Learning Objectives

1. Patients presenting with suspected pulmonary-renal syndrome should be tested for both anti-MPO (p-ANCA) and anti-PR3 (c-ANCA)
2. Pulmonary renal syndromes present with a spectrum of immunohistochemical features, with rare instances of overlap.
3. Immunohistochemical patterns of overlap may have prognostic implications for patients.

Introduction

Necrotizing crescentic glomerulonephritis is found in anti-glomerular basement membrane (GBM) disease (Type 1), immune complex (Type 2) deposition and anti-neutrophil cytoplasmic (ANCA)-related disease (Type 3).

ANCA positive glomerulonephritis is typically characterized on renal biopsy as pauci-immune, with mild or absent glomerular staining for immunoglobulin, or complement staining by immunofluorescence.

Rare cases of concomitant p-ANCA plus 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, bilateral breath sounds with expiratory wheeze.

Laboratory investigation revealed proteinuria, a creatinine of 3.52.

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, bilateral breath sounds with expiratory wheeze.

Case Presentation

A 63 year-old man with a 60 pack-year smoking history presented to a community hospital complaining of shortness of breath, bilateral breath sounds with expiratory wheeze.

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

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. 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 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.

References