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A Rare Case of Pulmonary Renal Syndrome Diagnosed as Microscopic Polyangiitis

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Abstract: ***Microscopic Polyangiitis (MPA) is a systemic pauci-immune vasculitis primarily affecting glomerular capillaries, leading to necrotizing glomerulonephritis and often involving the kidneys and lungs. MPA is a significant cause of Pulmonary-renal syndrome, with a male predominance and typically presenting in individuals aged 50-60 years. Clinical manifestations include rapidly progressive glomerulonephritis characterized by a rapid decline in glomerular filtration rate, microscopic hematuria, erythrocyte casts, proteinuria, and hypertension, along with lung involvement such as diffuse alveolar hemorrhage. Although the pathogenesis remains unclear, environmental factors and genetic predisposition are believed to play roles. Diagnosis is confirmed histologically, and treatment involves remission induction and maintenance strategies using immunosuppressive therapies. A clinical case of a 45-year-old male with MPA presenting with shortness of breath, hemoptysis, and renal involvement highlights the disease's complexity and the importance of prompt, accurate diagnosis and treatment to improve patient outcomes.***

Keywords: Microscopic Polyangiitis, Pulmonary-renal syndrome, Glomerulonephritis, ANCA vasculitis, Immunosuppressive therapy

1. Introduction

Microscopic Polyangiitis (MPA) is a systemic pauci - immune vasculitis of glomerular capillaries leading to necrotizing glomerulonephritis. The kidney and lung are the most typical organ involved in MPA. Notably MPA is a major cause of Pulmonary - renal syndrome. MPA has slight male predominance (F: M - 1: 1.8) with an average age of onset of 50 - 60 years. The main clinical presentation in MPA is rapidly progressive glomerulonephritis (RPGN) characterized by rapid decrease of glomerular filtration rate (GFR), microscopic haematuria, erythrocyte cast, presence of proteinuria (usually less than 3g) and hypertension. Lung manifestations consist of diffuse alveolar haemorrhage due to pulmonary capillaritis.

The pathogenesis of MPA is unknown. There is increasing evidence that environment factors in association with a genetic predisposition are involved in the pathogenesis of MPA. Histological confirmation of vasculitis remains still the gold standard of the diagnosis of MPA. The therapy of MPA consist of remission induction strategies in order to achieve remission of the disease and remission maintenance strategies.

2. Clinical Case

A 45 year - old male presented to LG hospital, Ahmedabad, Gujarat with chief complain of shortness of breath since 1 week. Which was insidous in onset and gradually progressive associated with hemoptysis, 3 to 4 episodes in 3 days, around 40 to 50ml in quantity and weight loss. There is no complain of fever, palpitation, pedal edema and chest pain. No past history of diabetes mallitus, hypertension, ischemic heart disease, tuberculosis. There is history of an ulceration over the right leg 1 year ago which resolve by surgical debridement and heal with scar. No history of addiction. No significant Family history.

On examination patient was well alert and oriented to time, place and person. His vitals were taken which recorded a pulse rate of 76/min in the right radial artery and O2 saturation was 88% on room air and respiratory rate was 24/min. He was afebrile and Blood Pressure recorded was 116/74 mmHg. His respiratory examination revealed abnormal respiratory sounds - extensive crackles in bilateral lower lung fields. On physical examination over Right leg healing ulcer scar was present.

3. Diagnosis

The following investigations were done during the course of admission -

Hemogram

Investigations			
Hb	8	8.4	9.9
Tlc	9870	8700	11150
Apc	5.77	2.33	2.51
Rbc	3.12	3.71	3.84
Hct	27	27.5	31
MCV	84	85.5	81

LFT and RFT

Investigations			
S. urea	97	78	96
S. creat	2.25	2.14	1.68
Total Bilirubin/ Direct Bilirubin	1.8/0.9	1.2/0.4	1.6/0.5
SGPT	25	24	70
ALP	81	82	108
Na	134	141	139
K	4.7	4.8	4.3

- ESR - 82 mm/hr, CRP - 34.6 mg/dl
- Sputum AFB, CBNAAT, Cs, fungus were negative. H1N1 and covid was negative.
- Urine RM showed blood (+4), Albumin (+4), rbc (20 - 22) and absent pus cells.
- 24 - hour urine protein excretion was 1044mg.

- Chest X - ray film showed reticular shadows and loss of volume especially affecting the lower lung fields.
- HRCT Thorax: Diffuse confluent and scattered ground glass opacities in bilateral lung fields. Mild bronchiectatic changes in both lung fields. No pretracheal, paratracheal, hilar or paraspinal lymphadenopathy is seen. No pleural or pericardial effusion is seen.
- ECG and 2D echo was normal.
- USG KUB: Rt kidney: 92× 45mm, Lt kidney: 104× 40mm, Bilateral increased cortical echogenicity with preserved corticomedullary difference.
- Anti - ASO, IgM for atypical organisms, RA Factor, cANCA, ANA and viral markers were negative.
- p - ANCA with Myeloperoxidase antibody (MPO) of 134 AU/mL (normal< 26), negative Anti - PR3 and Anti - GBM and normal C3 and C4 levels.
- Renal biopsy: Immunofluorescence; IgA, IgG, IgM, C3, kappa, lambda were negative. HPE and IF; Pauci immune crescentic glomerulonephritis with one fibrocellular crescent.

History of hemoptysis with persistent without any identifiable cause of acute kidney injury lead to diagnosis of pulmonary renal syndrome. with high level of P ANCA with renal biopsy revealed active pauci - immune crescentic glomerulonephritis with no history of allergy and other features diagnosed as MPA.

Management:

After confirming diagnosis
Inj MPS (1gm) iv OD was given for 5 days followed by starting oral medication
T. prednisolone (20mg) 3 - 0 - 0
T. endoxan (50mg) BD

Following therapy Improvement in clinical condition and renal function was seen and patient was discharged and advised for regular follow up for to assess disease progression and monitor drug toxicity.

4. Discussion

Microscopic polyangiitis (MPA) is a form of ANCA vasculitis. ANCA vasculitis is caused by host - derived auto - antibodies against shielded neutrophilic antigens. These antibodies are suspected to react against granules present in neutrophils and monocytes.70% of MPA cases have a positive ANCA at the time of diagnosis. MPA has a variety of manifestations affecting many different organs; however most cases of MPA have been associated with renal and lung involvement. Most patients present with a rapidly progressive glomerulonephritis picture, which presents as a loss of renal function in days to weeks, a urine analysis that has protein and red blood cells, and histological findings on biopsy that shows crescent cellular formation on glomeruli. Treatment of MPA required immunosuppressive therapy like steroid, cyclophosphamide, mycophenolate mofetile, rituximab etc. Plasmapheresis has also been known to help those patients that have severe kidney disease, a creatinine > 4.0, and active vasculitis. After starting treatment it is important to access disease progression and drugs complication.

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