



BMH Med. J. 2017;4(4):122-125 **Case Report**

## **Air Leak Syndrome An Unusual Presentation Of Desquamative Interstitial Pneumonitis**

Shaik Mastan, Neena Jacob, Nasser Yusuf, Shehla Basheer, CP Rauf

Chest Hospital, Kozhikode, Kerala, India

**Address for Correspondence:** Dr. Shaik Mastan, Chest Hospital, Kozhikode, Kerala, India. Email: mastansk84@gmail.com

### **Abstract**

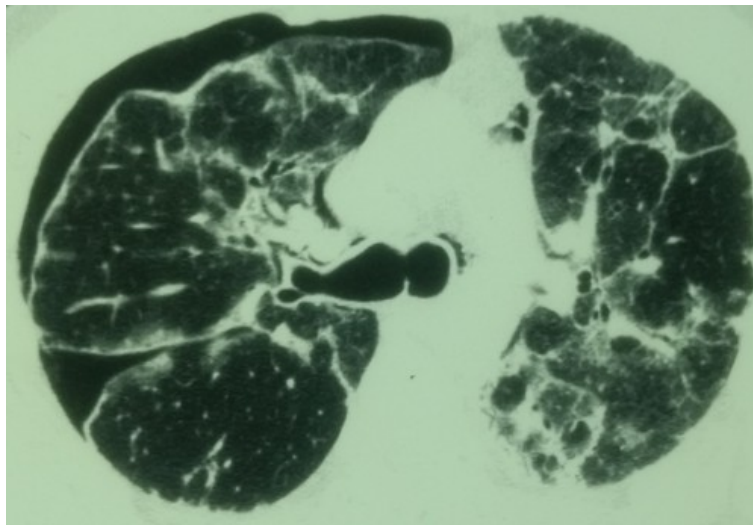
Desquamative interstitial pneumonia (DIP) is a less common type interstitial lung disease, a type of idiopathic interstitial pneumonias, which has frequent association with smoking exposure and has good response to corticosteroid therapy. Here we are reporting a case of DIP presented as pulmonary air leak syndrome which was managed by chest tube drainage and steroid therapy.

**Keywords:** Air Leak Syndrome; Desquamative interstitial pneumonia

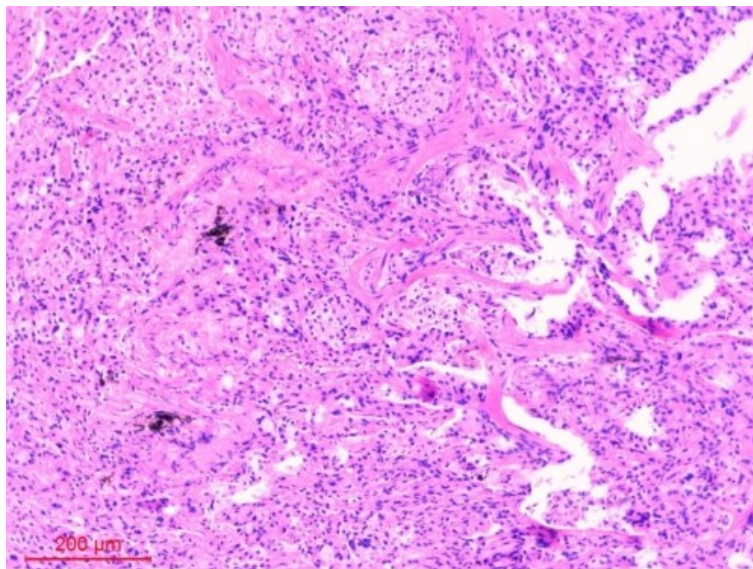
A young male presented to emergency department with sudden increase in dyspnea and chest discomfort for the last 2 days. There was history of two and a half months fever, dry cough and progressive dyspnea. He was a non smoker but there was exposure to passive smoking for 9 months. He was not having exposure to organic or inorganic dusts or fumes or drugs. He was admitted and evaluated for his breathlessness 1 month back elsewhere and was treated symptomatically (**Figure 1**). On examination, he was tachypneic, with a respiratory rate of 34/min, Pulse rate 94/min, blood pressure 110/70 mm Hg, SPO2 86% on room air, decreased breath sounds on right hemithorax and basilar rales on left side. Laboratory investigations including peripheral blood cell counts, liver, and renal function tests were within normal limits. Serologic tests for antinuclear auto antibodies (ANA), antineutrophil cytoplasmic antibodies (C-ANCA and P-ANCA), rheumatoid factor and retroviral antibody were negative. Pulmonary function tests showed moderate restriction (forced vital capacity decreased to 56% of predicted value). Chest X-ray was showing large pneumothorax on the right side with acino-nodular opacities in Left lung fields. However, chest computed tomography (CT) showed diffuse ground-glass opacities all over the Left lung fields which are predominantly subpleural with patchy areas of consolidation and pneumothorax on the right side with pneumomediastinum, without nodules or honeycomb pattern (**Figure 2**). The patient was in severe respiratory distress. We performed video-assisted thoracic surgery (VATS) guided lung biopsy and chest tube was inserted followed by talc pleurodesis at a later date. Histopathology revealed diffuse infiltration of alveolar spaces with macrophages (**Figure 3**), these macrophages were filled with foamy cytoplasm (**Figure 4**). The patient was diagnosed with DIP and prednisolone (60 mg/day) was administered resulting in improvement of symptoms. The chest tube was removed and patient was discharged from the hospital with a maintenance dose of prednisolone (30 mg/day), was advised to avoid smoking (active/passive) and to follow up in the outpatient clinic.



**Figure 1.** HRCT Showing ground glass opacities predominantly subpleural in lower lobes (elsewhere)



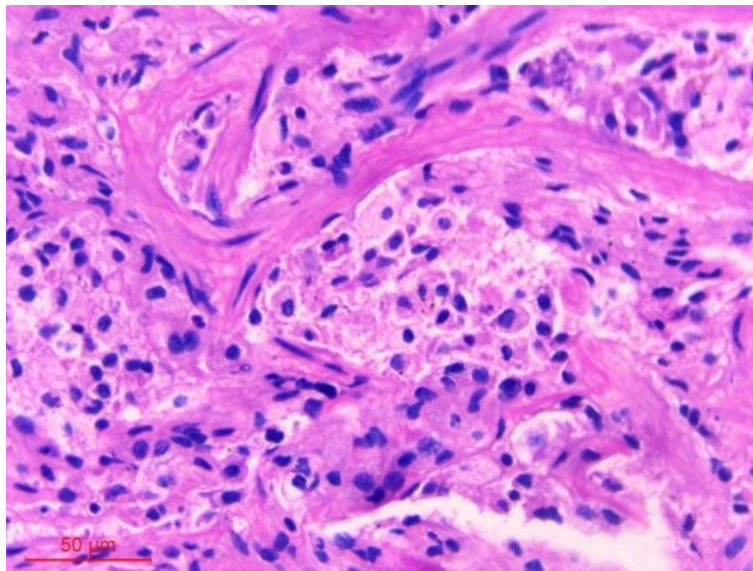
**Figure 2.** CT scan showing right side pneumothorax and pneumomediastinum (at presentation)



**Figure 3.** HPE Low power view of DIP showing diffuse involvement of alveoli with infiltration of macrophages

## Discussion

DIP is a rare form of idiopathic interstitial pneumonia characterized by a large number of pigmented macrophages in the alveoli with interstitial fibrosis and/or inflammation first described by Liebow et al in 1965 (1). The lesions are characterized by massive proliferation and desquamation of large alveolar cells, by slight thickening of the walls of distal air spaces, by the absence of necrosis and by minimal loss of tissue (1).



**Figure 4.** HPE Higher power view showing alveoli filled with macrophages with foamy cytoplasm.

The predominant findings on high resolution CT scans in patients with DIP are ground glass opacities which usually affect the lower lobe and subpleural regions. These together with the few cystic changes seen in patients with DIP, have been found to be the features which distinguish DIP from UIP and are thus a basis for distinguishing between the two conditions in most cases (1,2). The relatively good preservation of morphological structures of the underlying parenchyma in such areas is commonly seen on the HRCT scan of patients with DIP.

Histopathological examination in DIP is characterized by the presence of alveoli studded with large number of macrophages. The macrophages are usually evenly distributed, often contain light brown pigment and are also called smoker's macrophages. Finely granular iron may be present in the cytoplasm. Earlier the macrophages were thought to be desquamated type II epithelial cells hence the misnomer "desquamating interstitial pneumonia" (4). The lung architecture is maintained so that the alveoli are readily recognized. The alveolar walls display a minimal inflammation with mild infiltration of lymphocytes, plasma cells, and occasional eosinophils. There may be metaplasia of type II the alveolar lining cells. Interstitial fibrosis is only mild when present.



**Figure 5:** Chest X-ray after 3 months of steroid treatment

Most patients with DIP show a good response to corticosteroid therapy with smoking cessation and has good prognosis (2). Even spontaneous remission has been described in some cases. The literature contains only few cases that were refractory to corticosteroid therapy, showed near fatal tension pneumothorax despite steroid therapy (3). Our patient had pneumothorax with mediastinal emphysema, presented as air leak syndrome and was managed with VATS lung biopsy and corticosteroid therapy. Pneumothorax is a possible complication of DIP (3). Kim et al. (5) reported the initial improvement of DIP after initiation of corticosteroid therapy and pneumatocele formation during the course of corticosteroid tapering. For patients with DIP, a standard guideline regarding the dose and duration of corticosteroid has not been suggested yet. Therefore, corticosteroid therapy needs to be adjusted according to individual circumstances and the severity of disease.

Patient is on regular follow up since 3 months, he is asymptomatic with marked radiological clearing (**Figure 5**) and improvement in pulmonary function.

### References

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