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AN UNUSUAL PRESENTATION OF RAPIDLY PROGRESSIVE GLOMERULONEPHRITIS IN A PATIENT WITH SICKLE CELL DISEASE

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ABSTRACT

We present an unusual presenting manifestation of sickle cell disease admitted with anuria. On evaluation, he was found to have rapidly progressive renal failure. Renal biopsy was suggestive of pauci immune crescentic glomerulonephritis. Despite the measures of immunosuppresssion, patient continued to be dialysis depndent. Crescentic glomerulonephritis is rare in sickle cell disease.

INTRODUCTION

Renal manifestations of SCD include proteinuria, hematuria, concentrating and diluting dysfunctions, papillary necrosis and chronic kidney disease. Amongst the causes of proteinuria, FSGS accounts for 10-20% of cases followed by non immune MPGN.[1] Immune MPGN is rare which was reported in only 12 cases in literature.[2] Crescentic GN in SCD is rare Herein, we report a patient with rapidly progressive glomerulonephritis with crescents on renal biopsy.

CASE DETAILS

A 30 years aged man presented to the emergency department with sudden onset of anuria of 15 days duration. History of pedal edema and breathlessness were present. There was no history of fever, nephrotoxic drug ingestion immediately preceding the onset of these symptoms. He was evaluated elsewhere and was given blood transfusion in view of anemia. At admission on examination, he had tachypnea, tachycardia, BP was 130/80 mm of Hg. Pallor and pedal edema were present. Bilateral crepitations were present in lungs. On evaluation, his hemoglobin was 7.5gm/dl, total WBC was 17200cells/mm3, platelets of 6.1lakhs/mm3. Peripheral smear showed moderate anisopoikilocytosis with microcytes, fragmented RBC, sickle cells. His reticulocyte count was 2.5% and immediate sickling test was positive. His complete urine examination showed albumin3+, RBC 10-15/hpf. Serum levels of C3 was 68mg/dl and C4 was 6mg/dl. Serology for ANA and antidsDNA were negative. Liver function tests showed serum bilirubin of 2.1mg/dl and direct bilirubin was 1.2 mg/dl with enzymes being within normal limits. Ultrasound abdomen showed normal sized kidneys. Chest xray and ECG were normal. His blood urea was122mg/dl and serum creatinine was 6mg/dl. He was initiated on hemodialysis and was placed on supportive treatment along with course of antibiotics. Inview of severe renal insufficiency with active urine sediment and hypocomplementemia, a possible diagnosis of rapidly progressive glomerulonephritis (RPGN) was considered. Renal biopsy showed 10 glomeruli of which 8 glomerulii had circumferential cellular crescents. Endocapillary proliferation with lobular accentuation, thickening of basement membrane along with splitting were also present. IFTA was 40% and interstitium showed scattered infiltrates of lymphocytes (fig 1,2).On Immunofluorescence, there was weak C3 in mesangium and capillaries. A diagnosis of crescentic MPGN was considered. Serology for ANCA was negative. Patient was given parenteral methylprednisolone followed by oral steroids and cyclophosphamide. Despite the immunosuppression, he remained anuric. At the end of 2 months follow up he continued to be dialysis dependent.

DISCUSSION

Renal manifestations in sickle cell disease include proteinuria due to underlying glomerulopathy, hematuria, acute kidney injury and chronic kidney disease, urinary concentrating defects, abnormalities in urinary acidification, potassium excretion and supranormal proximal tubular function, renal tubular acidosis, interstitial fibrosis, tubular hemosiderosis and rarely renal medullary carcinoma.[1-3,6] Sickle cell nephropathy has been well described in literature which includes both the glomerular and tubular abnormalities in patients with SCD.

Glomerular manifestations in SCD include FSGS, nonimmune MPGN, post infectious glomerulonephritis, minimal change disease and amyloidosis.[2,4] Rarely

immune complex MPGN and recently a case of IgA nephropathy presenting with isolated hematuria was also reported.[5] However, crescentic GN is rare and has been reported in 22 year aged postpartum woman, a known case of MPGN with crescentic transformation.[7] But SCD presenting as with rapidly progressive glomerulonephritis with biopsy proven crescentic MPGN has not been reported.

Paucity of literature exists regarding MPGN in cases of SCD. Though nonimmune MPGN is common in SCD, immune MPGN is rare. Immune complex MPGN was reported in only 12 cases till now in literature.[7] Nonimmune MPGN is thought to be secondary to the fragmentation of RBC in the capillary loops, which are further phagocytosed by the mesangial cells. This is followed by laying of new basement membrane beneath the capillary endothelium by the active mesangium.[7] Experimentally, similar changes were observed in rats due to underlying thrombotic microangiopathy.

Crescentic MPGN is itself a rare entity. Small focal crescents are commonly seen with MPGN but crescents in >50% of glomeruli is very rare in MPGN. Sharma etal reported crescentic MPGN developing in the course of MPGN in a 22 years aged young woman. D'Amico and Ferrario described six variants of MPGN, namely classic, nodular, exudative, focal segmental, MPGN with massive deposits and crescentic MPGN. Crescentic MPGN was reported in renal grafts more often than in native kidneys. Few reports of crescentic MPGN were reported with hepatitis B, hepatitis C, and urticarial vasculitis.

In our patient, crescentic MPGN was reported. His serum for ANCA was negative. He was given immunosuppression with steroids and cyclophosphamide, but there was no improvement in renal function causing the patient to be dialysis dependent. A similar case of crescentic MPGN in a known case of MPGN was reported in postpartum woman where in the patient improved with the treatment.

Points of interest in this patient are two fold

1. Occurrence of crescentic MPGN in a case of sickle cell disease.
2. Crescentic MPGN may be the first clinical manifestation of Sickle cell disease.

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