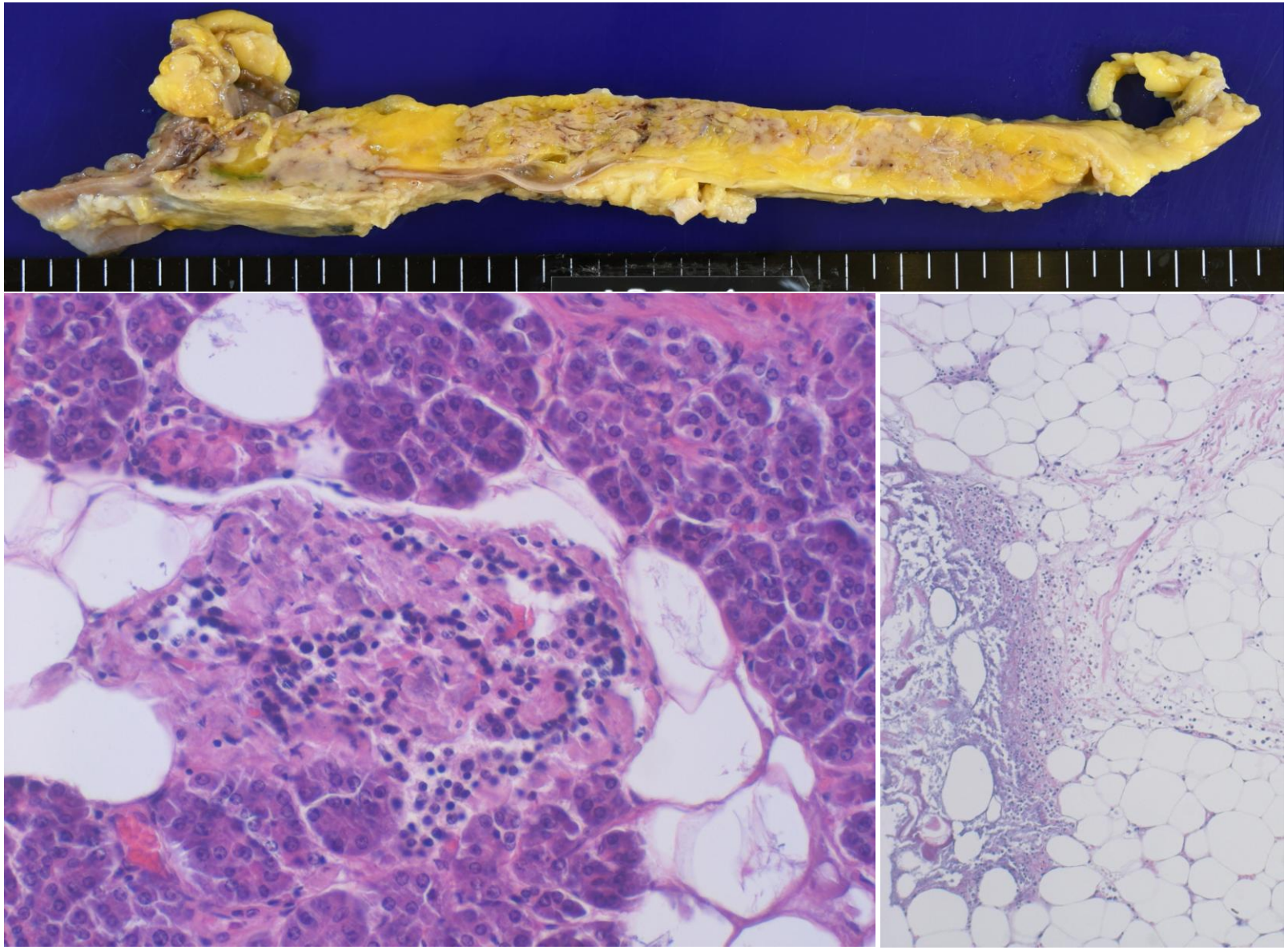
Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. The duodenum shows mucosal hemorrhage, and features of erosive duodenitis are seen microscopically (right: H&E). The pancreas is grossly unremarkable.



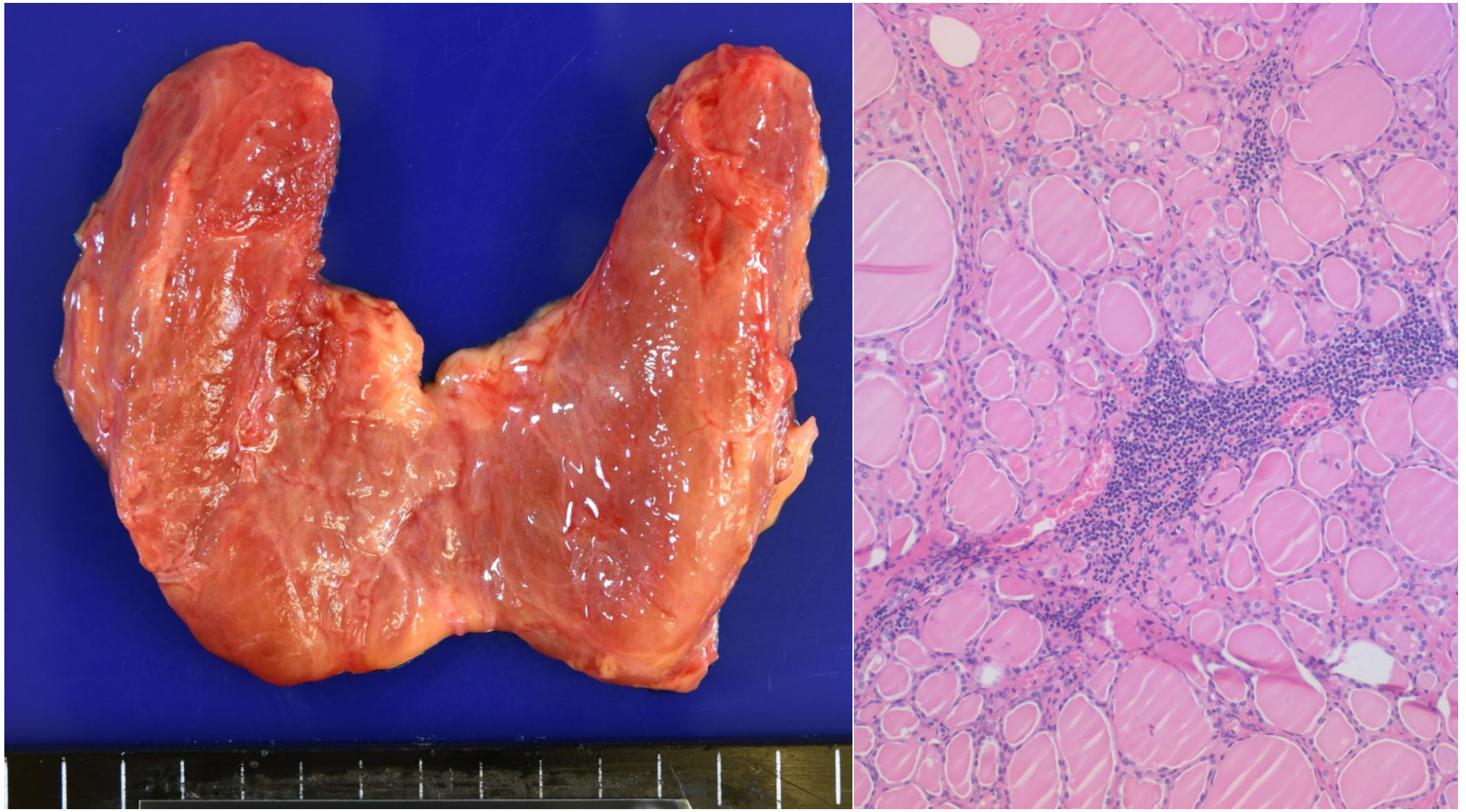
Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. The colon shows mucosal hemorrhage (left: gross finding).
Microscopically, intramucosal hemorrhage is observed (right: H&E).



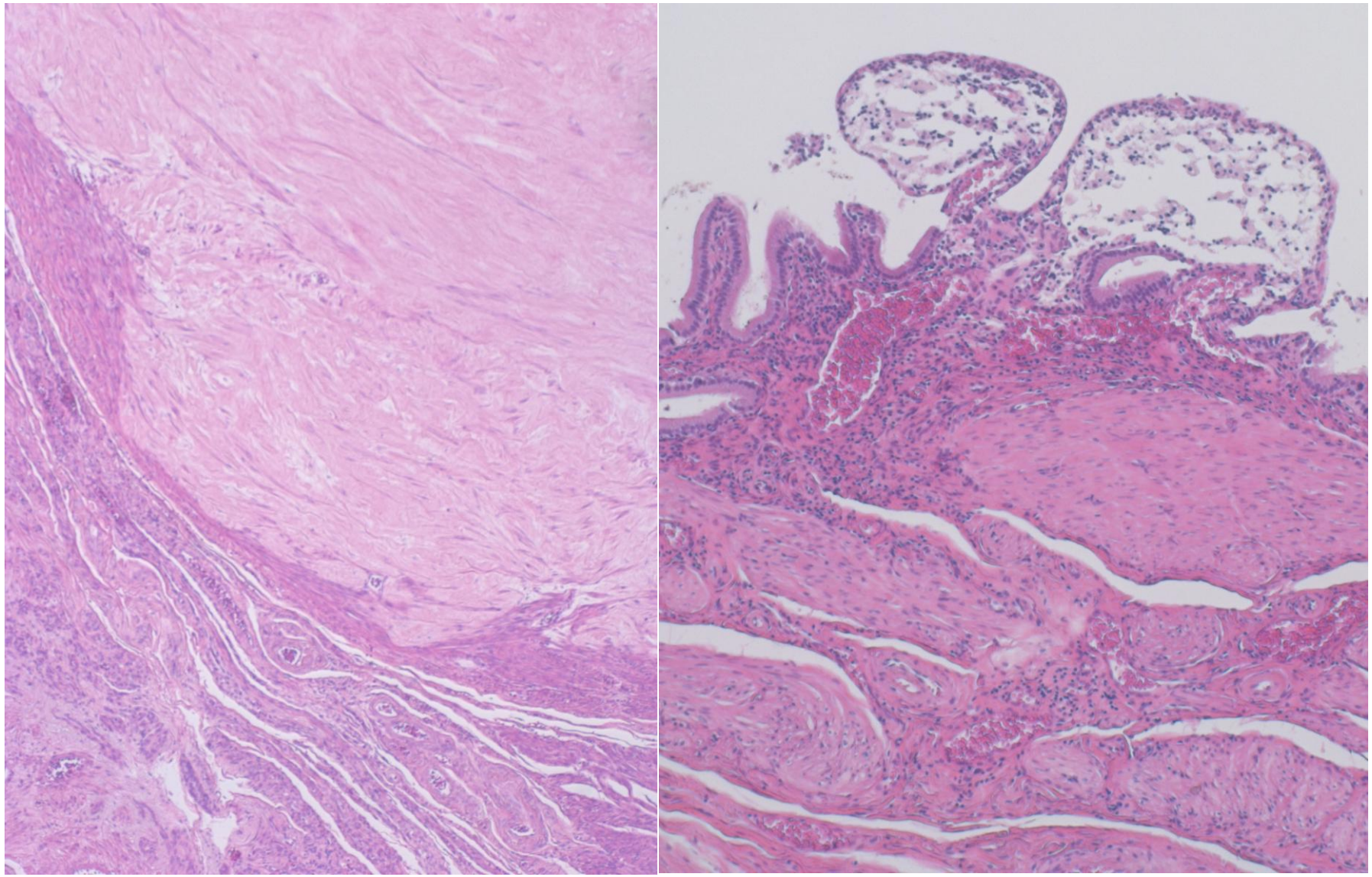
Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. Age-related osteoporosis is noted grossly (left) and microscopically (right: H&E).



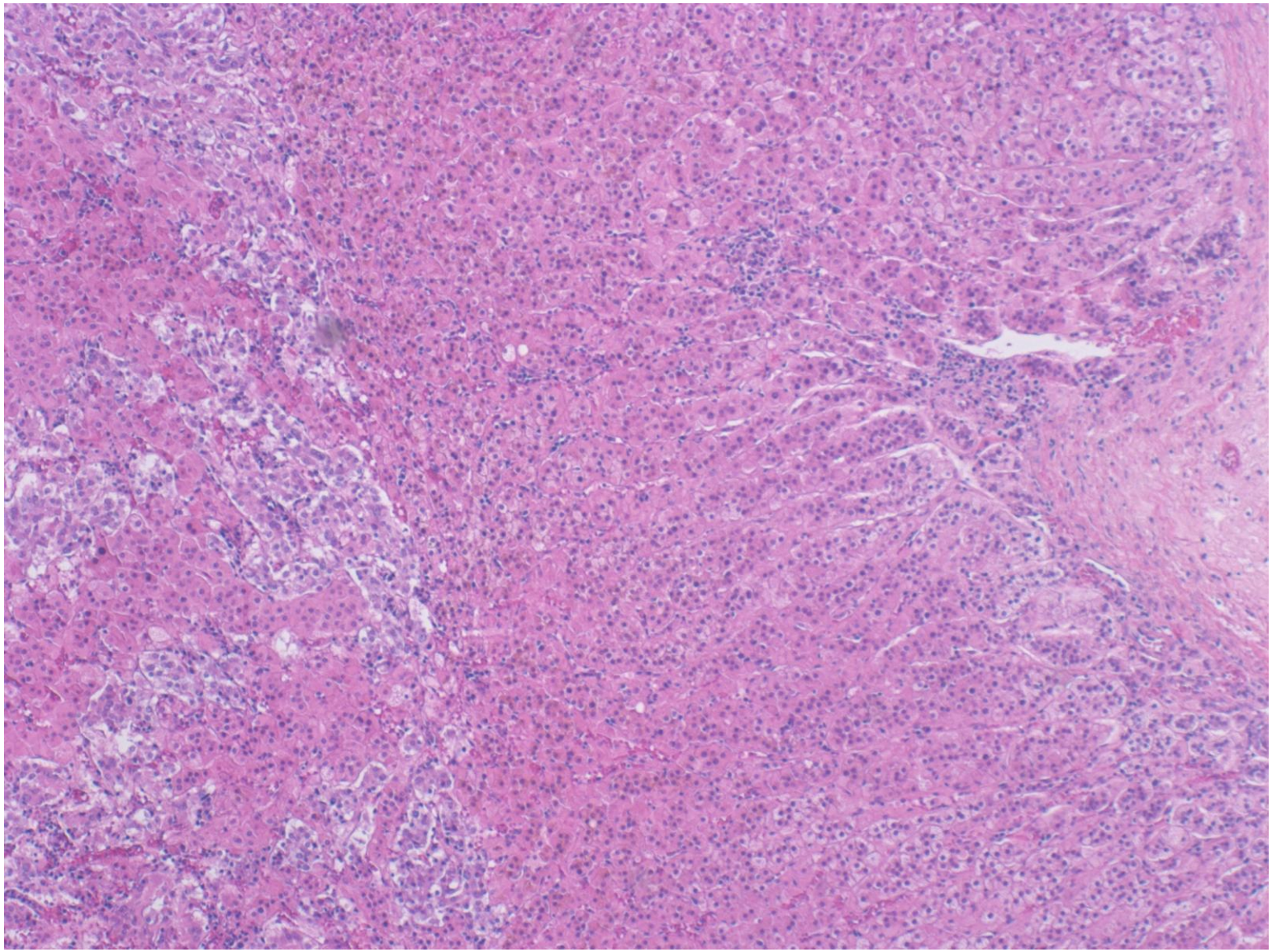
Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. The cut surface of the pancreas after formalin fixation is shown on the top. Microscopically, amyloid deposition of Langerhans islets is focally noted (bottom left: H&E). Fat necrosis is focally associated (bottom right: H&E).



Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. Lymphocytic thyroiditis (left: gross finding, right: H&E). The thyroid weighs 16 g.



Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. Hyalinized leiomyoma of the uterus (left) and cholesterolosis of the gallbladder mucosa (right) (H&E).



Idiopathic pulmonary fibrosis seen in a 90 y-o female patient. The adrenal cortex shows lipid depletion, representing a strong stress during the agonal phase (H&E).

Final anatomical diagnoses (90 y-0 female patient)

1. Idiopathic pulmonary fibrosis (UIP)
   a) diffuse interstitial fibrosis with honeycomb appearance
2. Aspiration pneumonia (bilateral lower lobes, marked)
   acute exacerbation for the last 33 days
   a) Bronchopneumonia
   b) Pleural effusion (left: 50 mL, right: 260 mL)
3. Middle lobe syndrome
4. Mucosal hemorrhage of the gastrointestinal tract
5. [type 2 diabetes mellitus]
   a) lipomatosis of the pancreas
   b) Amyloid deposition of the islets, mild
6. Left ventricular hypertrophy (heart: 320 g)
7. Leiomyoma of the uterus (multifocal, hyalinized and calcified)
8. Lymphangiectasia of the gastric fundus (focal)
9. Fatty change of the liver (centrilobular, mild, liver: 680 g)
10. Fat necrosis of pancreas, mild
11. Ascites retention (200 mL)
12. Cholesterolosis of the gallbladder
13. Lymphocytic thyroiditis (16 g)
14. Osteoporosis
15. Candidal glossitis
16. Aortic atherosclerosis with calcification

Direct cause of death: respiratory failure