# Vision loss followed by multisystem organ failure: A complex case of pauci-immune vasculitis

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**Background**

Pauci-immune vasculitis is a rare disease state with a wide range of systemic manifestations and is the most common cause of rapidly progressive glomerulonephritis. This disease is frequently associated with ANCA positivity and the type and antibody can be associated with more specific clinical syndromes. This case reports one such example as a hallmark educational case.

**Case Summary**

- 69-year-old female
- PMH of hypertension, GERD, and pulmonary node who presented with painless atraumatic vision loss in the right eye after referral from her ophthalmologist. Additionally noted malaise, 30-lb weight loss over three months, headaches, and jaw pain.
- On presentation, she was found to be in acute renal failure with elevated inflammatory markers.
- Initial concern was for temporal arteritis and patient was started on high dose steroids, temporal artery biopsy was negative.
- Rheumatologic evaluation and renal biopsy were consistent with pauci-immune vasculitis/glomerulonephritis and serology showed p-ANCA positivity.
- Patient was initiated on immunomodulatory therapy with rituximab.
- Had possible cardiac arrest vs vasovagal event which she received CPR for and returned to baseline.
- She subsequently developed respiratory failure during her stay and new onset HFrEF, consistent with Takotsubo cardiomyopathy. She underwent medical optimization without invasive cardiac testing during hospitalization.
- Is on waitlist for renal transplant otherwise has remained stable without further vision loss and heart function returned to baseline.

**Hospital course:**

- **6/22/20:** Transfer from nearby hospital for painless right eye vision loss, elevated BUN/Creatinine, malaise, 30 lb weight loss over 3 months, headaches, jaw pain. Started on IV steroids due initial concern for Giant Cell Arteritis.
- **6/23/20:** Right Temporal Artery Biopsy completed and was negative.
- **6/24/20:** Kidney Biopsy completed and rheumatologic evaluation consistent with Pauci-immune vasculitis/glomerulonephritis and serology showing p-ANCA positivity
- **6/26/20:** Patient's renal function continued to worsen despite IV fluids, started on dialysis and plasmapheresis. Nephrology, Hematology-Oncology following.
- **6/27/20:** 2nd plasmapheresis infusion given.
- **6/28/20**: 1st infusion of Rituximab given, will receive 3 more Rituximab infusions weekly.
- **7/2/20**: Overnight 07/1-07/2 patient was hypoxic requiring oxygen. CXR showing pulmonary vascular congestion and interstitial edema. Plan for dialysis because of fluid overload. Later in the day during dialysis patient had cardiac arrest vs vasovagal event for which she received CPR for and returned to baseline shortly. Transferred to ICU for closer monitoring.
- **7/3/20:** Overnight 7/2-7/3 patient's Hgb 6.9, transfused 2 units pRBCs and given DDAVP. Did have right perinephric hematoma which could have contributed to anemia. Downgraded to floors.
- **7/4/20:** Rapid Response for hypoxia. Diuresis with Bumex.
- **7/5/20:** Transferred back to ICU due to hypoxia, placed on BiPAP.
- **7/6/20:** CTA negative for PE, showing pulmonary edema. Thoracentesis completed. Echo showing EF 25-30% concerning for takotsubo cardiomyopathy. Cardiology consulted. Second dose of Rituximab received.
- **7/7/20**: Per Cardiology patient had Takotsubo vs obstructive CAD. Started on beta blocker.
- **7/9/20:** Patient weaned off oxygen s/p thoracentesis, was transudative. Downgraded from ICU to floors.
- **7/13/20:** Third dose of Rituximab given
- **7/20/22:** 4th dose of Rituximab given, patient discharged on oral prednisone to follow-up with Nephrology, Cardiology, Hematology-Oncology, Ophthalmology as an outpatient.

**Epidemiology and Clinical Outcomes**

- Represents 80% of RPGN, 7-10 cases per million people per year in the US.
- Predilection for whites compared to blacks, equal representation in men and women.
- Without treatment, 1 year mortality rate of 80%.
- With aggressive immunosuppression 5-year survival rate 75%.

**Pathophysiology**

- ANCA binds target antigens PR3 and MPO activates neutrophils and monocytes.
- Production of ROS and elastase injurious to endothelial cells.
- Release of NETs (neutrophil extracellular traps) damaging endothelial cells and glomeruli.

**Diagnosis**

- Generalized non-specific symptoms like fever, malaise, anorexia, weight loss, myalgias.
- Asymptomatic hematuria to RPGN.
- Most cases, elevated creatinine in combination with active urine sediment.
- Kidney Biopsy showing fibrinoid necrosis with crescent formation.

**Management**

- Without vital organ involvement or life-threatening disease glucocorticoids + methotrexate or Rituximab.
- With vital organ involvement or life-threatening disease glucocorticoids + cyclophosphamide +/- Rituximab.
- Add plasma exchange if pulmonary hemorrhage, RPGN, or Ant-GBM antibody positive.

**Conclusion**

- Pauci-immune vasculitis can have devastating effects on multiple organ systems, and thus it's important to recognize early symptoms to diagnose and treat properly to potentially limit morbidity and mortality.

**References**

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