

AORTIC ANEURYSM AND PR3ANCA VASCULITIS

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62 year old Caucasian male with history of hypertension and rheumatoid arthritis and normal renal functions at baseline presents with back pain and reduced urine output for 4 days. Review of systems was positive for smoking history and recent hemoptysis. Examination revealed BP140/90mm Hg, Heart rate 125/minute with no signs of fluid overload. Labs showed pH 7.28, Hb11.5g/dl, potassium of 7mmol/l, Blood urea (BUN) 145mg/dl, Creatinine (Cr) 14.5mg/dl, Urine protein/creatinine ratio 4 in oliguric urine and many RBCs in urinalysis. Ultrasound showed normal sized kidneys. Echocardiogram showed normal Left ventricular size and function. Bilateral ground-glass infiltrates were seen in CT chest. CT scan of abdomen showed infra-renal abdominal aortic aneurysmal leak that was repaired with stent graft. Renal functions worsened on day 2 with BUN 148, Cr 14.8 and started on hemodialysis (HD). Found to be C-ANCA /PR3ANCA positive at 94(normal <6U/ml) and pulse doses of high dose steroids was started for the possible diagnosis of ANCA vasculitis. Patient subsequently underwent a renal biopsy. Biopsy revealed severe findings of pauci-immune necrotizing and crescentic glomerulonephritis typical of Wegener's granulomatosis (WG). Also associated with extensive capillaritis, interstitial hemorrhage and tubular necrosis which is a manifestation of PR3-ANCA associated disease. Aggressive management was continued with plasmapheresis and oral cyclophosphamide at dose 2.5mg/kg. Patient was discharged on oral steroids and cytoxan with outpatient HD. As the aneurysmal repair was done much earlier when the rest of the renal workup was pending, pathologic sampling of the aorta was not obtained.

We strongly suspect that the aortic aneurysm in our patient is caused by PR3ANCA vasculitis due to the acute presentation of various symptoms and clinical findings that can be clustered into a single diagnosis. WG is known to mostly affect only small and medium sized vessels. Few case reports are reported in literature of WG induced large vessel vasculitis leading to periaortitis, aortic aneurysm and dissection. Progressive renal failure is commonly observed in patients with WG. End-stage renal disease occurs in approximately 20 to 25% of patients.