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Clinical Image

Pulmonary Alveolar Proteinosis

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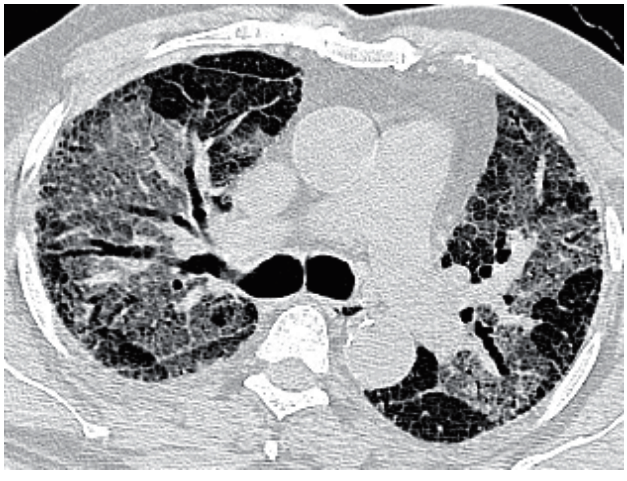
In March, 2012, a 68-year-old gentleman presented to the Intensive Care Unit (ICU) with respiratory distress, fever and chest pain for three days. As his oxygen saturation could not be maintained, even with non-invasive ventilation, he was intubated and put into mechanical ventilatory support. Initial chest x-ray A-P view, done in the ICU showed "bat wing" appearance of pulmonary edema. But cardiac evaluation, including an echocardiogram, excluded any cardiac involvement. A high resolution computed tomography (HRCT) of lung revealed "diffuse bilateral ground glass opacities with thickened interlobular septae, giving rise to typical crazy paving appearance, in bilateral upper lobes, lateral segment of right middle lobe and involves areas of lower lobes in geographical pattern" [Figure 1 & 2], suggestive of pulmonary alveolar proteinosis. Periodic acid-Schiff (PAS) staining of the broncho-alveolar lavage (BAL) were also positive. Repeated lobar lavages were done by fibre-optic bronchoscope (FOB), with temporary clinical improvement. As our hospital does not posses the facility for whole lung lavage, and other advanced therapeutic options, the patient was referred abroad for further management.

Pulmonary Alveolar Proteinosis (PAP) is a diffuse pulmonary disease where lipoproteinaceous material, primarily surfactant and surfactant apoproteins, are accumulated in distal airway and alveoli, resulting in impaired gas transfer.[1,2] It is a rare disorder with estimated prevalence of 0.37 per 100,000.[1] The clinical course ranges from spontaneous resolution to respiratory failure. It has three distinct forms, congenital (mutation in genes encoding for surfactant proteins and granulocyte-macrophage colony-stimulating-factors), secondary (to immunosuppression, hematological cancers or toxic inhalations) and acquired or idiopathic (autoimmune disease, associated with GM-CSF autoantibodies)[1,2] Most patients present with progressive exertional dyspnoea and cough. Fever, chest pain, or hemoptysis are less common presentations. Inspiratory crackles, cyanosis and digital clubbing are usual physical findings. Routine blood counts, chemical analysis, and urinlysis are usually normal. Chest radiograph usually reveals bilateral airspace disease with an ill-defined nodular or confluent pattern, often with perihilar predominance or "bat wing" appearance of pulmonary edema but without other radiographic signs of left-sided heart failure. HRCT shows patchy, ground-glass opacifications with superimposed interlobular septal and intralobular thickening, a pattern commonly referred to as "crazy paving".[1] Lipoproteinaceous material (phospholipid surfactant and surfactant apoproteins) in the BAL are PAS-positive.[2]



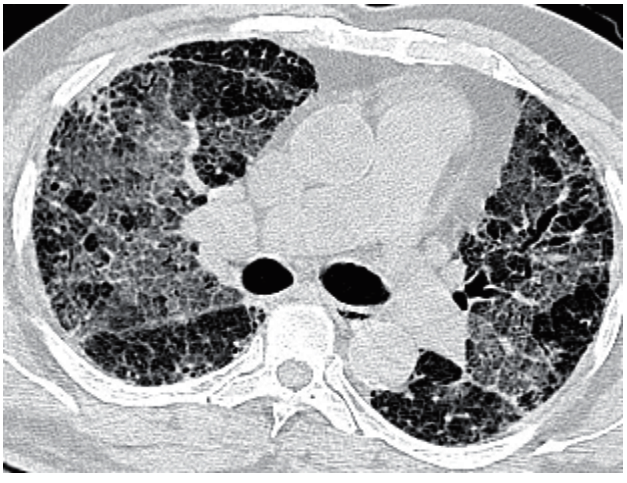


Figure 1 & 2: *HRCT of lung revealed diffuse bilateral ground glass opacities with thickened interlobular septae giving rise to typical "crazy paving" appearance.*

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Treatment of PAP depends on the underlying cause. Treatment for the congenital form is supportive, whereas secondary form depends on the treatment of the underlying cause.[1] Acquired PAP has successfully been treated with whole-lung lavage, and still remains the standard of care.[1,2] If whole-lung lavage under general anesthesia is unavailable, or contraindicated, multiple segmental or lobar lavage by FOB is considered safer. Other potential therapeutic approaches are subcutaneous or inhaled GM-CSF, rituximab etc.[2] Successful treatment with lung transplantation has also been reported.[1]

References:

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