A rare case of secondary pulmonary alveolar proteinosis with adenocarcinoma of lung with retroviral disease

Sir,

Pulmonary alveolar proteinosis (PAP) is a rare disorder characterized by alveolar accumulation of abnormal surfactant. We report, so far, the first case of a patient with retroviral disease presenting with secondary PAP and adenocarcinoma of the lung together from India.

A 35-year-old man, diagnosed with human immunodeficiency virus infection two years ago, on antiretroviral therapy, presented with complaints of dyspnea, cough, fever, and weight loss. He was treated with antituberculous therapy since six months on presumptive diagnosis of tuberculosis based on clinical symptoms and radiology, chest X-ray [Figure 1], and computed tomography (CT) scan findings.

Sputum reports for all infections were negative. Bronchoalveolar lavage (BAL) was performed. The BAL fluid showed alveolar macrophages 27%, lymphocytes 32%, neutrophils 41%, and sediment on standing periodic acid stain positive and diastase resistant, suggestive of PAP, negative for infections including tuberculosis, *Pneumocystis carinii* pneumonia (PCP), and malignancy. He refused a therapeutic lavage and transbronchial biopsy.

On follow-up, about 2 months later, he presented with increased symptoms. A repeat CT scan revealed an increase in ground glass opacities and increased consolidation in the right lower lobe [Figure 2]. A transbronchial biopsy and washings were taken. Bronchial washings were negative for infective etiology such as tuberculosis, PCP, bacteria, fungi, and atypical mycobacteria. Histopathological examination was suggestive of adenocarcinoma of the lung staining focally positive for thyroid transcription factor-1. The patient was referred to a higher oncology center for further management.

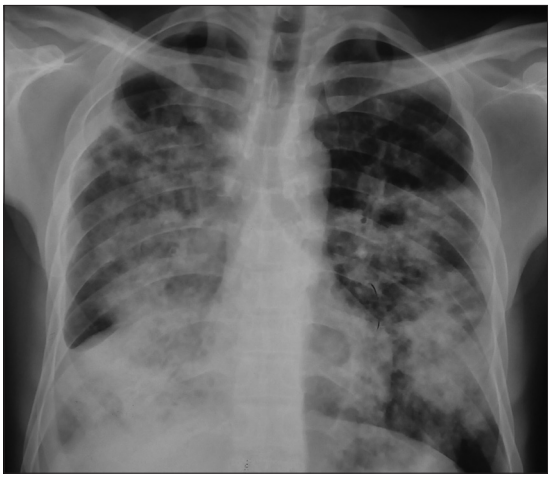

Figure 1: Chest X-ray showing bilateral alveolar opacities in all lung fields and right lower zone consolidation

Secondary PAP has been reported in association with systemic disorders associated with immunodeficiency, including acquired immunodeficiency syndromes. In patients with acquired immunodeficiency syndrome (AIDS), PAP may be result of the primary pathology[1] or due to super added infections like PCP.[2]

The altered immune function in AIDS patients leads to an altered ability of the macrophages to clear out accumulated lipoid material within alveoli and an increased susceptibility to opportunistic infections. A systematic review of Indian cases of PAP was done by Hadda *et al*.[3] in 2016. As per this review, out of 30 reported cases, 24 had primary PAP. Remaining six patients were labeled as secondary PAP related to cyclosporine or mycophenolate, cotton dust exposure, sandstone exposure, glass cutting, and fiber exposure or nocardia. Concomitant infections were seen in four patients, *Mycobacterium tuberculosis* in two and *P. carinii* in two patients.

Similarly, Khan *et al*.[4] also published a case series from another Indian tertiary care hospital. This study had five patients with idiopathic PAP and one patient with secondary PAP due to Nocardia.

In neither of these two studies was a case with AIDS, PAP, and pulmonary cancer reported.

PAP has been described in association with cancers, both pulmonary (small cell cancer, non-small cell cancer, squamous cell carcinoma, and adenocarcinoma) and extra-pulmonary.

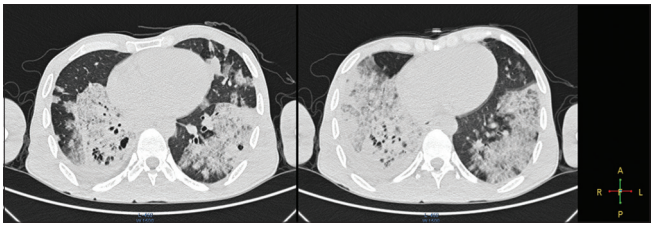

Figure 2: Computed tomography scan images showing consolidation and ground glass appearance bilaterally

The lung cancer may occur prior to[5] coincidentally with[5] or after many years of diagnosis of PAP.[6]

In both PAP and adenocarcinoma, the alveoli are filled with a low-density material, the alveolar septa thicken but the underlying parenchymal architecture remains normal.

Solid lung cancers causing PAP have not been extensively studied. As the patient was otherwise tolerating his antiretroviral therapy over two years with a normal CD4 count, considering the slow progression of the disease, the PAP was likely secondary to malignancy.

The importance of diagnosing the etiology for the management of a disease cannot be overemphasized especially in a rare disorder like PAP. However, due to the complex pathophysiology of PAP, diagnostic modalities to differentiate the underlying cause in presence of two potential causes of secondary PAP are lacking. The presentation of retroviral disease with adenocarcinoma of lung with PAP is extremely rare and to the best of our knowledge, this is the first reported case in India.

Acknowledgment

We would like to thank doctors and nursing and supporting staff of Intensive Respiratory Care Unit, Department of Chest Medicine and EPRC.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Aditi Dushyant Punwani, Sonal Prabhakar Karpe, Amita Umesh Athavale, Prabodh D Garg, Mahesh R Jansari

Department of Chest Medicine, EPRC, KEM Hospital, Mumbai, Maharashtra, India
E-mail: sonalkarpe@yahoo.co.in

Submitted: 12-Mar-2020 **Accepted:** 02-Apr-2020
Published: 30-Oct-2020

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Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/lungindia.lungindia_142_20

How to cite this article: Punwani AD, Karpe SP, Athavale AU, Garg PD, Jansari MR. A rare case of secondary pulmonary alveolar proteinosis with adenocarcinoma of lung with retroviral disease. Lung India 2020;37:550-1.