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Case Report

A rare presentation of rheumatoid arthritis with simultaneous occurrence of MPO-ANCA associated vasculitis: a case report

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Introduction

Secondary amyloidosis and the use of non-steroidal anti-inflammatory drugs (NSAIDs) are the common causes of renal failure in rheumatoid arthritis (RA) [1,2]. Pauci-immune crescentic glomerulonephritis is a rare renal complication of RA. Because of its rarity and the presence of overlapping features, the diagnosis can be easily missed. Other than pauci-immune crescentic glomerulonephritis, mesangial proliferative and membranous glomerulonephritis have been identified in autopsy studies of patients with RA [1]. In addition, there have been published cases of pauci-immune necrotizing glomerulonephritis in patients with acute rheumatic flares and deteriorating renal function [1-3]. Patients with pauci-immune necrotizing glomerulonephritis on long term treatment for RA develop severe forms of RA, such as erosive RA with involvement of multiple organs. We present a case of simultaneous RA and myeloperoxidase positive glomerulonephritis.

Case Presentation

A 42-year-old male with rheumatoid arthritis of 19 years duration (diagnosed at the age of 23 years) had been clinically in remission for more than 10 years and off treatment for 10 years. He was admitted to our hospital with a history of small and large joint pain and swelling of 2 months duration. The joints involved were bilateral small joints of the hand and the ankle and knee joints. He had a history of pleuritic type chest pain and haemoptysis and haematuria for one week. Systemic evaluation did not reveal any other abnormalities.

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On examination, he was afebrile. He was pale but not icteric. There was no lymph node enlargement. His blood pressure was 120/80 mmHg and pulse rate was 80bpm. Oxygen saturation was 99% on room air. Rest of the cardiovascular system examination did not reveal any abnormalities. Respiratory system examination revealed bilateral lower zone fine crepitations, more on the right lung.

Although the patient gave a history of bilateral small and large joint swelling for 2 months, examination of the joints did not reveal any features of synovitis. Abdominal and nervous system examination was unremarkable. His investigations on admission are given in Table 1.

Table 1: Blood investigations on admission

Investigation	**Value (normal range)**
WBC (10^9/L)	12.3(4.0-10.0)
Hb (g/dL)	10.8(11.0-15.0)
PLT (10^9/L)	403(150-450)
Blood Urea (mmol/L)	4.9(1.8-6.3)
Serum Creatinine (µmol/L)	129(53-88)
Serum Sodium (mmol/L)	136(136-145)
Serum Potassium (mmol/L)	4.7(3.5-5.1)
SGOT(U/L)	299(15-37)
SGPT(U/L)	111(12-78)
Gamma GT(U/L)	441(5 – 40)
INR	0.95(<1.1)
APTT (sec)	32(21-35)
C-reactive protein(mg/L)	93(0-5)
ESR (mm/1st hour)	86(0-29)
Albumin(g/dL)	3.9
Rheumatoid factor	negative
urine protein creatinine ratio	784
urine full report	protein - 3+ pus cells -10-12/HPF red cells - 80-90/HPF dysmorphic red blood cells - negative

The chest X-ray showed features of diffuse air space opacity in the mid and lower zones (Figure 1). HRCT showed features of "bilateral airspace opacity and crazy paving" (Figure 2). These features represent pulmonary haemorrhage. Ultrasound scan of the abdomen showed a right sided pelvic kidney, early renal parenchymal changes on the left side and an echogenic liver. P-ANCA was positive and C-ANCA was negative. Antinuclear antibody was negative.

A multi-disciplinary team, which included a nephrologist, rheumatologist, respiratory physician and physician, assessed the clinical features and biochemical markers. A probable diagnosis of P-ANCA vasculitis causing pulmonary renal syndrome was made. It was confirmed by a renal biopsy. Therefore, anti-glomerular basement disease was excluded. According to the decisions taken at the MDT meeting, intravenous methyl prednisolone 1g daily was given for 3 days. Following this, oral prednisolone 1mg/kg/day was initiated. The dose of prednisolone was tapered according to the clinical improvement. It was also decided to continue a maintenance dose of prednisolone of 10mg daily for one year [4].

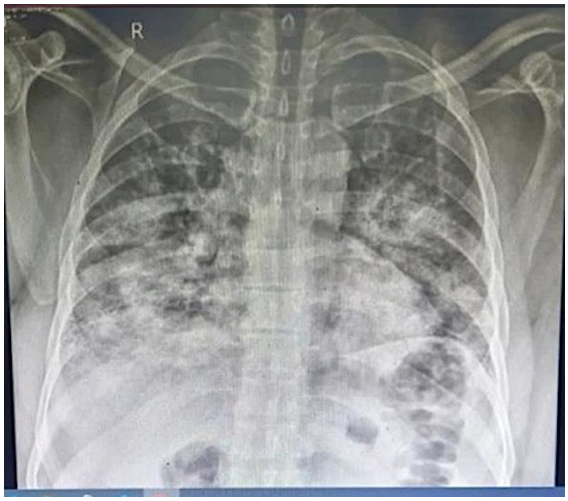

Figure 1: Diffuse air space opacity in mid and lower zones

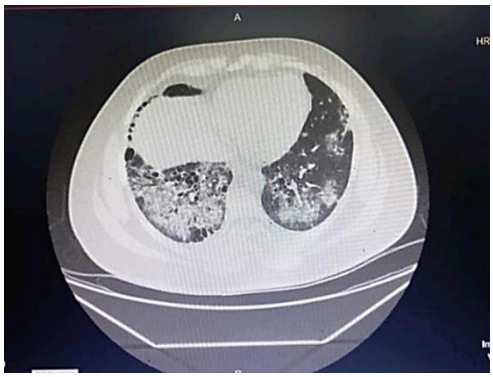

Figure 2: HRCT showed features of "bilateral airspace opacity and crazy paving"

Renal biopsy showed segmental necrotizing glomerular nephritis consistent with pauci immune small vessel vasculitis. Intravenous cyclophosphamide 500mg 2 weekly for 2 doses was given. It was decided to continue intravenous cyclophosphamide 1000mg monthly for 6 months and change to mycophenolate mofetil later.

Discussion

The presentation of rheumatoid arthritis varies widely from patient to patient. Anti-MPO antibodies were positive in 12% of patients with rheumatoid arthritis with different levels of disease activity [5,6]. The incidence of ANCA in RA is as high as 40% and individuals have expressed both c-ANCA and p-ANCA staining patterns [7].

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Disease severity is increased in RA patients with P-ANCA positivity and inflammatory markers are usually higher in them. They develop complications related to the vascular and pulmonary system. It is hypothesized that management with methotrexate and infliximab can influence the activity of crescentic glomerular nephritis [8].

We present a case of a patient with RA who presented with haemoptysis, pulmonary involvement, glomerular proteinuria and abnormal renal function not attributable to anti-rheumatoid drugs.

After assessing the clinical features and biochemical markers, a diagnosis of P-ANCA vasculitis causing pulmonary renal syndrome was made and it was decided to further confirm the diagnosis by renal biopsy. As P-ANCA became positive, it was decided that anti-glomerular basement disease could be excluded. Renal biopsy was performed by the Consultant Nephrologist and the microscopic appearance is shown in Figure 3.

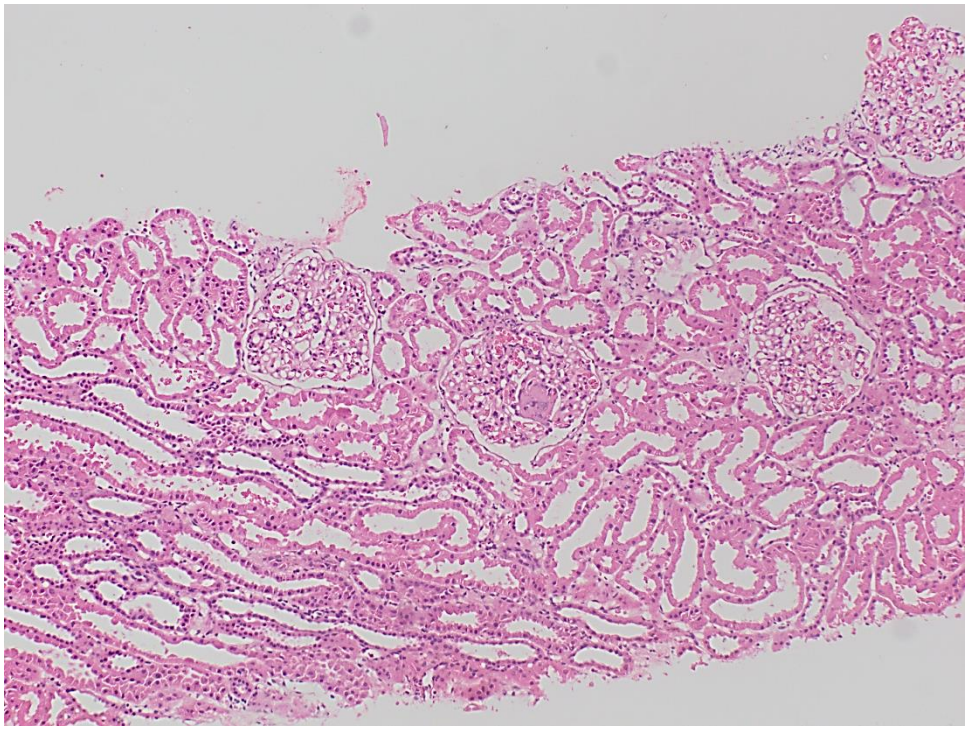

Figure 3: Renal biopsy showing segmental necrotizing glomerular nephritis consistent with pauci immune small vessel vasculitis

Progression to end stage renal disease can be prevented by early recognition through full renal work up in a patient with rheumatoid arthritis and appropriate management.

Conclusion

Acute kidney injury or acute tubular necrosis secondary to haemodynamic instability causes an elevation in serum creatine. If there is continuous rise in serum creatine even after adequate intervention, other causes have to be considered. A full renal work up is

recommended if there is a continuous rise in serum creatine even with adequate interventions such as removal of offending agents and adequate hydration. Serology, complement level measurement and urine and serum electrophoresis are included in the renal workup. If the findings of these are inconclusive, a renal biopsy should be considered. Pulmonary haemorrhage can manifest as a complication of pauci-immune crescentic glomerular nephritis in these patients. Patients who develop haemoptysis or have clinical examination changes, such as bilateral lung fine crepitations, need to undergo further work up and plasma exchange as a treatment. RA patients with ANCA-associated vasculitis (AAV) with antibodies against the myeloperoxidase (MPO) protein (MPO-AAV) will need long term steroids, cyclophosphamide and prophylaxis against *Pneumocystis jiroveci* pneumonia.

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