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Idiopathic Pleuroparenchymal Fibroelastosis (IPPFE), a rare clinic-pathologic entity that needs more attention: Series of Egyptian patients

Dalia Abd El-Kareem
Cairo University, Egypt

Idiopathic Pleuroparenchymal Fibroelastosis (IPPFE) is an entity recently classified by the American thoracic society/ European respiratory society as a rare Idiopathic Interstitial Pneumonia (IIP). This might be uncertain, based on some clinical experiences. As part of a large Egyptian study that included patients with Diffuse Parenchymal Lung Diseases (DPLD), we encountered 6 patients with DPLD diagnosed as IPPFE by surgical lung biopsy over a period of one year. Clinical data, High Resolution chest Computed Tomography (HRCT) findings and histologic criteria from thoracoscopic lung biopsies were correlated in a multidisciplinary approach. Most of our patients were young age, with female predominance and living in the same area (Upper Egypt). Exertional dyspnea and cough were the main presenting symptoms. Low body weight, flat chest wall and stretched skin were the main signs. HRCT showed upper lobe volume loss, traction bronchiectasis, visceral pleural thickening and ground glass opacity. Histologic features included thickening of the visceral pleura, sub-pleural parenchyma and interlobular septa with deposition of large amounts of elastic fibers (by elastic stain) and Non-Specific Interstitial Pneumonia (NSIP) histologic pattern. IPPFE should get more attention being more prevalent than we used to know. Although other IIPs may represent initial phase in IPPFE or may coexist together but it should still be considered as a separate entity. Multidisciplinary approach is required for diagnosis. Further studies to reach etiologic factors are highly needed.

Biography

Dalia Abd El-Kareem is a Lecturer of Pathology at Faculty of Medicine, Cairo University in Egypt. She has completed her MD in Pathology and Pulmonary Pathology in 2016. She has completed her Graduation from Medical School in 2007 from Cairo University. She has received training in Pulmonary Medicine and Respiratory ICU at Cairo University Hospitals and then joined Department of Pathology, completed her Pathology training and received MSc degree in Pathology in 2013. She was also trained in the field of Pulmonary Pathology at University of Texas Medical Branch (UTMB), Galveston, USA.

dalia_ak@kasralainy.edu.eg