Essential Thrombocythemia Presenting as Paraneoplastic Glomerulonephritis

Dear Sir,

A 44-year-old male was admitted with reduced urinary output and bilateral pedal edema for 1 week. He had bipolar affective disorder for the last 14 years but was not receiving any medications. Examination showed facial puffiness, bilateral pedal edema, and moderate splenomegaly. Hemoglobin was 11.3 g/dl (normocytic, normochromic), total white blood cell count 23,600/μl, platelet count 7.5×10^9/L, and erythrocyte sedimentation rate 68 mm in 1 h. Peripheral smear showed normocytic normochromic red blood cells with mild leukocytosis and thrombocytosis. He had nephrotic range proteinuria, hypoalbuminemia, and moderate renal failure. Renal biopsy revealed focal segmental glomerulosclerosis [Figure 1 lower panel]. Bone marrow biopsy showed a cellular marrow with megakaryocyte hyperplasia and areas of megakaryocyte dyspoiesis [Figure 1 upper panel]. HIV, hepatitis B, and hepatitis C serologies were negative. Antinuclear antibody was negative. The Janus kinase 2 gene (*JAK2*) *V617F* mutation was positive, but BCR/ABL fusion gene was negative. He was diagnosed to have essential thrombocythemia with paraneoplastic focal segmental glomerulosclerosis and was treated with high-dose steroids, hemodialysis, and hydroxyurea. His renal failure stabilized with treatment and he was continued on hydroxyurea.

Glomerular diseases are associated with many solid and hematologic malignancies. Paraneoplastic glomerular lesions are most likely due to abnormal products produced by tumor cells although the exact pathogenesis is unclear. The treatment of these cancer-associated glomerulopathies is primarily targeted at treating the underlying malignancy.

Myeloproliferative disorders include chronic myelogenous leukemia, polycythemia Vera (PCV), and essential thrombocythemia. Among the myeloproliferative neoplasms, PCV, essential thrombocythemia, and primary myelofibrosis are associated with focal segmental glomerulosclerosis. Glomerular disorders associated with myeloproliferative disorders are usually late complications and tend to have a poor renal prognosis. Failure to recognize paraneoplastic glomerulonephritis can lead to undue delay in initiating anticancer medications.

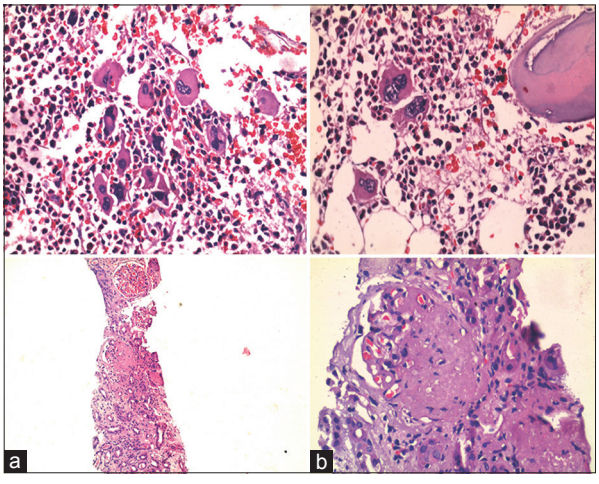

Figure 1: Upper panel – bone marrow trephine biopsy showing cellular marrow with megakaryocyte hyperplasia and megakaryocyte dyspoiesis (H and E, ×40). Lower panel – renal biopsy core with 1/11 glomeruli showing features of focal segmental glomerulosclerosis lower panel a (H and E, ×10). Glomerulus with segmental sclerosis with adhesion to the capsule lower panel b (H and E, ×40)

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Conflicts of interest

There are no conflicts of interest.

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