

Background

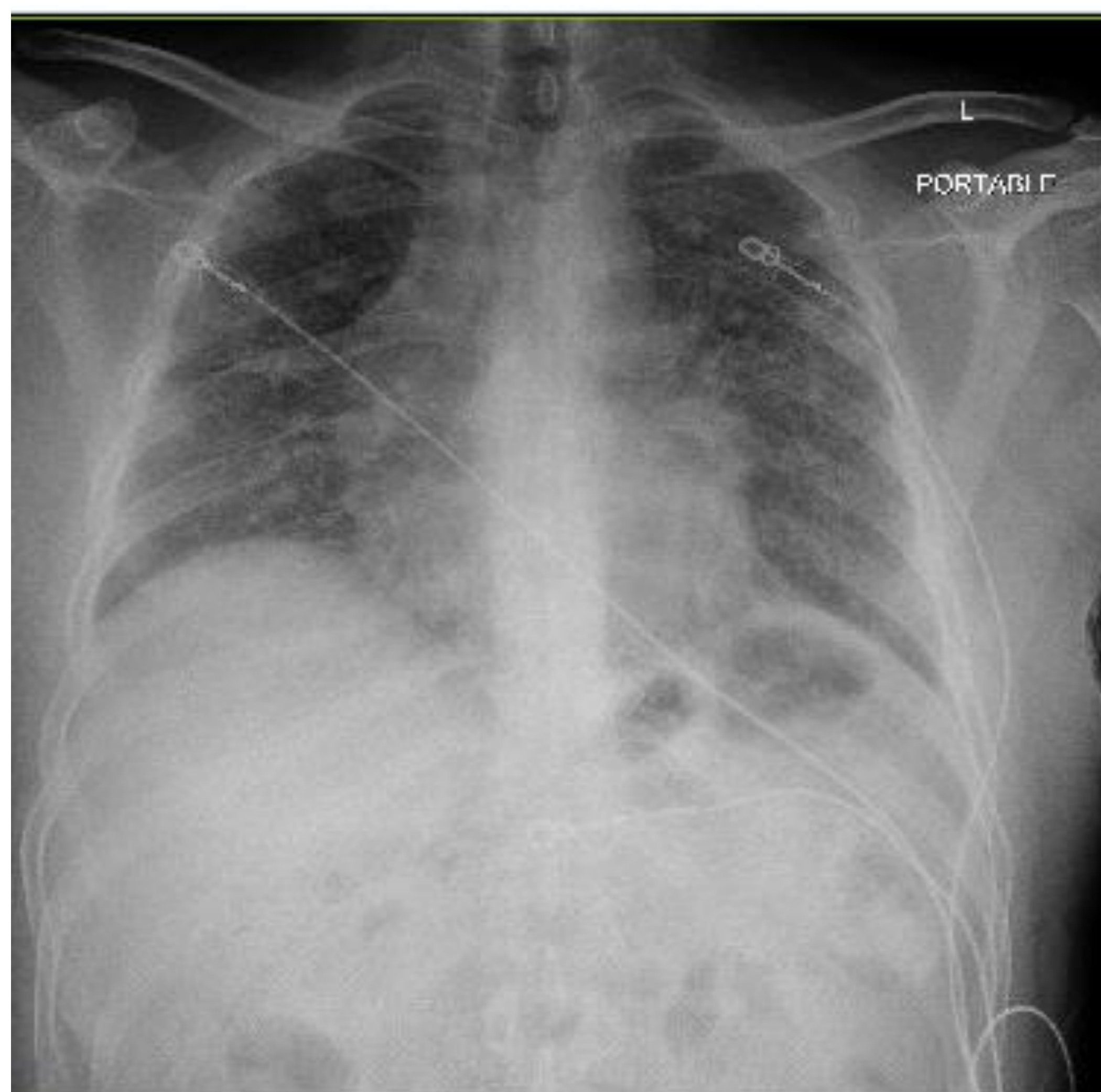
Pulmonary alveolar proteinosis, also known as PAP, is a rare lung condition which leads to the buildup of surfactant in the alveoli, preventing gas exchange resulting in hypoxemia. PAP is classified as either autoimmune, congenital, or secondary to environmental exposure. Diagnosis of PAP involves testing for anti-GM-CSF antibodies, chest x-ray, CT scan, and a lung biopsy. Once diagnosis is established, many require whole lung lavage (WLL) with about 20L of saline in order to clear out copious amounts of protein from the lungs. This necessitates lung isolation techniques and possible V-V ECMO support. These patients exhibit significant hypoxemia requiring high flow oxygen therapy and if refractory to standard treatments, will require lung transplant.

Case Presentation

43 year old male with significant environmental exposure to silica; he was a granite and marble cutter for over 10 years. He developed significant shortness of breath requiring 5L of home oxygen. He presented to South Shore University Hospital with increasing shortness of breath and hypoxemia. He underwent right VATS with lung biopsy and BAL, with ECMO standby. Pathology came back positive for acute on subacute silicosis and extensive secondary pulmonary alveolar proteinosis. He continued to deteriorate in the hospital with respiratory failure and hypoxemia with multiple reintubations. He was transferred to Northwell LIJ where he underwent whole lung lavage with 20L of normal saline and with lung isolation and ECMO stand by. This patient exemplified secondary PAP due to silica exposure and likely also had a component of autoimmune PAP.



CT and CXR on Admission



Discussion

There are numerous anesthetic challenges during management of patients with PAP. One-lung ventilation in a patient with already compromised pulmonary function can lead to hypoxemia. During whole lung lavage, the lavaged lung is often dependent, which also contributes to hypoxemia. During drainage, blood flow increases back to the nonventilated lung increasing shunting. In some instances, ECMO is necessary for adequate oxygenation. Hemodynamic instability also occurs when large amounts of lavage fluid cause a mediastinal shift, affecting filling of the heart and cardiac output. Large lavage fluid volume can also increase pulmonary vascular resistance. Other issues during WLL include hypothermia, hydro- or pneumothorax, and loss of lung isolation with leakage of lavage fluid into the ventilated lung.

References

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