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Case Report:

Pulmonary manifestation of primary biliary cirrhosis: A Case Report

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Abstract:

Primary biliary cirrhosis (PBC) is a chronic, progressive autoimmune liver disease characterized by the destruction of small intrahepatic bile ducts, resulting in impaired bile flow and the accumulation of toxic bile acids. Although the primary pathology of PBC primarily affects the liver, it is increasingly recognized that this condition can also have extrapulmonary manifestations, including pulmonary involvement. A 45 year old male patient was admitted with complaints of exertional breathlessness, and occasional cough ,fatigue since 25 years. The symptoms had increased over the last 2 days. There was history of hospitalisation 1 year back wherein patient was diagnosed with liver abscess which was drained. On examination, the patient was tachypneic (respiratory rate :30 /min), pulse rate was 120/min, BP was 110/70 of mm of Hg, and SpO_2 was 95% on 2L O2/min. In conclusion, this case report emphasizes the importance of considering pulmonary manifestations in patients with primary biliary cirrhosis (PBC) and highlights the association between PBC and progressive pulmonary fibrosis. Early recognition and management of these complications are crucial for optimizing patient outcomes.

Keywords: Primary biliary cirrhosis, autoimmune liver disease, hepatomegaly

Introduction:

Primary biliary cirrhosis (PBC) is a chronic, progressive autoimmune liver disease characterized by the destruction of small intrahepatic bile ducts, resulting in impaired bile flow and the accumulation of toxic bile acids. Although the primary pathology of PBC primarily affects the liver, it is increasingly recognized that this condition can also have extrapulmonary manifestations, including pulmonary involvement.

Pulmonary manifestations in PBC are relatively rare but can significantly impact a patient's quality of life and overall prognosis. These manifestations can range from mild respiratory symptoms to severe

complications such as pulmonary hypertension and fibrosis. The exact mechanisms underlying the development of pulmonary involvement in PBC are not yet fully understood, but it is believed to be multifactorial, involving immune dysregulation, chronic inflammation, and vascular abnormalities. Clinically, patients with PBC may present with respiratory symptoms such as dyspnea, cough, and fatigue. These symptoms are often non-specific and can be mistakenly attributed to other comorbidities or the effects of advanced liver disease. Therefore, the diagnosis of pulmonary involvement in PBC requires a high index of suspicion, thorough evaluation, and exclusion of other respiratory diseases.

Case Report:

A 45 year old male patient was admitted with complaints of exertional breathlessness, and occasional cough ,fatigue since 25 years. The symptoms had increased over the last 2 days. There was history of hospitalisation 1 year back wherein patient was diagnosed with liver abscess which was drained. On examination, the patient was tachypneic (respiratory rate :30 /min), pulse rate was 120/min, BP was 110/70 of mm of Hg, and SpO_2 was 95% on 2L O2/min. There was grade 4 clubbing and respiratory system examination revealed bilateral basal crepitations. Serum ANA was positive, and ANA profile revealed positive antimiochodrial antibodies (AMA M2 +). RA factor was positive, anti-CCP test was negative. Total IgE was 114.26 iu. Serum Lipid profile was normal (s.cholesterol 122mg%, triglycerides was 50 mg%),liver function test was normal, USG abdomen was suggestive of liver abscess in right lobe of liver. CECT Scan thorax (Photograph 1) showed cystic bronchiectasis in bilateral lower lobes, along with bilateral upper lobe fibrosis. Sputum culture suggestive of growth of pseudomonas. Patient was diagnosed as a case of PBC with PBC related progressive pulmonary fibrosis with secondary respiratory infection.

Discussion:

Pulmonary manifestations in PBC are relatively rare but can significantly impact a patient's quality of life and overall prognosis. These manifestations can range from mild respiratory symptoms to severe complications such as pulmonary hypertension and fibrosis. The exact mechanisms underlying the development of pulmonary involvement in PBC are not yet fully understood, but it is believed to be multifactorial, involving immune dysregulation, chronic inflammation, and vascular abnormalities. Clinically, patients with PBC may present with respiratory symptoms such as dyspnea, cough, and fatigue. These symptoms are often non-specific and can be mistakenly attributed to other comorbidities or the effects of advanced liver disease. Therefore, the diagnosis of pulmonary involvement in PBC requires a high index of suspicion, thorough evaluation, and exclusion of other respiratory diseases.

This case report aims to present a detailed clinical account of a patient with primary biliary cirrhosis who developed pulmonary manifestations. We describe the patient's clinical presentation, diagnostic workup, and treatment course, highlighting the challenges encountered in establishing the diagnosis and managing the pulmonary complications. By sharing this case, we hope to increase awareness among healthcare professionals about the potential pulmonary manifestations of PBC and promote early recognition and appropriate management of these conditions.

This case report highlights the pulmonary manifestations of primary biliary cirrhosis (PBC) in a 45-year-old male patient. The patient presented with exertional breathlessness, occasional cough, and fatigue, which had worsened over the past 2 days. The history of a previous hospitalization for a liver abscess raises the possibility of an underlying liver disease, which led to the suspicion of PBC in this case.

PBC is a chronic autoimmune liver disease characterized by progressive destruction of small intrahepatic bile ducts. While the primary pathology of PBC primarily affects the liver, it is increasingly recognized that this condition can have extrapulmonary manifestations, including pulmonary involvement. The presence of respiratory symptoms and the findings on examination, such as tachypnea, bilateral basal crepitations, and grade 4 clubbing, raise suspicion for pulmonary involvement in this patient.

The diagnosis of PBC in this case was supported by the presence of positive serum antinuclear antibodies (ANA) and antimiochondrial antibodies (AMA M2+). ANA positivity is a common finding in autoimmune diseases, while AMA M2 is highly specific for PBC. Additionally, the absence of other autoimmune markers, such as rheumatoid factor (RF) and anti-CCP antibodies, helps to exclude other rheumatologic conditions.

The presence of cystic bronchiectasis in the bilateral lower lobes, along with bilateral upper lobe fibrosis, on the CT scan of the thorax is consistent with PBC-related progressive pulmonary fibrosis. The development of fibrotic changes in the lungs is thought to be due to chronic inflammation and immune dysregulation seen in PBC.

Conclusion:

In conclusion, this case report emphasizes the importance of considering pulmonary manifestations in patients with primary biliary cirrhosis (PBC) and highlights the association between PBC and progressive pulmonary fibrosis. Early recognition and management of these complications are crucial for optimizing patient outcomes.

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