Journal of Pathology and Disease Biology

Extended Abstract
Vol. 3, Iss. 2
2019

Pathology Congress 2017: Idiopathic Pleuroparenchymal Fibro elastosis (IPPFE), a rare clinic-pathologic entity that needs more attention: Series of Egyptian patients - Dalia Abd El-Kareem - Cairo University, Egypt

Dalia Abd El-Kareem
Cairo University, Egypt.

Idiopathic pleura parenchymal fibro elastosis (IPPFE) is a unit freshly secret by the American thoracic society / European respiratory society as a rare idiopathic interstitial pneumonia (IIP). This could be uncertain, based on certain clinical experiences. In a huge Egyptian study counting patients with verbose parenchymal lung disease (DPLD), we met 6 patients with DPLD diagnosed as IPPFE by surgical lung biopsy over a period of one year. The clinical data, the results of the high resolution thoracic computed tomography (HRCT) and the histological criteria of the thoracoscopic pulmonary biopsies were correlated in a multidisciplinary approach. Most of our patients were young, predominantly female and living in the same region (Upper Egypt). Exercise dyspnea and cough were the main presenting symptoms. Low body weight, a flat chest wall and tight skin were the main signs. HRCT has shown loss of upper lobe volume, traction bronchiectasis, visceral pleural thickening, and opacity of the frosted glass. Histological features included thickening of the visceral pleura, sub pleural parenchyma and interlobular septa with deposition of large amounts of elastic fibers (by elastic staining) and the histological pattern of nonspecific interstitial pneumonia (NSIP). IPPFE should receive more attention being more widespread than we knew. Although other IIPs may represent the initial phase of IPPFE or coexist together, they should nevertheless be considered as a separate entity. A multidisciplinary approach is required for diagnosis. Further studies to achieve etiological factors are needed.

Pleuroparenchymal fibro elastosis (PPFE) is a rare pulmonary fibrosis which is characterized clinically by a predominant fibrosis of the upper lobe. PPFE is a slowly progressing disorder and its first symptom is dyspnea or a dry cough. Chest pain owing to pneumothorax might be the primary symptom in some patients. Pleuroparenchymal fibro elastosis (PPFE) is a infrequent disorder categorized by pleural and parenchymal fibrosis underlying the upper lobe, the latter being intra-alveolar with an elastosis accompanying the alveolar walls. The purpose of this study was to review cases meeting the published imaging and histological criteria, and to identify any common clinical features that might suggest an underlying etiology for a condition that was previously considered to be 12 patients (seven women, median age 57 years). , the symptoms presented were shortness of breath (11 of 12 patients) and dry cough (six of 12 patients). Seven patients reported recurrent infections during their illness. Five have demonstrated non-specific autoantibody positivity. Two patients obligated a domestic past of interstitial lung disease (ILD).

High-resolution computed tomography of lung disease distant from the parenchymal pleura was present in six of 12 patients (coexisting fibrosis, n = 5; bronchiectasis, n = 1). Out of seven patients whose tissues were taken from the lower lobes, four patients presented less intense changes in PPFE (one with additional characteristics of hypersensitivity pneumonitis) and three presented usual interstitial pneumonia. PPFE is a distinct clinical pathological entity, clinical data suggesting a link with recurrent pulmonary infection. Genetic and autoimmune mechanisms can also contribute to the development of these changes. The PPFE can also have a more diffuse implication than that previously reported and coexist with different models of ILD.

Pleuroparenchymal fibro elastosis (PPFE) is an unusual lung disease with unique clinical, radiological and pathological features. Elected a sporadic idiopathic interstitial pneumonia in 2013, its name raises to a combination of fibrosis involving the visceral pleura and fibro-elastic changes predominant in the sub pleural pulmonary parenchyma. Though a sum of disease

This work is partly presented at 14th Asia Pacific Pathology Congress, November-13-14, 2017 held at Osaka, Japan.

connotations have been described, no single cause of PPFE has been unequivocally identified. A diagnosis of PPFE is most often made by identifying the characteristic anomalies on the CT scans. The first changes are systematically localized in the upper lobes near the pulmonary apices, the same places where the subsequent progression of the disease is also most visible. When severe enough, the disease causes a progressive loss of volume in the upper lobes which, in combination with a decrease in body mass, produces a lamellar thorax. Formerly measured as a gradually progressing entity, it is now documented that around patients with PPFE follow an inexorably progressive course which leads to irreversible respiratory failure and early death. In the absence of effective drug therapy, lung transplantation remains the only treatment option for this disorder. This review focuses on improving the early recognition of the disease and the evaluation of its pathophysiological impact and discusses working approaches for its management.

It has been recommended that acute or subacute lung damage, counting diffuse alveolar damage, causing exuberant interstitial inflammation, is at the heart of the pathological cascade culminating in PPFE. However, the exact nature of the harmful stimuli involved in triggering this process remains unknown. The presence of diffuse alveolar lesions has also been reported in the context of post-transplant PPFE. The failure of parenchymal lung lesions to adequately resolve the risks favoring the repair of aberrant tissues, which can leave dense and permanent obliterated fibro elastosis and atelectasis which ultimately contribute to the development of PPFE. PPFE and alveolar fibro elastosis developing in post-transplant restrictive allograft syndrome (SAR) have common pathological and genetic characteristics. The reasons why such injuries should produce well-defined chronic fibrous anomalies, mainly sub-pleural, rich in elastin, remain unexplained.

There is currently no cure for pulmonary fibrosis, but treatments and therapies are constantly improving. The average life expectancy of someone with pulmonary fibrosis is three to five years, but if detected early, treatment can help slow the progression of the disease. Pleural condensing happens once the lining of the lungs thickens by scar tissue. Pleural thickening is usually first detected through a chest x-ray, but a CT scan can detect the condition earlier.

Biography: Dalia Abd El-Kareem is a lecturer in pathology at the Faculty of Medicine at the University of Cairo in Egypt. She obtained her doctorate in pathology and pulmonary pathology in 2016. She obtained her medical degree in 2007 at the University of Cairo. She received training in pulmonary medicine and respiratory intensive care at Cairo university hospitals, then joined the pathology department, completed her pathology training and obtained a master's degree in pathology in 2013. She was also trained in the field of pulmonary pathology at the University of Texas Medical Branch. (UTMB), Galveston, United States.

This work is partly presented at 14th Asia Pacific Pathology Congress, November-13-14, 2017 held at Osaka, Japan.