A case of Wegener Granulomatosis which has relapsed with only eye involvement

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INTRODUCTION. Wegener's granulomatosis (WG) is a disease which is characterized by multisystemic involvement including glomerulonephritis together with necrotizing granulomatous vasculitis of the upper and lower respiratory pathways. Eye involvement is common but retinal vasculitis has rarely been described. Herein we will present a case with the diagnosis of WG which developed retinal vein occlusion in its clinical follow up.

CASE. Seventy-six-year-old female patient with a diagnosis of hypertension was admitted to the emergency department because of impaired state of well being, left leg numbness and widespread joint pain. The patient's blood pressure was 210/140 mmHg and her laboratory test results were as follows; serum urea: 134 mg/dl, creatinine: 5.3 mg/dL, total protein: 5.6g/dl, albumin: 2.3g/dl, Hb 7.7 g/dL, hematocrit: 36% leukocytes: 19600/mm^3, platelet 466000/mm^3 sedimentation: 115mm/hr, CRP: 17, ANA (-), Antids-DNA (-), c-ANCA (+; 66.51 U / ml) RF (-) IgG: 1360 mg/dL, IgA: 415mg/dl, IgM: 41mg/dl, C3: 136 mg/dL (79 to 152), C4: 28mg/dl (16-38), Urinalysis; density: 1000, protein +, microscopy. 20 erythrocytes, 18 leukocytes were detected and 1375 mg/L/day proteinuria was present in 24-hour urine. The length, parenchymal thickness and echogenicity of right and left kidney were 109x42 mm, 15 mm, grade 1 and 103x46 mm, 12 mm, grade 2 respectively. There was no renal stenosis on Doppler ultrasound examination. Millimetric bilateral nodules and mediastinal lymph nodes within the upper limits of normal were observed in CT imaging. The patient who was consulted to Rheumatologist 2013 were diagnosed as WG in January 2013. Paucar-immune cresentric glomerulonephritis was detected with kidney biopsy. Patient treated with high doses of steroids, cyclophosphamide, and 4 sessions of plasmapheresis due to the rapidly progressive glomerulonephritis. However, our patient appeared to need hemodialysis and continuous dialysis has become necessity. Renal

replacement therapy (RRT) needed 3 times a week. After receiving chronic hemodialysis program cyclophosphamide was suspended and corticosteroid therapy was tapered and discontinued.. In November 2013, the patient has evolved headache and bilateral visual loss. The patient was assessed by Ophthalmologists and central retinal venous thrombosis and multiple hemorrhages, narrowing in the arteries, veins, thickening, exudates, and capillary proliferation was detected in corneal endothelium. Temporal artery Doppler ultrasonography and cranial MRI was reported as normal. After Rheumatology consultation patient was evaluated as recurrence of WG with retinal vasculitis.

CONCLUSION. This case report showed that recurrence might be encountered with ocular involvement in patients with WG. It should be kept in mind that early termination of the immunosuppressive therapy increase the risk of relapses in different tissues and organs.

KEYWORDS: Wegener's granulomatosis, retinal vasculitis, relapse