Isolated Pauci-Immune Pulmonary Capillaritis- a Rare Cause of Diffuse Alveolar Hemorrhage

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Diffuse alveolar hemorrhage (DAH) can result from various etiologies. It may present as acute respiratory failure. In most cases, it is initially diagnosed and treated as multifocal pneumonia. We present a rare case of isolated pauci-immune pulmonary capillaritis (IPPC) as a cause of DAH. A 55-year-old female presented with 3 days of sudden onset of progressive dyspnea with associated dry cough. She denied fever, chest pain, wheezing and palpitations. Her past medical history includes hypothyroidism, hypertension, carpal tunnel syndrome, osteoarthritis, morbid obesity and a hospitalization with a similar presentation earlier. Her home medications include levothyroxine, lisinopril, ferrous sulfate and albuterol nebulization. She has a former smoking history of 15 pack-years. On examination, she was afebrile and hemodynamically stable. She was hypoxic with acute respiratory distress. Her lung auscultation revealed fine bibasilar rales. Her lab workup revealed hemoglobin 9.0 g/dL, and hematocrit 28.8%, WBC 9100/µL and lactic acid 1.5 mmol/L. The chest X-ray showed cardiomegaly with diffuse interstitial and alveolar airspace opacities suspicious for diffuse pulmonary edema, with superimposed consolidation. Her CT scan one month and 6 years prior showed findings of diffuse confluent consolidative and ground-glass opacities throughout the lungs bilaterally with relative sparing of bases. She was treated with vancomycin, cefepime, oxygen, non-invasive ventilation in ICU with preliminary diagnosis of acute hypoxemic respiratory failure secondary to hospital acquired pneumonia. Respiratory virus and vasculitides panels (ANCA, ANA, anti-GBM antibodies) were negative, HIV was non-reactive, blood cultures were negative, and creatinine remained stable. Despite antibiotics and solumedrol, she did not improve clinically. A bronchoscopy showed diffuse alveolar hemorrhage. A lung biopsy showed capillaritis, alveolar hemorrhage, and no granuloma. A diagnosis of IPPC was considered based on pulmonary capillaritis without systemic involvement and negative vasculitides serology. After multidisciplinary discussion, she was given pulse dose of steroids and started on IV cyclophosphamide in addition to prednisone for three months followed by methotrexate. She was discharged with follow up with pulmonary and rheumatology and has had significant clinical improvement. IPPC is a rare disease of unknown etiology which is characterized by inflammation limited to the pulmonary vasculature without any systemic involvement. ANCA and ANA are negative. It has been reported to be successfully treated with cyclophosphamide. Our case also demonstrates its response to steroid and cyclophosphamide and maintenance methotrexate therapy. Further studies are needed to understand the pathogenesis and to establish the treatment guidelines for the disease.