**A Case of Pauci-Immune Glomerulonephritis**

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**Patient Presentation**

- A 66 year old female with a history of hypertension presented to her primary care physician with symptoms of epistaxis, intermittent hematuria, dyspnea on exertion, and fatigue of two weeks in duration.
- Physical exam was remarkable for elevated blood pressure of 182/90mmHg and mild distress.
- Lab work significant for hemoglobin of 8.8g/dl, hematocrit of 25.2%, BUN of 63mg/dl, and creatinine of 6.0mg/dl. She had no prior findings of anemia or kidney dysfunction.
- Sent to the ED for the above findings.
- Urinalysis significant for hematuria, proteinuria, and dysmorphic RBCs.
- Renal ultrasound demonstrated loss of corticomedullary differentiation. Chest X-ray and CT abdomen pelvis unremarkable.
- Serum myeloperoxidase positive.
- Renal biopsy was obtained (Figure 1) and a diagnosis was made.
- Pathology report: Diffuse acute and early sclerosing necrotizing and crescentic glomerulonephritis consistent with pauci-immune ANCA associated glomerulonephritis; cellular crescents, fibrocellular crescents, and/or fibrinoid necrosis identified in approximately 45 of 61 glomeruli (74%).

**Background**

- Rapidly progressive glomerulonephritis (RPGN) is a nephritic syndrome characterized by rapidly worsening renal function with glomerular crescent formation on pathology.
- RPGN is broadly divided into pauci-immune glomerulonephritis (PIGN), anti-glomerular basement membrane GN, and immune-complex GN.
- Pauci-immune glomerulonephritis carries high morbidity and mortality with frequent progression to ESKD and premature death.

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**Hospital Course and Follow Up**

- On day four of admission patient became hypoxic and increasingly dysgenic, precipitating transfer to a facility with therapeutic plasma exchange capability.
- Received four sessions of TPE and started on cyclophosphamide at tertiary center, discharge on methylprednisolone in stable condition.
- Returned to tertiary center as an outpatient for plasmapheresis treatments, maintenance with steroid therapy complicated by Cushing’s syndrome.

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**Discussion**

- High dose glucocorticoids are effective in inducing remission in PIGN. Cyclophosphamide and rituximab are also used for induction.
- Plasmapheresis is indicated in instances of severe kidney injury and alveolar hemorrhage. Newer data suggests plasma exchange might not confer benefit to ANCA associated vasculitis patients, and that lower dose steroid regimens may be non-inferior[1].
- Maintenance therapy options include cyclophosphamide, methylprednisolone, azathioprine, mycophenolate mofetil, and rituximab.
- More data is needed regarding safe and efficacious management in these patients.

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**References:**