Focal Segmental Glomerulosclerosis in a Patient with Polycythemia Rubra Vera

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Dear Sir,

We report a case of focal segmental glomerulosclerosis (FSGS) with polycythemia rubra vera. The relationship between polycythemia and a renal lesion is discussed. To the best of our knowledge, polycythemia vera in association with FSGS has not been reported so far. A 40-year-old female presented with throbbing headache, giddiness, visual blurring, chest pain and off and on facial flushing of 2 years duration. While undergoing investigations for surgery for a thyroid nodule, the patient was found to have a high hematocrit (Hb 18 g%) count. At that time urine examination did not reveal any proteinuria. One year later she was found to be hypertensive and urine examination showed significant proteinuria (2.0 g in 24 h). On examination the patient was plethoric with no facial or pedal edema, blood pressure was 160/100 mm Hg and the spleen was enlarged. Fundus examination revealed a hyperemic disc and tortuous vessels. Hb was 20.0 g%, hematocrit (Hct) 65%, red blood cell (RBC) count $62 \times 10^5/\text{cm}^3$, total leukocyte count (TLC) $32 \times 10^3/\text{cm}^3$, platelets $4.1 \times 10^5/\text{cm}^3$ and the erythrocytic sedimentation rate was 7 mm in the 1 h. Bone marrow was hypercellular and showed erythroid hyperplasia with increased megakaryocytes. ^{51}Cr RBC volume was 38 ml/kg (normal 27-32 ml/kg). Urine examination showed 10-12 RBC/high power field, 6-8 white cells/high power field. Urinary protein excretion was 1.8 g in 24 h. Blood urea nitrogen was 18 mg%, serum creatinine 2.0 mg%, serum uric acid 5.6 mg%, serum albumin 3.2 g% and serum cholesterol 230 mg%. Renal ultrasonography showed normal sized kidneys. Arterial blood gas analysis showed PO_2 93.6 mm Hg and oxygen saturation of 96.5%. Kidney biopsy was done which showed 15 glomeruli with focally accentuated mesangial proliferation and a mild increase in mesangial matrix, 5 glomeruli showed segmental areas of sclerosis and hyalinosis with synechia formation. Focal areas of tubular atrophy and mild interstitial fibrosis were seen. Blood vessels displayed intimal fibroelastosis. Biopsy was consistent with FSGS.

The patient was initially subjected to phlebotomies. She showed symptomatic improvement. Hb decreased to 12.8 g%, Hct 37%, TLC 15,000/cm, platelet 4.5×10^5 /cm. Serum creatinine decreased to 1.4 mg%; however, proteinuria persisted (1.6 g/24 h). Subsequently she was given myelosuppressive therapy with ^{32}P . The hematological profile after 2 years of follow-up and 3 phlebotomies sittings and 3 doses of ^{32}P therapy is: Hb 13.0g%, Hct 39%; TLC 9×10^3 /cm, and platelets 2.4×10^5 /cm. Serum creatinine is 1.4 mg% and urinary protein excretion is 0.24 g in 24 h.

Our patient fulfils the criteria proposed by the polycythemia vera study group for labelling it as a case of polycythemia vera. Significant proteinuria was an important presenting feature and renal histology revealed FSGS. Symptoms due to polycythemia vera were present for at least 2 years before the detection of proteinuria. At the initial detection of polycythemia there was no proteinuria which

appeared a year later. The occurrence of a glomerular lesion in such a situation would be secondary to polycythemia vera.

Renal hemodynamic alterations in polycythemia vera (renal vasodilation, increased effective renal blood flow) could result in FSGS as in any other nonprimary FSGS [1]. Plomely et al. [2] described 3 patients with histologically defined glomerulonephritis in polycythemia vera, all of them had diffuse mesangial proliferation with proteinaceous deposits, focal tubular atrophy with interstitial fibrosis and arteriolar hyalinization.

In our case, serum creatinine and proteinuria decreased with the normalization of hematocrit. We have not rebiopsied the patient, but in view of the reduction in proteinuria, we expect that the histological changes might also show resolution. Long-term follow-up of our case would be required to show whether this proteinuria remission indicates resolution of the renal lesion and good prognosis.

References

Rennke HG, Klein PS: Pathogenesis and significance of non-primary focal and segmental glomerulosclerosis. *Am J Kidney Dis* 1989; 13: 443-456.

Plomely RF, Sullivan JR, Whitworth JA, Kincaid-Smith PS, Fairly KF, Brown PW: Polycythemia vera and glomerulonephritis. *Aust NZ J Med* 1983;13:125-129.