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Exploration To Delineate The Scannographic Aspects Of Alveolar Proteinosis: A Rare Case Report

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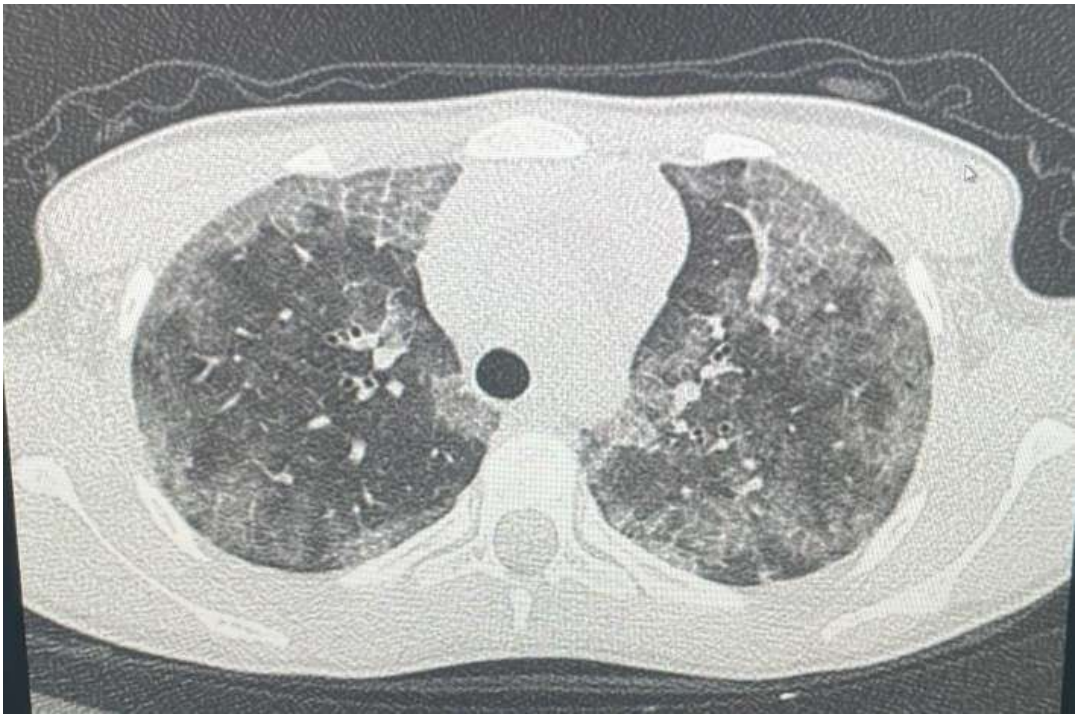
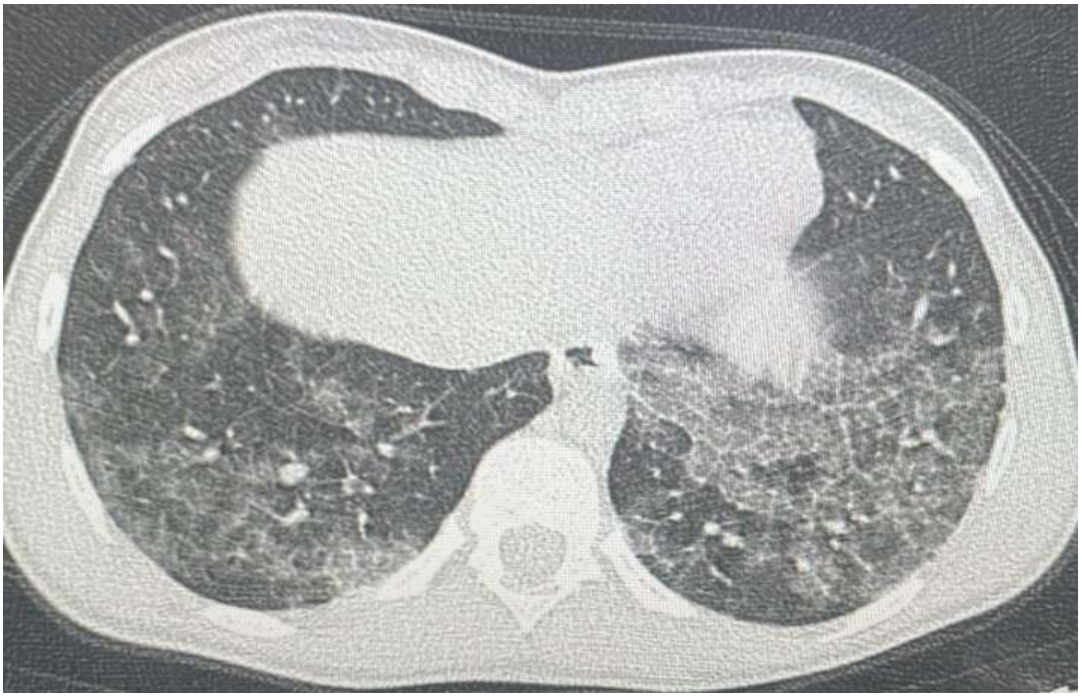
Alveolar Proteinosis (AP) is an uncommon disease characterized by the accumulation of surfactant components within the alveoli, hindering gas exchange. AP can be categorized into three groups:
1. Autoimmune AP, which is the most prevalent form and is distinguished by the presence of serum anti-GM-CSF antibodies.
2. Secondary AP, associated with an underlying condition, commonly hematologic, as well as toxic inhalation or infections.
3. Genetic AP.

In light of a rare case of alveolar proteinosis observed in a patient admitted to the radiology department of Cheikh Zaid University Hospital in Rabat, the authors aim to discuss the role of computed tomography (CT) and the scannographic aspects of this pathology.

I. Case Presentation:

A 14-year-old patient with diagnosed alveolar proteinosis under bronchoalveolar lavage treatment presented with dyspnea, thoracic pain, and asthenia in January 2021. A thoracic CT scan without contrast enhancement revealed the following findings:
- Diffuse pulmonary infiltration (alveolo-interstitial syndrome) characterized by a ground-glass and confluent appearance predominantly subpleurally in the lower lobe, suggestive of probable alveolar proteinosis.
- No mediastinal adenopathy.
- No pleuropericardial effusion.
- No parenchymal abnormalities.





II. Discussion:

Alveolar proteinosis is a rare disease with varying prevalence rates among different countries, ranging from 4 to 40 cases per million inhabitants, with an annual incidence of 0.2 per million inhabitants. Autoimmune AP shows a male predominance with a sex ratio exceeding 2. Nowadays, surgical lung biopsy is rarely required for AP diagnosis.

Indeed, thoracic CT scan and bronchoalveolar lavage (BAL) are generally sufficient to establish a positive diagnosis of AP, considering a compatible clinical presentation and after excluding other causes of diffuse infiltrative lung disease (DILD). The autoimmune nature of AP can be confirmed by the presence of positive serum autoantibodies against GM-CSF.

Radiologically, alveolar proteinosis appears as bilateral and often asymmetric consolidations on standard chest X-rays, with more pronounced abnormalities in the perihilar regions, resembling "butterfly wings" or "bat wings," without other suggestive signs of heart failure (cardiomegaly, Kerley B lines, pleural effusion). Pleural effusion is rare except in cases of complications. The scarcity of functional and clinical signs often contrasts with the profusion of radiological lesions.

Thoracic CT scan is essential for a definitive diagnosis of AP. It typically reveals ground-glass opacities distributed in a geographic pattern (involving healthy and diseased lobules), sometimes accompanied by alveolar parenchymal consolidations. Thickening of inter- and intralobular septal lines, creating a "crazy paving" appearance, is a classic feature of AP.

Although the crazy paving pattern is not pathognomonic of AP, it should prompt further investigation through BAL, with specific tests targeting AP. Differential diagnoses for alveolar proteinosis include infections (such as Pneumocystis pneumonia, tuberculosis), neoplastic conditions (lipid-laden adenocarcinoma), drug-induced pneumonias (amiodarone, bleomycin, gold salts), and other idiopathic DILD (sarcoidosis, nonspecific interstitial pneumonia).

BAL is the second essential examination for AP diagnosis. Bronchoscopic examination appears normal, while BAL fluid is classically described as "milky." It contains a substantial amount of acellular eosinophilic proteinaceous granular material that is periodic acid-Schiff (PAS) positive. Additionally, spumous macrophages with PAS-positive intracellular inclusions can be found. Electron microscopy of BAL from an AP patient reveals concentrically lamellated phospholipid structures called "lamellar bodies."

Treatment of AP primarily relies on therapeutic whole lung lavage. In refractory cases, GM-CSF and rituximab may be considered.

III. Conclusion:

Three types of AP can be distinguished: autoimmune, secondary to an underlying disease, and genetic. CT scanning serves as the gold standard for AP evaluation, displaying a crazy paving pattern, while BAL shows a milky appearance with lipoproteinaceous material. Therapeutic whole lung lavage is the cornerstone of treatment, with GM-CSF and rituximab reserved for refractory forms.