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### RESEARCH ARTICLE

#### AN UNUSUAL CAUSE OF RENAL FAILURE.

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#### Manuscript Info

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#### Abstract

A 58-year old female, a known asthmatic, presented with acute breathlessness for 1 week, and was found to have acute kidney injury. Renal biopsy revealed pauci-immune necrotising glomerulonephritis and necrotising vasculitis. She was positive for P-ANCA, and a diagnosis of ANCA-associated vasculitis with rapidly progressive glomerulonephritis was made.

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#### Introduction:-

##### Presentation of case:-

A 58-year old female, a known case of bronchial asthma, which was well controlled with short acting inhaled beta agonists for the last 8 years, now presented with an acute worsening of dyspnoea for the last one week, which was associated with paroxysmal nocturnal dyspnoea. She also complained of a gradual painless swelling of both feet for the last one week. She also had a history of diminished urine output for one week, with no change in the colour of urine, and no history of burning micturition.

On examination, she had bilateral pitting pedal oedema, and a blood pressure of 160/100 mmHg. Her jugular venous pressure was elevated, and she had a positive hepatojugular reflux. Auscultation of the cardiovascular system did not reveal any murmurs or added sounds, but inspiratory crackles were present bilaterally in the lung bases. The remainder of the systemic examination was unrevealing.

A complete haemogram revealed microcytic hypochromic anaemia with a haemoglobin of 6.3 g/dL, along with thrombocytosis (platelet count 612,000/mm<sup>3</sup>). She had renal failure, with a serum creatinine of 3.9 mg/dL and a blood urea of 184 mg/dL. A repeat serum creatinine 24 hours later was 6.2 mg/dL. Urinalysis revealed 2+ albuminuria, along with 3-4 RBCs per high power field. Ultrasound of the kidneys showed bilaterally increased cortical echoes, with the right kidney measuring 8.7x4.5 cm and the left kidney measuring 9.1x4.1 cm. A renal biopsy was performed, which revealed pauci immune necrotising glomerulonephritis and necrotising vasculitis, raising the possibility of ANCA associated vasculitis. In view of this, indirect immunofluorescence for P-ANCA and C-ANCA was performed, and she was found to be positive for P-ANCA, thus confirming the diagnosis of ANCA-associated vasculitis. Because of the long-term history of bronchial asthma, the possibility of Eosinophilia with Granulomatosis and Polyangiitis (Churg-Strauss) vasculitis was considered; however there was no eosinophilia on the peripheral smear and the absolute eosinophil count was only 153/mm<sup>3</sup>.

##### Differential diagnosis

1. Rapidly progressive renal failure
2. Acute exacerbation of bronchial asthma

**Clinical diagnosis:-**

Rapidly progressive renal failure

**Pathologic discussion:-**

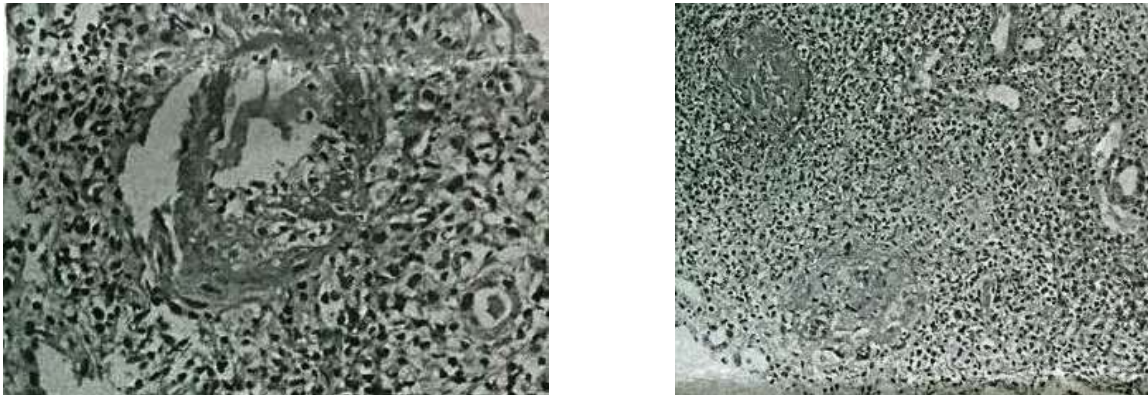
An ultrasound-guided renal biopsy was performed on the left kidney, and the biopsy specimen was sent for light microscopy and immunofluorescence.

On light microscopy, nine glomeruli were identified in the section studied. Four were globally sclerotic and three were in varying stages of sclerosis. The Bowmans capsule of these glomeruli was ruptured and there was a dense inflammatory cell infiltrate in the surrounding interstitium. Segmental fibrinoid necrosis of the capillary tuft was identified in two glomeruli. Dense lymphocytic infiltration was present in the entire tubulointerstitial compartment, and 20% of the tubules were atrophic and filled with PAS positive hyaline casts.

Another section which showed 4 glomeruli was stained for IgG, IgM, IgA, C3, C1q, kappa and lambda light chains, all of which were negative.

**Impression:-**

Necrotising glomerulonephritis and necrotising vasculitis, pauci-immune. The possibility of ANCA associated vasculitis is suggested.



**Fig. 1:-** Histopathology of renal biopsy

**Discussion of management:-**

In view of the patient's deteriorating renal parameters and hypervolaemia, the right internal jugular vein was cannulated and a haemodialysis catheter inserted. She was initiated on immunosuppression, with both intravenous cyclophosphamide and three days of pulse methylprednisolone, which was later switched to oral steroids. After two cycles of haemodialysis her urine output improved and she was euvolaemic. Her haemodialysis catheter was therefore removed, and the patient is currently not on renal replacement therapy.

Antineutrophil cytoplasm antibody (ANCA)-associated vasculitis is a small-vessel vasculitis that includes<sup>1</sup>

1. Granulomatosis with polyangiitis (GPA) – this was formerly called Wegener's granulomatosis
2. Microscopic polyangiitis (MPA)
3. Eosinophilic granulomatosis with polyangiitis (EGPA) – this was formerly called Churg–Strauss syndrome

Renal-limited ANCA-associated vasculitis can be considered the fourth entity, though it eventually corresponds to a kidney-limited form of MPA or GPA.<sup>2</sup>

The precise cause of ANCA-associated vasculitis remains unknown. It has been hypothesised that an infectious agent, such as *Staphylococcus aureus* for GPA, (over)activates the immune system<sup>3</sup>.

Treatment of severe ANCA-associated vasculitis comprises two phases:

1. Remission induction therapy based on the combination of glucocorticoids and another immunosuppressive agent (often cyclophosphamide)
2. Maintenance therapy

Newer biologic agents such as rituximab appear promising in the treatment of ANCA-associated vasculitis.<sup>4</sup>

**Final diagnosis:-**

ANCA-associated vasculitis, presenting as rapidly progressive glomerulonephritis.

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