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The Bugs were not to Blame; Granulomatosis with Polyangiitis Presenting as a Renal Mass

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Abstract

Granulomatosis with Polyangiitis (GPA) is an anti-neutrophil cytoplasm antibody (ANCA) associated vasculitis that rarely presents as a solitary renal mass. We present a 57-year-old male who presented with abdominal pain and was found to have a complex right renal lesion on abdominal imaging. He later developed fevers, hypoxic respiratory failure and acute kidney injury. Chest imaging showed confluent consolidative changes in the mid to lower lung zones. He was started on antibiotics but continued to worsen. He was noted to have diffuse alveolar hemorrhage on bronchoscopy with autoantibodies to proteinase-3 (PR-3) returning positive. He was started on rituximab and steroids with dramatic improvement. Kidney biopsy revealed crescentic and necrotizing glomerulonephritis associated with ANCA-associated vasculitis. GPA presenting as a renal mass presents a diagnostic challenge. Prompt diagnosis is however paramount given how fast GPA can progress.

Keywords: Granulomatosis with polyangiiti • Solitary renal mas • Diffuse alveolar hemorrhage • Necrotizing crescenteric glomerulonephritis

Introduction

Granulomatosis with Polyangiitis (GPA), previously known as Wegener's granulomatosis, is an anti-neutrophil cytoplasm antibody (ANCA) associated vasculitis of unknown etiology. It has an annual incidence of 12.8 per 1 person million years in the US [1]. Typical presentation is described as nasal involvement with pulmonary renal syndrome. We present a rare case of GPA that initially manifested as a renal mass.

Case Presentation

57 year old male with no significant past medical history presented with fever, cough and dyspnea. He was admitted to the hospital 2 weeks prior with abdominal pain and was found to have a 4-centimeter right renal complex lesion (Figure 1). Biopsy showed necrotizing mass with multinucleated giant cells (Figure 2), and xanthogranulomatous pyelonephritis was suggested. Patient received empiric antibiotics with a plan for nephrectomy. Urine cultures remained negative. Two days after discharge, he developed fevers, cough and dyspnea that prompted him to return to the hospital. He endorsed anorexia, night sweats and unintentional weight loss for 2 weeks.

Patient was febrile and hypoxic in the emergency room. Initial labs showed normocytic anemia, leukocytosis, hypoalbuminemia and acute kidney injury. Urinalysis showed 2+ proteinuria, 3+ hematuria and 6-10 white blood cells. Chest imaging was significant for confluent consolidative changes in the mid to lower lung zones and small to moderate bilateral pleural effusions (Figure 3).

Broad spectrum antibiotics were initiated for suspected pneumonia. He was noted to have worsening anemia requiring blood transfusions. Fevers persisted despite antibiotics. Hypoxic respiratory failure and acute kidney injury continued to worsen. Taking into consideration the recent biopsy of

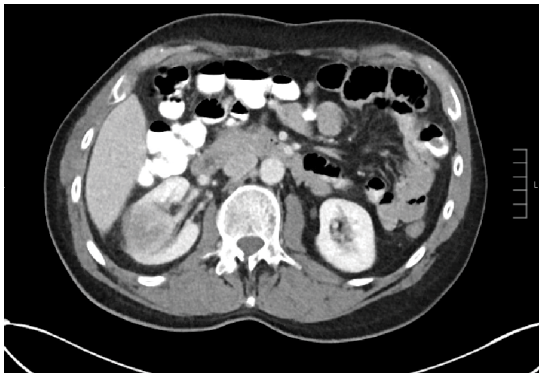
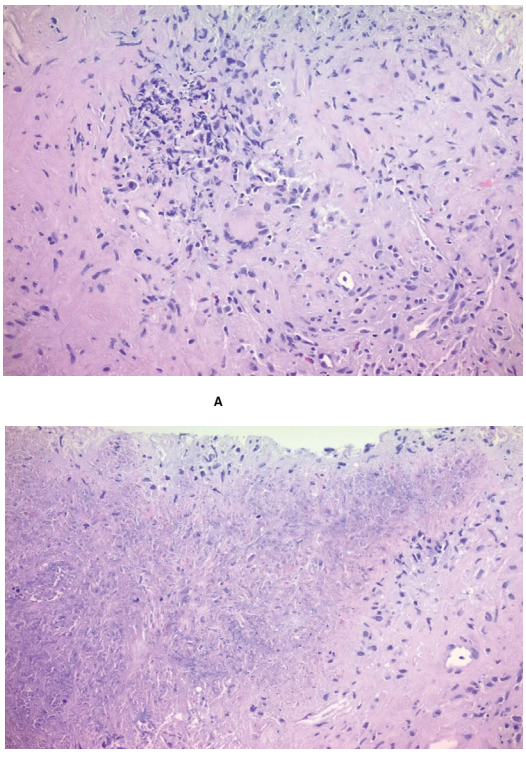


Figure 1. Complex right renal cyst noted on initial presentation.



A

B

Figure 2. Initial renal mass biopsy with a destructive inflammatory process including 'dirty necrosis' (A) and multinucleated giant cells (B).

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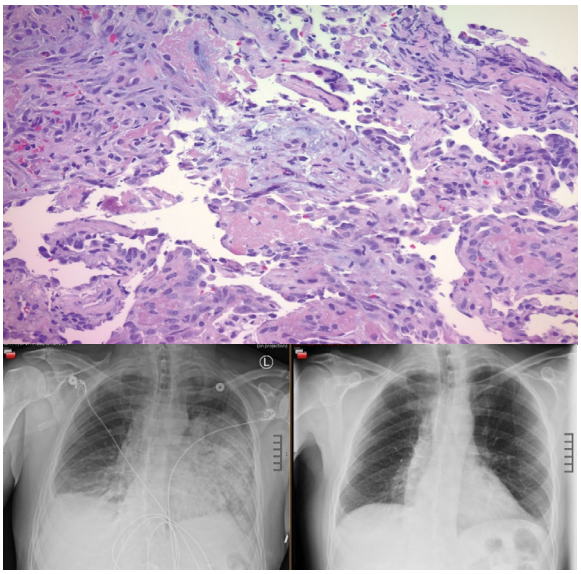
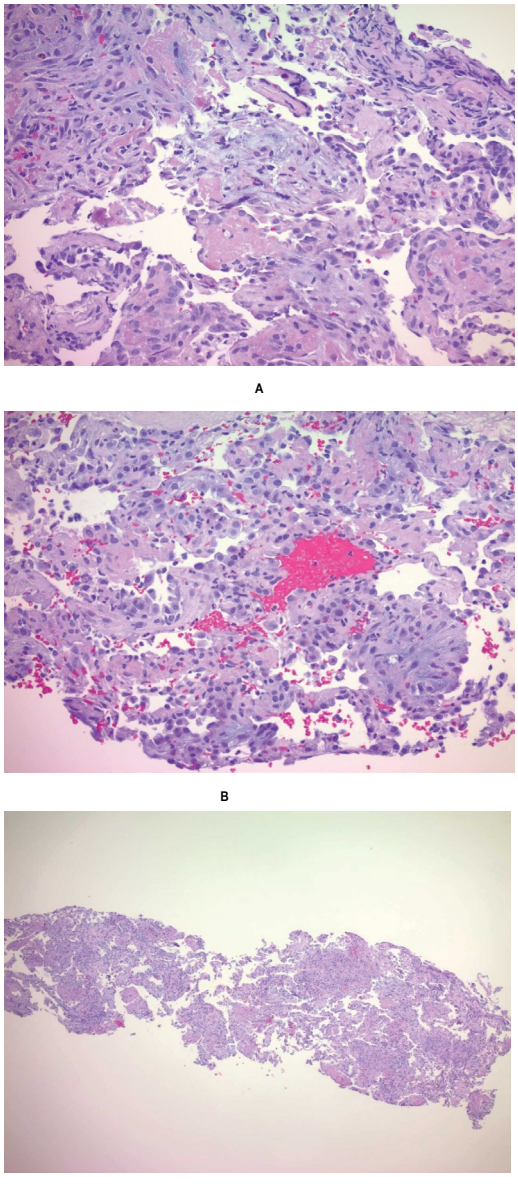


Figure 3. CXR on initial presentation (right), CXR on readmission 2 weeks later (left).



A

B

C

Figure 4. Transbronchial lung biopsy with organizing pneumonia, reactive pneumocytes, airspace fibrin and fresh hemorrhage low (A), high power (B) and few interstitial neutrophils (C).

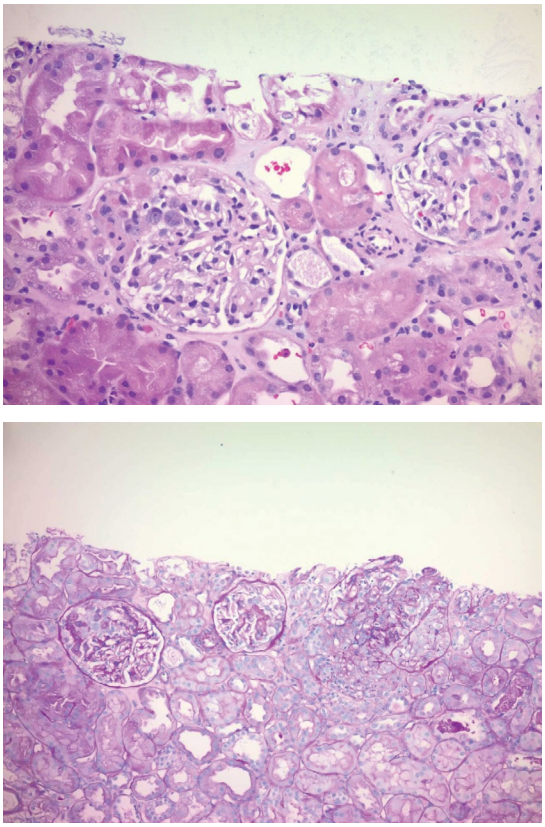


Figure 5. Subsequent renal biopsy with necrotizing crescentic glomerulonephritis (H&E stain (top), PAS stain; bottom).

renal mass which showed necrosis and occasional multinucleated giant cells, ANCA-associated vasculitis with diffuse alveolar hemorrhage (DAH) and rapidly progressive glomerulonephritis (RPGN) was suspected. Autoantibodies to proteinase-3 (PR-3) were found to be positive. Bronchoscopy showed evidence of diffuse alveolar hemorrhage; transbronchial biopsies showed organizing pneumonia (OP), airspace fibrin mixed with fresh blood, diffusely reactive pneumocytes and interstitial neutrophils (Figure 4). In the context of his prior renal biopsy and high titer anti-PR-3, lung biopsy findings were interpreted as compatible with an ANCA-associated vasculitis. Biopsy of the contralateral kidney showed crescentic and necrotizing glomerulonephritis associated with ANCA-associated vasculitis (Figure 5). Patient was started on high dose steroids and rituximab. 3 days after initiating steroids, his respiratory

status dramatically improved. Renal function stabilized, and the patient did not require hemodialysis. He was discharged home in stable condition.

Discussion and Conclusion

GPA presentation can be elusive at times. Pulmonary manifestations range from asymptomatic nodules to diffuse alveolar hemorrhage. Radiographically, we can encounter pulmonary infiltrates, nodules and cavitary lesions. Microscopic examination of the lung can show large necrotizing fibroinflammatory lesion, with central basophilic “dirty” necrosis, palisading histiocytes, multinucleated giant cells and sometimes giant cell vasculitis in the vessels away from the area of necrosis [2]. Our case demonstrates a rare presentation with DAH and OP [3-5]. Renal manifestations are usually rapidly progressive GN with hematuria, red cell casts and proteinuria. GPA manifesting as a solitary renal mass is exceptionally rare, with only fifteen cases reported in the literature [3]. In patients presenting with RPGN, end stage renal disease usually ensues within weeks of onset, therefore prompt diagnosis and treatment are essential. 75 to 80% of patients with GPA stain positive for cytoplasmic ANCA on immunofluorescence and have antibodies directed against PR-3. About 10-15% have antibodies directed against myeloperoxidase and have perinuclear staining on immunofluorescence. Our patient presented with unilateral renal mass and constitutional symptoms, attributed to xanthogranulomatous pyelonephritis and absence of typical signs of GPA. He later developed DAH and RPGN which prompted autoimmune workup. PR-3 antibodies returned positive and the biopsy of contralateral kidney revealed findings consistent with GPA. GPA manifesting as a renal mass is extremely rare and presents a diagnostic challenge.

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