Unilateral IPF- A Rare Occurrence

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Abstract: Idiopathic Pulmonary Fibrosis (IPF) is a chronic non – neoplastic lung disease, which is rapidly progressive in nature, having a poor prognosis. It is restricted to the lungs and occurs in the elderly. We present a case of a 71 year old female, with progressive shortness of breath and dry cough. High resolution computed tomography (HRCT) of the chest revealed unilateral IPF of the right lung. She also had a grade 3 hiatus hernia, which only fuels the speculation of GERD being an important risk factor for the development of IPF. Till date only a handful of such cases have been reported.

Key Words: idiopathic pulmonary fibrosis (IPF), hiatus hernia, high resolution computed tomography (HRCT), micro-aspiration

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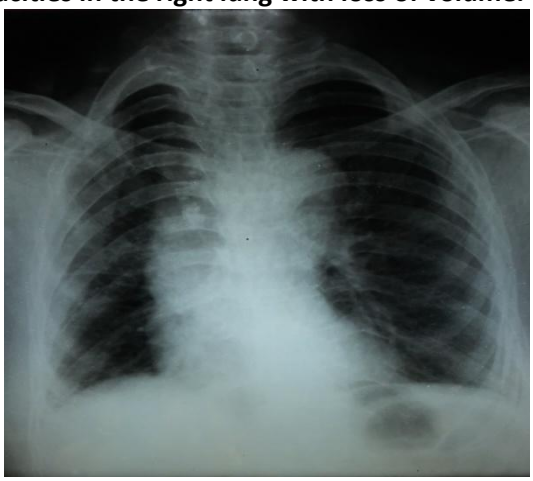
Introduction: Interstitial lung disease (ILD), also known as diffuse parenchymal lung disease (DPLD), refers to a group of lung diseases affecting the interstitium (the tissue and space around the air sacs of the lungs). It concerns alveolar epithelium, pulmonary capillary endothelium, basement membrane, perivascular and perilymphatic tissues. One of the main types of ILDs is IPF. Idiopathic pulmonary fibrosis (IPF) is defined as a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, and limited to the lungs. It is characterized by progressive worsening of dyspnea and lung function and is associated with a poor prognosis. Such is the debilitating nature of the disease that patients hardly survive for 3 or 4 years despite treatment. Asymptomatic GERD has been postulated as a risk factor for the progression of IPF.

CASE STUDY: A71year old woman presented to our out-patient department with complaints of progressive breathlessness, corresponding to MMRC grade 3 and dry cough. Physical examination revealed her vitals to be stable. She had a pulse of 84 per minute, a saturation of 94% and a blood pressure of 120/70mmHg. She had grade 2 clubbing and on respiratory system examination, she had fine end inspiratory crepitations all over the right hemithorax. A chest X-ray posterior anteriorview showed ill-defined opacities in the right lung.

She was further evaluated with High resolution computed tomography (HRCT) of thorax, which showed interlobular and intralobular septal thickening with honeycombing present in the right lung, predominantly in the basal areas. There was complete sparing of the left lung parenchyma. An incidental finding of hiatus hernia was also picked up. On comparative assessment with previous HRCT images, gradual progression of the right lung lesions was noted.

2 dimensional echocardiography of the heart was normal. Connective tissue disease profile was negative. The patient was further subjected to esophagogastroduodenoscopy which showed a grade 3 hiatus hernia. She was accordingly started on proton pump inhibitors with dietary modifications.Apart from respiratory complaints, she was also suffering from diabetes mellitus. Her glucose levels were controlled oral hypoglycemic medications.

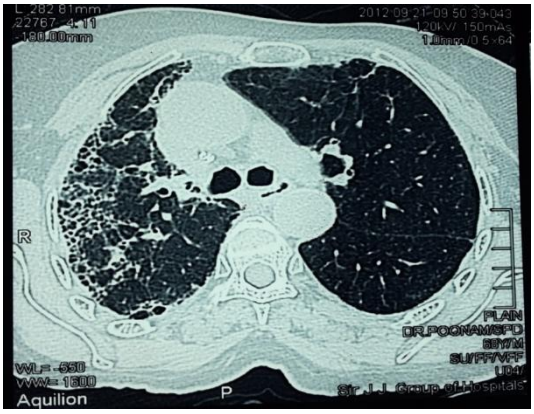
Figure 1 : Chest X ray PA view showing ill defined opacities in the right lung with loss of volume.

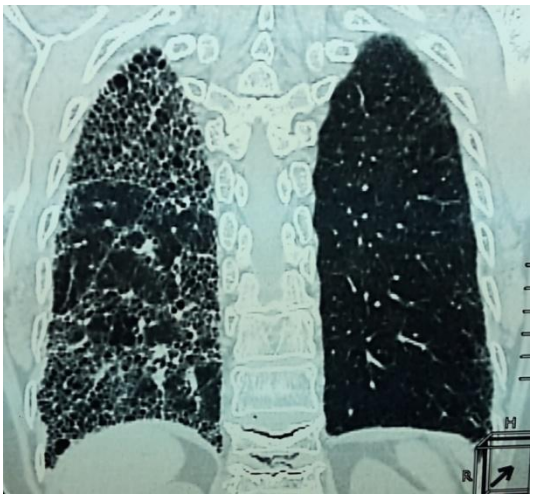


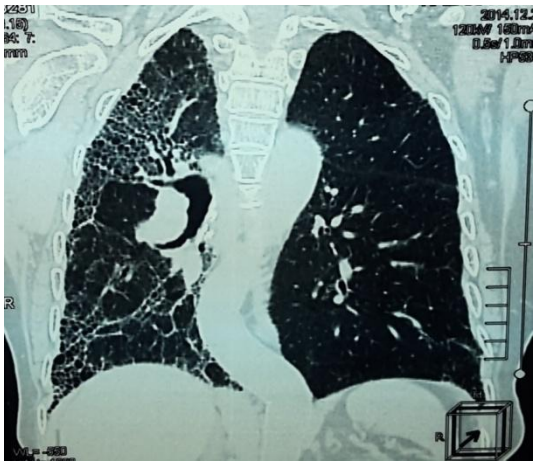
DISCUSSION: Interstitial lung disease (ILD), also known as diffuse parenchymal lung disease (DPLD)[1] refers to a group of lung diseases affecting the interstitium (the tissue and space around the air sacs of the lungs). It concerns alveolar epithelium, pulmonary capillary endothelium, basement membrane, perivascular

and perilymphatic tissues. One of the most common and fatal types of ILD is Idiopathic Pulmonary Fibrosis(IPF). Idiopathic pulmonary fibrosis (IPF) is defined as a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, and limited to the lungs.

Figure 2 a,b,c : HRCT Thorax showing the unilateral IPF. Complete sparing of the left lung can be appreciated







It is characterized by progressive worsening of dyspnea and lung function and is associated with a poor prognosis. It is associated with the histopathologic and/or radiologic pattern of Usual Interstitial Pneumonia(UIP)(2, 3, 4). The definition of IPF requires the exclusion of other forms of interstitial pneumonia including other idiopathic interstitial pneumonias and ILD associated with environmental exposure, medication, or systemic disease.

However by convention, IPF is generally considered to be a bilateral process. Unilateral affection of the pulmonary parenchyma is a very rare occurrence. Rare associations have been made between unilateral IPF and connective tissue diseases especially systemic scleroderma (5), as well as pulmonary vein thrombosis (6). Our patient was evaluated for the presence of underlying connective tissue diseases, the results of which were negative. She also did not have any pulmonary vessel, cardiac or airway pathology as demonstrated by HRCT Thorax and a 2D ECHO.

Idiopathic pulmonary fibrosis (IPF) is a rapidly progressive, fatal disease of unknown cause. Although there is no known cure, there have been indications that gastroesophageal reflux disease (GERD) is often present and may contribute to disease progression, possibly due to micro-aspiration of reflux gastric contents (7). Emerging data support a role for chronic micro-aspiration (i.e. subclinical aspiration of small droplets) in the pathogenesis and natural history of idiopathic pulmonary fibrosis. However, the precise relationship between chronic micro-aspiration and idiopathic pulmonary fibrosis remains unknown. Gastroesophageal reflux, a presumed risk factor for micro-aspiration, has been strongly associated with idiopathic pulmonary fibrosis with an estimated prevalence of 90% (8). Our patient also had grade 3 hiatus hernia, which was an incidental finding. The hernia was most probably the cause of reflux, which has led to the progression of the IPF.

We firmly believe defining the precise relationship between micro-aspiration and idiopathic pulmonary fibrosis is critically important because of its potential pathobiological and therapeutic implications. In general, current treatment strategies in idiopathic pulmonary fibrosis have focused on modulating the fibrotic tissue response after the injury, not on preventing the injury itself. Micro-aspiration may represent a source of repetitive injury in idiopathic

pulmonary fibrosis and may be modifiable with medical and/or surgical therapy.

Declaration Of Conflicting Interest : The authors declare that they have no conflicting interests.

References:

1. King TE (August 2005). "Clinical advances in the diagnosis and therapy of the interstitial lung diseases". Am. J. Respir. Crit. Care Med. 172 (3): 268–79.
2. American Thoracic Society; European Respiratory Society. Idiopathic pulmonary fibrosis: diagnosis and treatment: international consensus statement. Am J RespirCrit Care Med 2000;161:646–664.
3. American Thoracic Society, European Respiratory Society. American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias. Am J RespirCrit Care Med 2002;165:277–304.
4. Visscher DW, Myers JL. Histologic spectrum of idiopathic interstitial pneumonias. Proc Am ThoracSoc2006;3:322–329.
5. Viktor N. Lesnyak, Olesya V. Danilevskaya, Alexander V. Averyanov, and Lidia P. Ananyeva"Unilateral Pulmonary Fibrosis and Systemic Sclerosis", American Journal of Respiratory and Critical Care Medicine, Vol. 190, No. 9 (2014), pp. 1067-1068.
6. Raquel A Cavaco*, Sunny Kaul, Timothy Chapman, Romina Casaretti, Barbara Philips, Andrew Rhodes and Michael R Grounds Idiopathic pulmonary fibrosis associated with pulmonary vein thrombosis: a case report. *Cases Journal* 2009, 2:9156
7. Lee JS, Collard HR, Raghu G, et al. Does chronic microaspiration cause idiopathic pulmonary fibrosis Am J Med. 2010;123:304-311.
8. Joyce S. Lee, MD, Harold R. Collard, MD, Ganesh Raghu, MD, Matthew P. Sweet, MD, MS, Steven R. Hays, MD, Guilherme M. Campos, MD, FACS, Jeffrey A. Golden, MD, and Talmadge E. King Jr., MD Does Chronic Microaspiration Cause Idiopathic Pulmonary Fibrosis? Am J Med. 2010 April ; 123(4): 304–311