CHEST

# A 56-Year-Old Woman With Multiple Pulmonary Cysts and Severe Chest Pain

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A 56-year-old woman presented to our hospital with a 4-month history of worsening chest pain. She denied having any respiratory symptoms, such as dyspnea, sputum, cough, or hemoptysis, or any history of smoking or exposure to dusts. One year previously she had a vertebral fracture. There was no specific family history, including pulmonary or autoimmune diseases. Chest CT performed 3 years earlier showed multiple thin-walled pulmonary cysts, although no further investigations were performed.

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Physical examination confirmed right upper chest pain in the second rib that worsened during inspiration. She was afebrile with a pulse rate of 75 beats/min, a respiratory rate of 16 breaths/min, BP measurement of 110/63 mm Hg, and 97% oxygen saturation with ambient air. Routine laboratory test results demonstrated mild anemia, hypercalcemia, renal dysfunction, and proteinuria. Serologic and microbiological investigations did not reveal any autoimmune or infectious diseases. The serum kappa free light chain level and kappa to lambda ratio were extremely elevated at 42,900 mg/L (normal range, 3.3-19.4 mg/L) and 5,720 mg/L (normal range, 0.26-1.65 mg/L), respectively. Both serum and urine immunoelectrophoresis detected the presence of kappa light chains. Bone marrow aspiration revealed plasmacytosis as high as 27.2%, which was consistent with multiple myeloma (MM).

Pulmonary function testing showed the following: FVC, 1.54 L (63% predicted); $FEV_1$, 1.12 L (57% predicted); $FEV_1$ to FVC ratio, 0.73; residual volume, 1.10 L (74% predicted); total lung capacity, 2.65 L (69% of predicted); and residual volume to total lung capacity ratio, 41.5 (143.1% predicted), which demonstrated a combined obstructive and restrictive defect with air trapping. Chest CT scans revealed multiple thin-walled cysts of various sizes and small nodules. The cysts showed basilar and peribronchovascular distribution. The number, size, and wall thickness of the cysts worsened in 3 years (Fig 1).

She underwent a video-assisted thoracoscopic lung biopsy. Histologic analysis revealed diffuse extracellular eosinophilic amorphous deposits that were dominant in the perivascular and pleural regions. The deposits were negative on Congo red staining and showed no apple-green birefringence under polarized light. Immunohistochemical staining for light chains within the eosinophilic materials was positive for kappa but not lambda light chains (Fig 2). Electron microscopy showed granular electron-dense deposits (Fig 3).

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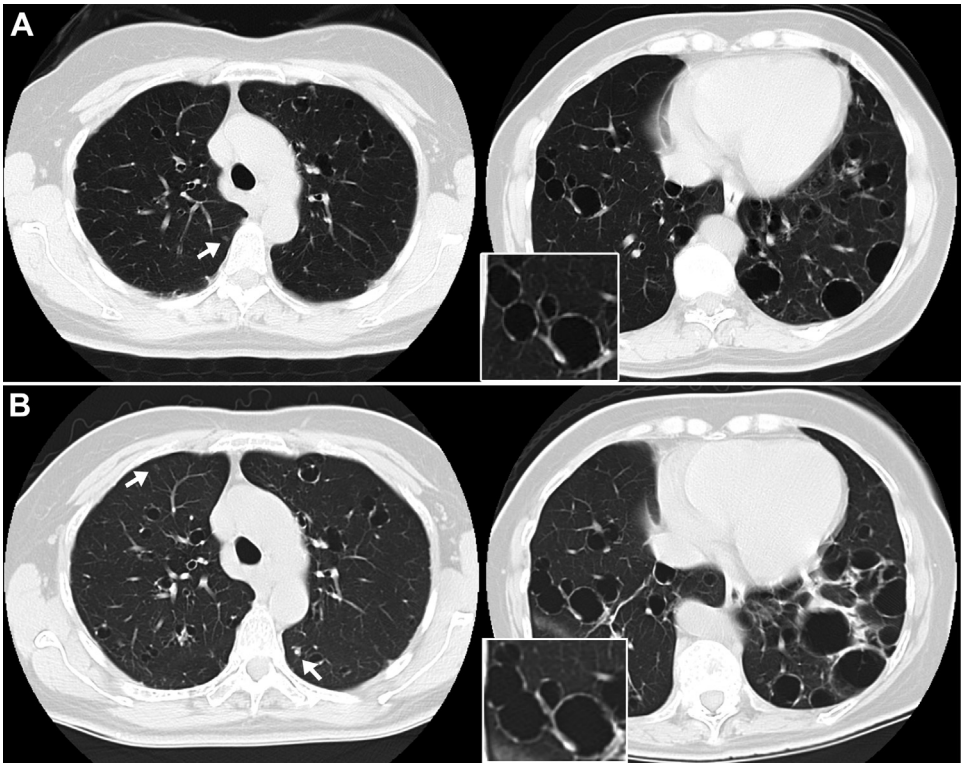


Figure 1 – *Chest CT imaging (A) 3 years before and (B) on admission showing multiple thin-walled cysts and small nodules (arrows). The size of the cysts was variable. The cystic lesions progressed in number, size, and wall thickness. No ground-glass opacities were seen. Inserts show higher magnifications of the cysts in right lower lobe.*

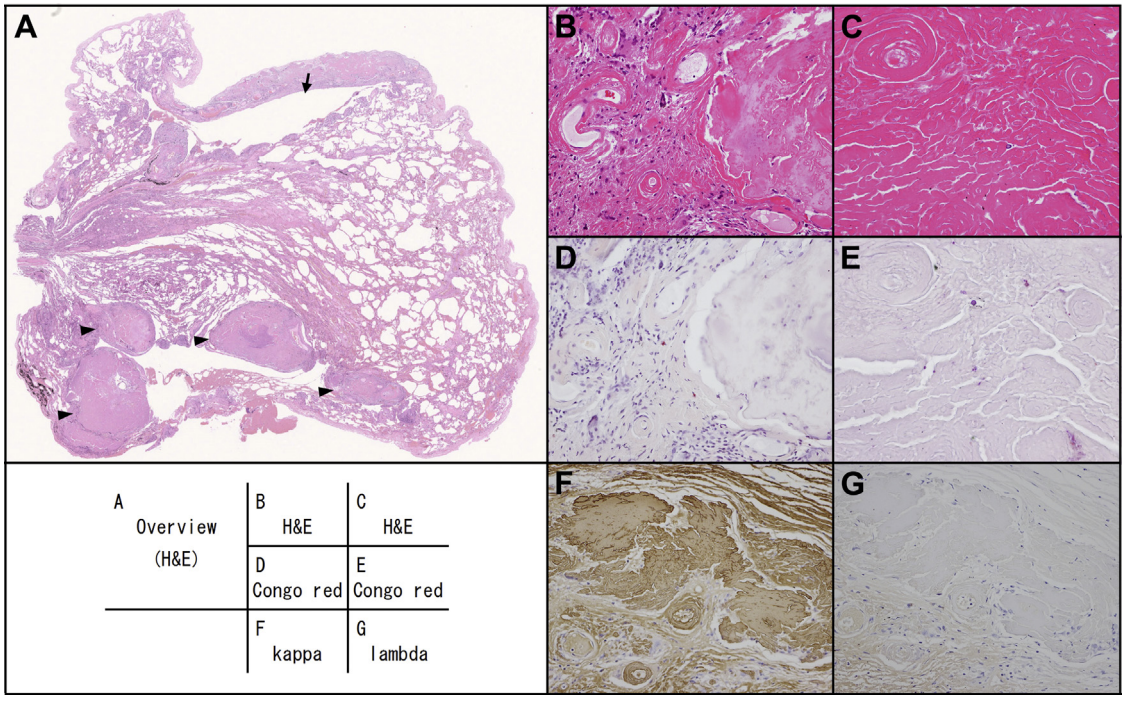


Figure 2 – *A, Diffuse deposits of amorphous eosinophilic materials in the peripheral lung and pleural regions with accompanying emphysematous cystic lesions (arrow) and nodular deposition (arrowheads) (H&E staining, panoramic view). B, Deposited materials in the pleural region (H&E staining, medium magnification). C, Highly magnified image of a nodular region of deposits (H&E staining). D and E, Deposits that are negative for Congo red staining (the serial sections to B and C, respectively). F, A deposit that is positive for kappa light chain immunostaining. G, A deposit that is negative for lambda light chain immunostaining.*