A Rare Case of Pulmonary Alveolar Proteinosis in a Pregnant Lady

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Abstract

Pulmonary alveolar proteinosis is a relatively rare syndrome of pulmonary surfactant clearance dysfunction that could present like asthma. A case of a middle-aged pregnant lady presented with asthma-like symptoms which was negative for autoimmune screening. Thoracic imaging showed ground-glass opacity with interstitial thickening. Bronchopulmonary lavage fluid analysis showed predominantly eosinophilic material within alveolar space. Lung biopsy revealed positive PAS stain for eosinophilic material. Its presentation in pregnancy could pose challenge to delivery. The associated maternal infection risk could compromise fetal survival.

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Case

This is a case of a 38-year-old lady, gravida 3 para 2 at 35 weeks of gestation, presented to our clinic with a three-month history of shortness of breath and dry cough. She is non-smoker and had past medical history of mild intermittent asthma. Further history shows that there were no symptom variability during daytime and night time. No associated constitutional symptoms. No symptoms suggestive of connective tissue disease including mucosal ulcers, arthritis, Raynaud’s or photosensitive rashes. Apart from that, no risk factors for pulmonary embolism were identified and she was not any oral contraception pills before.

On physical examination, patient was afebrile. She was not in respiratory failure and both lungs were clear. Upon investigation, the connective tissue screening was negative. Chest radiograph (CXR) showed bilateral hilar ill-defined opacities. Computed tomography (CT) of the thorax revealed a crazy paving pattern of ground glass opacity especially over the left lung, with superimposed interlobular septal thickening (Figure 1). The patient was treated initially with one week of antibiotic course for presumed community acquired pneumonia followed by trial of diuretic for presumed mild pulmonary edema due to unresponsiveness to the antibiotic therapy. Patient delivered uneventfully via elective caesarean (no capital letter) section one week after our visit.

The patient then underwent bronchopulmonary lavage three months post-partum. The lavage fluid appeared milky and turbid. Fluid analysis showed that the fluid was predominantly eosinophilic granular material within the alveolar spaces with amorphous non-foamy hyaline globules were identified (Figure 2). Culture and sensitivity showed no growth identified. Microscopic analysis of the transbronchial lung biopsy demonstrated diffuse eosinophilic granular material within the alveolar spaces which stained with Periodic Acid Schiff staining (Figure 3). The findings are in keeping with pulmonary alveolar proteinosis (PAP). Following the extensive bronchopulmonary lavage involving both lungs, she had marked improvement in lung functions.

Discussion

Pulmonary alveolar proteinosis is a rare pulmonary syndrome. The commonest age group for the onset of this syndrome is between 30 and 40 years with male predominance [1].

The annual incidence of pulmonary alveolar proteinosis was reported around 0.36 to 0.49 cases per million populations, with prevalence of 3.7 to 6.2 cases per million populations [2]. However, there are limited studies about the incidence and prevalence of such disease entity among our Asian community.

The usual presenting symptom appears to be progressive breathlessness, with chest radiographic evidence of bilateral alveolar infiltrates. The course of the disease varies. Some patients experience spontaneous remission; others develop progressive respiratory insufficiency. Milky lung lavage with PAS-positivity towards the lipoproteinaceous material clinches the diagnosis [3]. Periodic whole lung lavage is the most effective modality for such condition [4]. The whole lung lavage requires general anaesthesia and complete lung isolation with a
double-lumen endotracheal tube. Proteinaceous materials were washed out from one lung with saline while the other lung is being ventilated.\textsuperscript{[4]}

Relapses are commonly seen in this syndrome and thus long-term continuous follow-up is essential, particularly with the respiratory team.\textsuperscript{[5]} Prompt intervention, including repeat lung lavage, can be instituted in the case of relapse. Environmental triggers, if any, should be identified so as to avoid such triggers, for instance, insecticides and dusts.

**Conclusion**

This case highlights a rare case of pulmonary alveolar proteinosis presenting in pregnancy. However, respiratory function was not compromised and patient was able to undergo the lavage during postpartum period. Another point to emphasize here is clinicians should always first to consider pneumonia and other commoner respiratory diseases while approaching a pregnant lady with chronic dyspnoea and cough before thinking about such rare entity of pulmonary alveolar proteinosis. In spite of common cases of pneumonia and other commoner respiratory disease in pregnancy, pulmonary alveolar proteinosis is a rare differential diagnosis.

**Conflict of Interest Statement**

None

**References**