Necrotizing crescentic glomerulonephritis is found in A 63 year-old man with a 60 pack-year smoking history presented to a community hospital complaining of shortness of breath, non-exertional chest pain and recent history of hemoptysis. He had not been seen by a doctor in several years, had no known medical problems, and took no medications. He denied any similar health problems in his family. He was afebrile on physical exam and had a blood pressure of 160/110 mmHg. His heart rate was 108, respiratory rate 20, and there was no edema, weight gain, orthopnea or paroxysmal nocturnal dyspnea. Laboratory investigation revealed proteinuria, a creatinine of 1.4 mg/dL, and positive circulating c-ANCA antibodies. Protein/creatinine ratio 3.52.

The patient improved and was discharged on 10 mg of oral prednisone daily and hemodialysis to out-patient follow-up with rheumatology and nephrology.

CT Chest showed multiple bilateral pulmonary nodules, biopsy of which showed only hemosiderin-laden macrophages. Renal biopsy showed linear IgG staining suspicious for Goodpasture syndrome and necrotizing crescentic glomerulonephritis with 70% interstitial fibrosis on immunofluorescence microscopy. The patient was treated with hemodialysis, 7 rounds of plasmapheresis, rituximab, and solumedrol, complicated by upper gastrointestinal bleed requiring admission to the intensive care unit. After resolution he was continued on oral prednisone.

The differential diagnosis of Pulmonary-Renal Syndromes comprise Goodpasture syndrome, granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis, microscopic polyangiitis, SLE, sarcoidosis, TB and metastatic lung cancer. The typical presenting features of GPA vs GBM are hemoptysis with worsening renal function.

Rare cases have been seen with ANCA positive antibodies and linear immunofluorescence pattern staining have been discussed, however none with prior association with c-ANCA.

We report a case of systemic c-ANCA pulmonary-renal syndrome with anti-GBM-disease-typical linear staining on renal biopsy immunofluorescence.

Case Presentation

A 63 year-old man with a 60 pack-year smoking history presented to a community hospital complaining of shortness of breath, non- exertional chest pain and recent history of hemoptysis on a background history of chronic non-productive cough lasting many years. He had not been seen by a doctor in several years, had no known medical problems, and took no medications. He denied edema, weight gain, orthopnea or paroxysmal nocturnal dyspnea. Physical exam was significant for hypertension and course bilateral breath sounds with expiratory wheeze.

Laboratory investigation revealed proteinuria, a creatinine of 11.4 mg/dL, and positive circulating c-ANCA antibodies. Protein/creatinine ratio 3.52.

Discussion

The differential diagnosis of Pulmonary-Renal Syndromes comprise Goodpasture Syndrome, granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis, microscopic polyangiitis, SLE, sarcoidosis, TB and metastatic lung cancer.

Rare cases have been seen with ANCA positive antibodies and linear immunofluorescence in necrotizing crescentic glomerulonephritis, the majority of which are p-ANCA/MPO positive.

Patients presenting with suspected pulmonary-renal syndrome should be tested for both anti-MPO and anti-PR3 ANCA-related disease and anti-GBM disease. Further study of pulmonary-renal syndromes with ANCA vasculitis and linear immunoglobulin staining without anti-GBM serum antibodies are necessary to understand the pathogenesis and develop necessary treatment protocols.