A 45-Year-Old Woman with Acute Interstitial Pneumonia (Hamman–Rich Syndrome)

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A 45-year-old woman with no medical history was admitted to an intensive care unit within six days of having non-productive cough and dyspnoea. She had previously been prescribed with amoxicillin for presumed community-acquired pneumonia. Her chest X-ray revealed diffuse interstitial opacities and patchy airspace opacities (Figure 1a). A computed tomography (CT) scan of her chest revealed interlobular septal thickening with diffuse ground-glass opacities (Figure 1b). All cultures, including those of blood, urine, sputum, viral, fungal, and other atypical organisms, were unremarkable. Bronchoscopy revealed that the airways of the right and left lungs appeared normal. The bronchoalveolar lavage (BAL) fluid differential cell count was 77% neutrophils and 23% monocytes. All cultures and serologies from the BAL fluid were unremarkable, and methylprednisolone treatment was initiated. The patient failed to improve after receiving broad-spectrum antibiotics and steroids and required endotracheal intubation for hypoxemic respiratory failure; she required maximal mechanical ventilator support. Her secretion quality remained minimal and unremarkable post intubation. Her respiratory status continued to deteriorate, causing paralysis and necessitating inhaled epoprostenol. Her acute respiratory distress syndrome (ARDS) remained refractory to all treatments, and extracorporeal membrane oxygenation (ECMO) was initiated. After nine days of ECMO support, she passed away after her family opted to withdraw life support and transition goals of care towards comfort. Based on the idiopathic nature of our patient’s ARDS, despite receiving a thorough diagnostic workup, she was diagnosed with acute interstitial pneumonia (AIP) (Hamman-Rich Syndrome). AIP, which is a diagnosis of exclusion only after infections and other aetiologies have been ruled out, is classified as idiopathic interstitial pneumonia and is an uncommon and rapidly progressive form of diffuse lung injury originally described by Hamman and Rich in 1935 (1-3). AIP, which is not associated with cigarette smoking, typically affects healthy individuals over the age of 40 years (mean age: 50–55 years) without a history of lung disease and occurs with equal frequency in men and women (3). The in-hospital mortality rate due to AIP is more than 50%, with most of those who survive the initial hospitalization dying within six months of presentation (3).

References