Introduction

The idiopathic interstitial pneumonias (IIPs) are a group of diffuse parenchymal lung diseases (DPLDs) commonly referred as interstitial lung diseases (ILD). They result from lung parenchymal injury due to inflammation and fibrosis and though they primarily affect the interstitium, the airspaces, peripheral airways and vessels also can get affected. [1] IIPs share similar clinical, radiologic and histopathologic features [2]. IIP consists of major IIPs, rare IIPs and unclassifiable IIPs. Idiopathic pulmonary fibrosis (IPF), non-specific interstitial pneumonia (NSIP), respiratory bronchiolitis-interstitial lung disease (RB-ILD), cryptogenic organizing pneumonia (COP), desquamative interstitial pneumonia (DIP) and acute interstitial pneumonia (AIP) are classified under major IIPs whereas idiopathic lymphoid interstitial pneumonia (ILIP) and idiopathic pleuroparenchymal fibroelastosis (IPPF) are grouped as rare IIPs (Figure 1) [3].