Case Report

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A Rare Case of Anti Neutrophil Cytoplasmic Antibody Associated Vasculitis Presenting as Pyelonephritis

Internal Medicine Section

CHETAN PHADKE[1], SHREEHARSH GODBOLE[2]

ABSTRACT

Anti-Neutrophil Cytoplasmic Antibody (ANCA) Associated Vasculitis (AAV) is a condition affecting small and medium-sized vessels, predominantly impacting the kidneys. Microscopic Polyangiitis (MPO) is primarily associated with antibodies to myeloperoxidase. MPO frequently presents as glomerulonephritis, but pyelonephritis has not been mentioned hitherto in the literature as an initial presentation. Granulomatosis with Polyangiitis (GPA) is known to present as pyelonephritis, but not MPO. Such an unusual presentation may masquerade as an infectious condition when the underlying cause may, in fact, be an autoimmune one. This case report concerns a patient who presented with pyelonephritis. The patient also exhibited lung involvement that appeared to be pneumonia. Sepsis was suspected initially, and antibiotics were commenced. He also experienced renal failure and required haemodialysis. As the condition did not resolve, extensive investigations were conducted. An autoimmune panel and a renal biopsy revealed AAV as the underlying condition. Prompt treatment with glucocorticoids and rituximab resulted in a dramatic improvement in the patient's condition. The patient did not require further haemodialysis, and he subsequently recovered and was discharged. The patient is on regular follow-up and requires only a small maintenance dose of prednisone. This case report demonstrates that pyelonephritis may not always have an infection as its underlying cause. A high index of suspicion for an autoimmune cause may result in early treatment and can be life-saving.

Keywords: Biopsy, Microscopic polyangiitis, Renal insufficiency

CASE REPORT

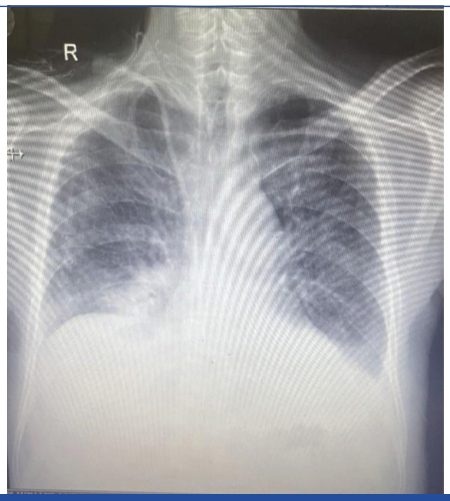
A 61-year-old Asian Indian male presented to the outpatient department with fever, cough, and bilateral flank pain that had persisted for the last month. The pain increased upon straining and during micturition, and was of moderate intensity. He had no history of diabetes mellitus, hypertension, tuberculosis, urinary tract infections, or any previous hospital admissions. On admission, his renal parameters and urine analysis were found to be abnormal, along with the presence of proteinuria. The laboratory investigations are provided in [Table/Fig-1]. He also exhibited anaemia and leucocytosis, while the urine culture showed no growth.

Laboratory parameter	Initial presentation	After treatment	Follow-up
Serum urea	137 mg/dL	99 mg/dL	40 mg/dL
Serum creatinine	7.39 mg/dL	2.1 mg/dL	1.4 mg/dL
Urinanalysis	protein 2+, pus cells 12-15/hpf, RBCs 4-5/hpf	protein trace, pus cells 2-3/hpf, RBCs 6-8/hpf	protein trace, pus cells 1-2/hpf, RBC, 1-2/hpf
Proteinuria	3.12 mg/mg (PCR)	2.89 mg/mg (PCR)	0.46 mg/mg (PCR)

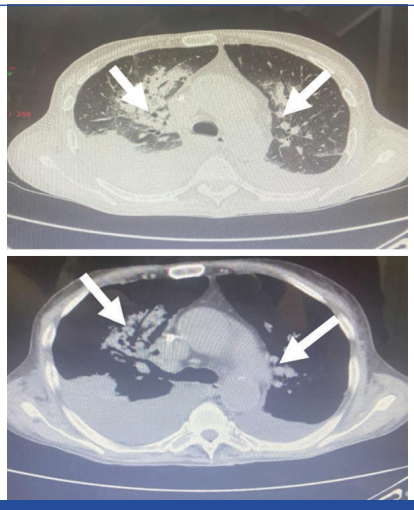
[Table/Fig-1]: Relevant laboratory parameters.
PCR: Protein creatinine ratio

The chest X-ray [Table/Fig-2] revealed significant confluent consolidative changes in the mid to lower right lung zones, consistent with pneumonia, as well as a small to moderate pleural effusion. A High-Resolution Computed Tomography (HRCT) scan of the chest suggested bilateral moderate pleural effusion with partial collapse of the bilateral lower lobes [Table/Fig-3]. An ultrasound of the abdomen showed bilateral normal-sized kidneys (right kidney: 11.0×5.3 cm; left kidney: 11.3×5.7 cm), with bilateral raised renal cortical echogenicity and mild perinephric fat stranding, suggestive of pyelonephritis, which was subsequently confirmed on a non-contrast Computed Tomography (CT) scan of the abdomen [Table/Fig-4].

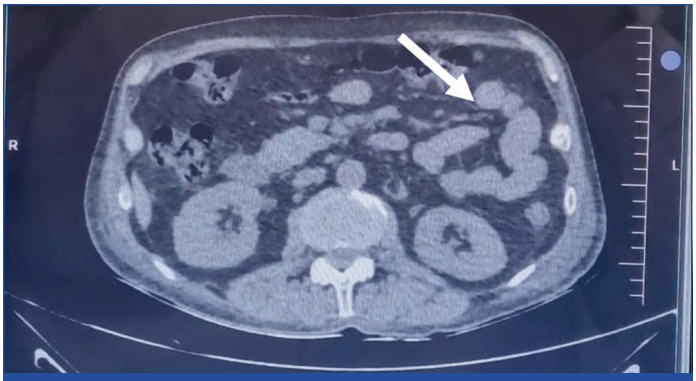
Antibiotics were started based on suspicion of sepsis. In light of his symptoms and laboratory parameters, the patient was initiated on



[Table/Fig-2]: Chest X-ray (PA) view (arrow shows consolidation).



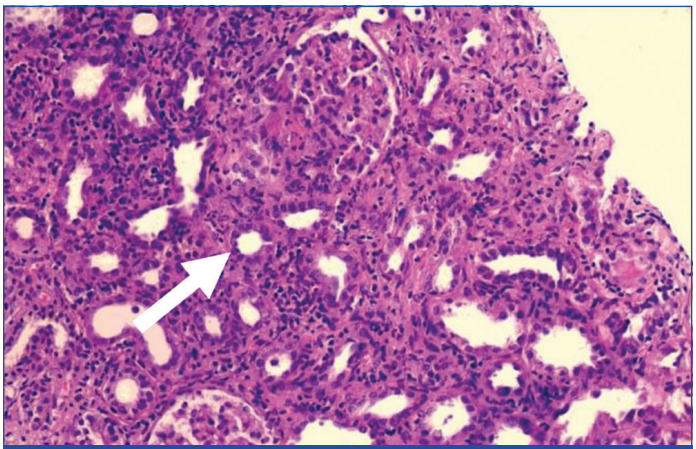
[Table/Fig-3]: HRCT Thorax (arrows indicate partial collapse).



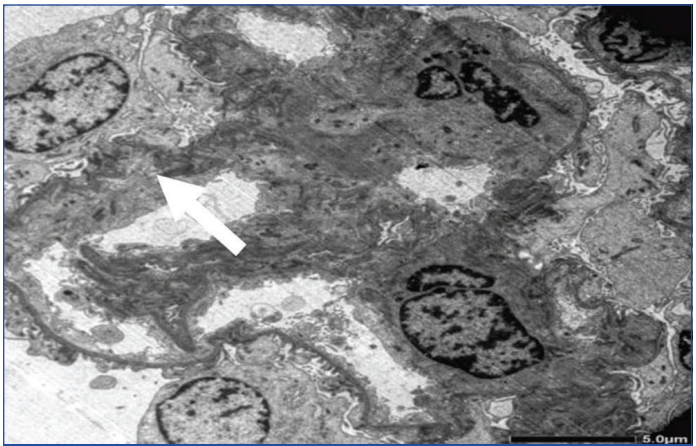
[Table/Fig-4]: Non-contrast Computed Tomography (CT) of the Abdomen (arrows show perinephric fat stranding).

haemodialysis. While the symptoms subsided, the pleural effusion persisted, and the chest X-ray showed no improvement. A pleural tap revealed transudative fluid, and tuberculosis was ruled out by a nucleic acid amplification test. An autoimmune panel revealed a normal C3 level (99 mg/dL), a normal C4 level (37 mg/dL), a negative antinuclear antibody result, and a negative ANCA for proteinase 3. However, the ANCA for myeloperoxidase proved to be positive.

A renal biopsy [Table/Fig-5,6] was performed. Light microscopy revealed 17 glomeruli, of which six exhibited crescents (four cellular and two fibrocellular), and seven demonstrated fibrinoid necrosis with perinecrotic neutrophils. The tubules showed severe acute injury with interstitial fibrosis and tubular atrophy affecting 25% of the sampled cortex. The arteries exhibited necrotising vasculitis. Direct immunofluorescence was negative. Transmission electron microscopy revealed a normocellular glomerulus, with thirty percent of the loops showing foot process flattening. No sclerosis, electron-dense deposits, or organised deposits were observed. Thus, the renal biopsy was suggestive of pauci-immune ANCA-Associated Vasculitis (AAV) with crescents. A diagnosis of ANCA AAV presenting as MPO was established.



[Table/Fig-5]: Light microscopy of renal biopsy, 200X, PAS (arrow showing fibrinoid necrosis with perinecrotic neutrophils).



[Table/Fig-6]: Electron microscopy of renal biopsy (arrow shows foot process flattening in glomerulus).

The patient was started on pulse methylprednisolone 500 mg for three days, followed by oral prednisone along with rituximab. Within a week of starting treatment, the patient improved dramatically. His renal function parameters improved, and his pleural effusions did not recur. The patient did not require haemodialysis thereafter. Two months into follow-up, the patient was doing well, with normal renal parameters and urine analysis. At the time of the latest follow-up, he was on a low dose of prednisone as a maintenance regimen.

DISCUSSION

ANCA-associated vasculitis (ANCA-AAV) is an inflammatory disorder of the small arteries characterised by vascular destruction and tissue necrosis that can affect various organs [1]. It predominantly impacts the kidneys [2]. Glomerulonephritis is a frequent manifestation of AAV, presenting in almost all cases of Myeloperoxidase (MPO) and frequently in granulomatosis with polyangiitis (GPA) [3]. MPO is primarily associated with antibodies to myeloperoxidase [4]. The worldwide reported prevalence of AAV ranges from 4.6 to 21.8 cases per one million person-years [5]. The average onset of AAV is 65 years, with prevalence peaking in the 70-75 age group [6]. Solid ANCA positivity, when accompanied by relevant clinical symptoms, may be sufficient to initiate remission induction treatment without a renal biopsy. However, biopsy is highly recommended and provides "gold-standard" diagnostic results, especially in cases of ambiguous serology or unusual clinical presentations [7].

While pyelonephritis associated with GPA has been described previously in the literature [8], there have been no reports of pyelonephritis due to Microscopic Polyangiitis (MPA). Due to the relapsing nature of AAV, disease activity needs to be monitored prospectively for adequate treatment [9]. Glucocorticoids in combination with rituximab or cyclophosphamide are recommended as the initial treatment for new-onset AAV (1B Recommendation) [10]. Maintenance therapy with either rituximab or azathioprine, along with low-dose glucocorticoids after induction of remission, is also recommended (1C Recommendation) [10].

CONCLUSION(S)

AAV presents in a myriad of ways. This case highlights the need to maintain a high index of suspicion for AAV in the differential diagnosis of renal failure, particularly in the presence of other systemic involvement. Prompt diagnosis and treatment can dramatically improve patient outcomes.

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PARTICULARS OF CONTRIBUTORS:
1. Resident, Department of Nephrology, Dr. D. Y. Patil Medical College, Pune, Maharashtra, India.
2. Resident, Department of Nephrology, Dr. D. Y. Patil Medical College, Pune, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Shreeharsh Godbole,
Resident, Department of Nephrology, Dr. D. Y. Patil Medical College,
Sant Tukaram Nagar, Pimpri, Pune, Maharashtra, India.
E-mail: shreeharshgodbole@gmail.com

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