PULMONARY ALVEOLAR PROTEINOSIS TREATED SUCCESSFULLY WITH WHOLE LUNG LAVAGE: A RARE CASE WITH RARE PROCEDURE

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ABSTRACT

Pulmonary Alveolar Proteinosis (PAP) is characterized by accumulation of surfactant in alveoli and terminal airways resulting in hypoxemic respiratory failure. A report describes a case of 42 years old male patient with PAP, presented with bilateral chest pain and dyspnea on rest since last four months. General examination, revealed that the patient had hypoxia but he was hemodynamically stable. Chest examination showed bilateral crepitations. Chest radiograph was suggestive of bilateral consolidation and cavitatory changes in right upper and lower zones. HRCT thorax was suggestive of diffuse ground glass opacities with interstitial septal thickening (crazy paving pattern). Bronchoalveolar Lavage was PAS positive confirming diagnosis. Whole lung lavage procedure was done for the management of PAP. Patient was asymptomatic after 3 months of the procedure.

KEYWORDS

Pulmonary Alveolar Proteinosis (PAP), Whole Lung Lavage, Crazy Paving Pattern

INTRODUCTION

Pulmonary Alveolar Proteinosis (PAP) is characterized by accumulation of surfactant in alveoli and terminal airways resulting in hypoxemic respiratory failure. PAP can be grouped into three categories as primary, secondary and congenital. Whole lung lavage remains the most widely used therapy for PAP. Herein, we present a case of secondary PAP in which we performed whole lung lavage procedure successfully with improvement of patient clinically as well as radiologically.

CASE REPORT

A 42 years old male patient working as a well digger, presented to the department of pulmonary medicine, B.J. Medical College, Ahmedabad, with breathlessness at rest and bilateral chest pain since 2 days. He had persistant bilateral chest pain and dyspnea on exertion since last 4 months with no complaints of cough and fever. He had no past history of hypertension, diabetes, tuberculosis and he was not having any addiction. He had no any gas or toxic substance exposure history. He didn't had similar illness in his family At the time of admission, his temperature was 36.9°C. Blood Pressure of 108/72 mmHg, Pulse rate 122/ min, Respiratory Rate 22 / min, SpO2, 76 on room air. There was no cyanosis, no clubbing and no distension of jugular vein.

Trachea was centrally placed and there was increase in respiratory movements. On auscultation, bilateral crepitations were present more on right side.

ABGA showed PaO2 56, PCO2 36, pH 7.36, SPO2 82.4%.

LFT, RFT, CBC, PT, RBS were within normal limits. Sputum for AFB was negative Chest R radiography PA view.

HRCT thorax showed diffuse ground glass densities in both lungs with interstitial septal thickening seen through it (crazy paving pattern). Consolidation was seen in bilateral lower lobes with 34x26x36 mm cavity in right lower lobe.
Bronchoscopy was done to confirm the diagnosis. Bronchoalveolar Lavage was done to confirm the diagnosis. BAL fluid showed PAS stain positive.

We planned whole lung lavage procedure with discussion with anesthetists and physiotherapists at our institute. We did the procedure in our Surgery OT under General Anesthesia, double lumen ET tube placed and no leaks were confirmed by Bronchoscope. First, right lung was selected and for this, patient was kept in lateral position with right side upwards. The left lung was ventilated during the procedure. One liter warm saline infused at the rate of 120 ml per minute and then, 4-5 minutes of chest percussion and oscillation was performed by physiotherapists. Then outlet clamp was removed allowing the fluid to freely gravitate in collection bottle. This cycle was performed 14 times and in each cycle, fluid showed decreased turbidity and increased transparency.

The fluid collected in each cycle, serially is shown as below

Patient was vitally stable and conscious, oriented to time, place and person on the very next day and extubated and put on simple oxygen mask. On the next day, patient's SPO2 was 94% at room air.

Patient was discharged on 3rd day of procedure and was followed up after one month.

We admitted the patient on the next follow up and did the Whole Lung Lavage procedure on the Left Lung. Then patient was maintaining SPO2 98% at room air. Patient was advised to avoid the aggravating factors of developing PAP on discharge. Follow up HRCT thorax was done in the next visit which showed significant improvement as compared to the pre procedure HRCT.

DISCUSSION
Pulmonary Alveolar Proteinosis (PAP) is characterized by accumulation of surfactant in alveoli and terminal airways resulting in hypoxemic respiratory failure. PAP can be grouped into three categories as primary, secondary and congenital. Primary PAP is caused by impairment of GM-CSF dependent surfactant clearance by alveolar macrophage and it is most common type and accounts for 90% of all cases. Secondary Pap is a consequence of comorbid condition that impairs surfactant clearance by alveolar macrophage. It is commonly associated with hematological disorders, primary myelodysplasia, infectious diseases, autoimmune diseases, immunosuppressants after organ transplantation, heavy inhalational exposure to inorganic dust e.g. silica, aluminium, titanium, and HIV infection.

PAP patients commonly present as progressive dyspnea of insidious onset in age group of 20 to 50 years, cough, fatigue, fever and sputum production.

On physical examination, crepitations especially in dependant areas and cyanosis are occasionally present. Digital clubbing is typically absent.

Chest radiograph usually reveals bilateral symmetrical alveolar opacities located centrally in mid and lower lung zones often with a peribulbar predominance resembling batwing appearance.

HRCT shows geographical pattern of ground glass opacification with superimposed interlobular septal and intralobular thickening commonly referred as crazy paving pattern that is characteristic of PAP. BAL fluid is opaque and has milky/waxy appearance and is stained positive with PAS Whole lung lavage remains the most widely used therapy for PAP.

Augmentation with recombinant human GM-CSF, in subcutaneous and inhaled administration forms, have demonstrated therapeutic efficacy with latter appearing more efficaceous.

CONCLUSION
Pulmonary Alveolar Proteinosis is a disease of irregularity of alveolar macrophage which leads to accumulation of surfactant protein in alveolus which leads to hypoxemia which can be successfully treated with whole lung lavage.

REFERENCES:
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