Idiopathic erythrocytosis in IgA nephropathy

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ABSTRACT

We report a case of idiopathic erythrocytosis in a 31-year-old male who was incidentally detected to have hypertension during his preemployment checkup. Urine routine showed proteinuria and hematuria. Biochemical parameters revealed raised serum creatinine, and histological findings of the renal biopsy showed IgAN.

Key words: Erythrocytosis, IgA nephropathy, nephrotic syndrome, polycythemia

Introduction

IgA nephropathy (IgAN) is one of the most frequent forms of glomerulonephritis (GN). There have been several case reports citing polycythemia vera in IgAN. Polycythemia can be associated with IgAN, focal segmental glomerulosclerosis and membranoproliferative GN. Here, we report a case of idiopathic erythrocytosis in a 31-year-old male who was incidentally detected to have hypertension during his pre-employment checkup. Urine routine showed proteinuria and hematuria. Biochemical parameters revealed raised serum creatinine, and histological findings of the renal biopsy showed IgAN.

Case Report

A 31-year-old male was referred to nephrology in view of hypertension, renal dysfunction, proteinuria, and active urinary sediments. His clinical examination revealed evidence of bilateral pitting pedal edema and blood pressure (BP) of 160/110 mmhg. His systemic examination was normal. There was no history suggestive of polyarthralgia, rash, skin ulcers chronic cough and drug intake. Complete blood count revealed hemoglobin of 19.2 g/dl, normal total count, and platelet count. Serum creatinine was 2.2 mg/dl and uric acid 10.6 mg/dl. The 24 h urine protein excreation was 7.5 g/day. Possible causes of secondary erythrocytosis were ruled out. Serum erythropoietin levels were normal and janus kinase 2 exon mutation was negative. The liver function, uric acid, electrolytes, glucose, complement 3 (C3), and complement 4 (C4) serum levels were normal. Coagulation and the levels of antistreptolysin O and high-sensitivity C-reactive protein were also normal. Antinuclear antibody profile was done, and antibodies to anti-myeloperoxidase, proteinase 3, double-stranded DNA, nucleosome and Sjögren's syndrome type A (SS-A) and SS-B antibodies tested negative. Ultrasound revealed normal-sized kidneys with right kidney of 9.6 cm and left kidney measured 9.4 cm. Radioisotope scan revealed a glomerular filtration rate (GFR) of 78 ml/min with a right and left differential of 33 ml/min and 35 ml/min, respectively. Bone marrow biopsy was normal. Renal biopsy showed chronic IgA nephropathy (IgAN), with M, mesangial hypercellularity; E, endocapillary proliferation; S, segmental glomerulosclerosis/adhesion; T, tubular atrophy/interstitial fibrosis (MEST) scoring of M1, E0, S1, T1.

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The patient was treated with two sittings of phlebotomy, started on amlodipine 5 mg and ramipril 2.5 mg for control of hypertension along with allopurinol 100 mg and aspirin 150 mg for hyperuricemia and polycythemia, respectively. He was followed up every 2 weeks on outpatient basis. At the end of 8 weeks, his blood pressure was within normal limits.

Discussion

To the best of our knowledge, there have been 24 cases of polycythemia associated with renal disease described so far and out of them only ten have been associated with IgAN.[1-6] The other renal lesions associated with polycythemia were focal segmental glomerulosclerosis, membranoproliferative glomerulonephritis, and rapidly progressive glomerulonephritis. Furthermore, of the ten cases of polycythemia associated with IgAN described so far, all happen to be males. Nearly 66% of presented with nephrotic syndrome and 34% had subnephrotic range of proteinuria. Only three cases reported to have normal renal function whereas others had either moderate to severe renal dysfunction. All the ten patients had hypertension, and three cases had hyperuricemia.[6]

Several cytokines and growth factors play a role in the progression of renal disease in polycythemia; one such report demonstrated abnormally upregulated mRNA expression of platelet-derived growth factor and insulin-like growth factor. Insulin-like growth factor has been implicated in exacerbation of polycythemia.[7] Furthermore, the increased blood viscosity and blood volume leads to vascular microthrombi and glomerular capillary occlusion thus reducing the GFR. Capillary occlusion leads to ischemia which eventually leads to chronic renal damage if not reversed on time. Interferon therapy used in polycythemia has also been reported to cause renal damage.[8]

This is the 11th case of polycythemia associated with IgAN being reported and this is the first case of idiopathic erythrocytosis being associated with IgAN. Striking similarities with other similar case reports are the preponderance in males (all ten case reports being males) and the presence of hyperuricemia, hypertension, and renal failure.

Conclusion

There has been an increased presence of IgAN worldwide and it has become one of the leading causes of renal failure in young adults. Hypertension and active urinary sediment is a common finding in most of these individuals, what makes this case report rare is the presence of idiopathic polycythemia as an additional cofactor. As only nine such reports have been made so far, it is essential to dig further into the pathogenesis of polycythemia in these individuals to know more about this causal relationship.

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Conflicts of interest
There are no conflicts of interest.

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