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Complete Remission of Autoimmune Pulmonary Alveolar Proteinosis Associated with Mycoplasma Infection: A Rare Case Report

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Case report

A 12 -year-old female presented to the emergency room difficulty in breathing while speaking along with hypoxia of 89% on room air. Upon further questioning, patient has been experiencing exercise intolerance, fatigability and diminished school performance for the last 4 months. She was previously healthy and she has had normal growth with weight and height the 76th and 97th percentiles. She had no allergies and did not use medication including bronchodilators. There was no family history of lung disease.

She was seen in at the emergency room a month prior with complain of dry cough and shortness of breath, at that time; chest radiography showed bibasilar infiltrates and interstitial markings (Figure 1A). A clinical diagnosis of atypical pneumonia was made and she was treated Azithromycin, Corticosteroids and Albuterol. A follow up chest X-ray on August-2016 showed resolved basilar infiltrates with some persistent interstitial prominence (Figure 1B).

Physical examination revealed tachypnea at rest, a few basal, bilateral inspiratory crackles and intermittent hypoxemia on minimal effort; oxygen therapy was started. The chest X-ray was performed and showed diffuse lower lobes reticulonodular opacities suggesting nonspecific interstitial process, possibly chronic fibrosis and Interstitial Lung Disease (ILD) was suspected (Figure 1C). Chest CT with contrast showed diffuse ground-glass opacity seen predominantly in the lower lobes with interlobular septal thickening and bilateral hilar adenopathy (Figure 1D).

Rheumatology and infectious workup including (viral and fungal pathogens) has been negative except for Mycoplasma IgM was weakly positive 1:80 and she was treated with a 5-day

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course of Azithromycin. However, patient continued to have intermittent hypoxemia and shortness of breath with effort. Broncho-Alveolar Lavage (BAL) was normal (Figure 2 A). Lung biopsy was performed by Video-Assisted Thoracoscopic Surgery (VATS) showed generalized marked distention of alveoli and bronchioles with eosinophilic granular substance which is PAS positive with diastase resistance. Occasional cholesterol clefts are seen within the alveolar contents. Only very focal collections of foamy macrophages are seen in few alveoli. The alveolar septa appear unremarkable; only hypertrophy of type II pneumocytes noted. The findings are characteristic for alveolar proteinosis. There is no evidence of pulmonary fibrosis or pneumonic infiltrate. No structures suggestive of Pneumocystis carinii are seen on GMS (Figure 2 B). Testing for surfactant protein C and ABCA 3 have also been negative.

Blood sample sent to Cincinnati's children hospital for further evaluation, which showed an elevated serum GM-CSF autoantibody concentration of 60.4 mcg/ml. (Ref range: <5.0 mcg/ml). The STAT5 phosphorylation index test was also abnormal suggesting lack of GM-CSF signaling in leukocytes. These results are indicative of autoimmune PAP.

At home, the patient was oxygen-dependent, requiring 1–2 liters of oxygen via nasal cannula to maintain oxygen saturation levels above 90%. To manage chronic respiratory symptoms, she was initiated on daily inhaled corticosteroids and as-needed albuterol for bronchodilation. A sleep study was performed due to concerns about nocturnal desaturation, which revealed frequent episodes of hypoxia during sleep, further supporting the need for continuous oxygen therapy and close respiratory monitoring.

The patient experienced multiple hospital admissions over the years due to recurrent pulmonary complications, including hypoxia, episodes of pneumonia, and wheezing. To manage worsening hypoxia, the patient underwent three separate whole lung lavages with normal saline over a three-year period, which contributed to a reduction in symptoms and decreased oxygen dependency. Despite the chronic nature of these respiratory issues, longitudinal follow-up revealed a gradual clinical improvement. After several years, the patient began to exhibit signs of remission, with stabilized pulmonary function as evidenced by a forced expiratory volume in one second (FEV_1) of 82%. Additionally, the patient reported no dyspnea on exertion, indicating a meaningful recovery in functional respiratory capacity.

Discussion

Pulmonary Alveolar Proteinosis (PAP) is an uncommon pulmonary condition characterized caused by surfactant accumulation that limits oxygen diffusion across alveoli. This disorder was initially documented in medical literature in 1958 [1].

PAP can be classified into autoimmune and non-autoimmune forms. The non-autoimmune category (about 10%) includes congenital causes and secondary cases linked to underlying diseases or environmental exposure. The autoimmune form, which represents the majority (about 90%), results from autoantibodies that neutralize Granulocyte-Macrophage Colony-Stimulating Factor (GM-CSF). This inhibition impairs alveolar macrophage function, preventing the effective breakdown and removal of surfactant within the lungs. This immune disruption results in impaired stimulation of alveolar macrophages. Consequently, these cells remain functionally immature and unable

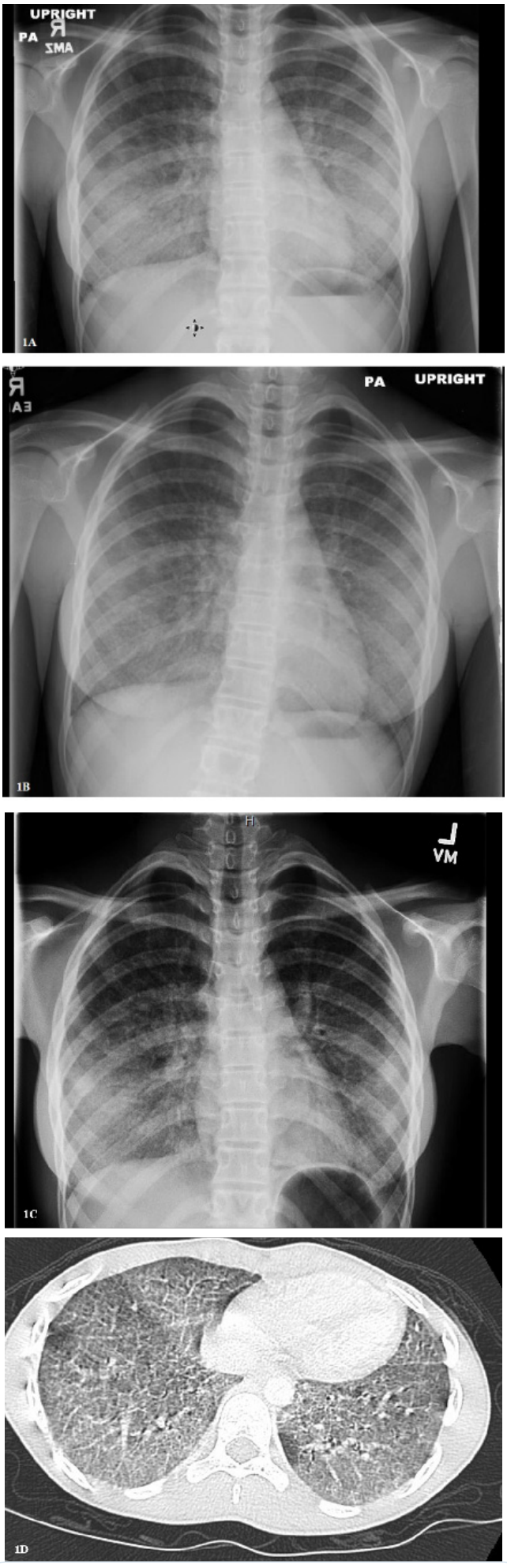

Figure A-D

to effectively degrade and clear surfactant material from the alveolar spaces. Congenital forms of PAP arise from genetic mutations affecting surfactant proteins or components of the GM-CSF receptor, with inheritance patterns that may be autosomal dominant or recessive. In contrast, secondary PAP develops as a consequence of underlying conditions such as hematologic malignancies, specific infections, or exposure to certain medications and inhaled particulates [2–4].

Dyspnea and exercise intolerance were the main clinical manifestations in our case which prompted further diagnostic studies. Chest X-rays with persistent of interstitial markings few months after empirically treatment for atypical pneumonia raised questions about chronic interstitial lung disease versus rheumatologic process.

Our patient was found to have Mycoplasma Pneumoniae on respiratory PCR. Studies showed Mycoplasma often occurs in PAP and it's difficult to ascertain whether is works as a trigger, a mere complication of the disease or incidental. Infections such as Cytomegalovirus, Mycobacterium tuberculosis, Nocardia species, and Pneumocystis jirovecii have all been implicated in reported cases of PAP. In several of these instances, deficiencies in either the number or functional capacity of alveolar macrophages were observed, suggesting a possible contributory role in disease development [5–7].

It has been proposed that *Mycoplasma pneumoniae* may bind directly to Surfactant Protein A (SP-A), facilitating persistent adhesion to type II alveolar cells, macrophages, and histiocytes [8]. This interaction could potentially disrupt normal macrophage function, initiating an immune response that leads to the production of GM-CSF autoantibodies, thereby contributing to the onset of autoimmune PAP.

Our patient has undergone Whole Lung Lavage (WLL) a year later. A lavage with 1 L saline solution in each lung was performed until the liquid was clear. Her lavage fluid was noted to be turbid and cloudy. Cultures were sent which were negative. Overall, the patient tolerated the procedure well. The outcome has been satisfactory so far, the patient still on supplemental oxygen of 1.5L at night and on room air during the day. She attends school normally. The patient will certainly need the support of a multidisciplinary team, and Whole Lung Lavage (WLL) has shown clinical effectiveness in managing PAP, particularly in adolescents and adults, with reported improvement rates ranging from 60% to 84% [9–10]. In select cases, Rituximab has also been utilized as an alternative therapeutic approach, demonstrating benefit in a limited number of adult patients documented in case reports [11].

Autoimmune Pulmonary Alveolar Proteinosis (PAP) generally carries a favorable outlook when managed with whole lung lavage, with five-year survival rates reported as high as 95% [12]. Early case series had suggested that spontaneous resolution might occur in up to 50% of patients [13,14], but more recent and larger cohort studies indicate this is much less common — likely affecting fewer than 10% of cases. Despite overall positive outcomes, mortality is still observed in a subset of patients, primarily due to complications from hematologic disorders (33%), infections (25%), respiratory failure (25%), and hemorrhagic events (13%) [12].

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