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Rapidly Progressive IgA Nephropathy Leads to End-Stage Renal Disease: A Case Report

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Abstract:- IgA nephropathy (IgAN), also known as Berger's disease, is one of the leading causes of chronic kidney failure and end-stage renal disease ESRD. It is a disease characterized by depositions of IgA in kidneys leading to inflammation in the glomeruli of the kidneys which progress to cause kidney damage. The Cases of rapidly progressing renal failure caused by IgAN are relatively rare. This case report is about a 38-year-old Asian male who developed a rapidly progressive IgA nephropathy which has led to end-stage renal disease (ESRD).

Keywords:- *IgA Nephropathy, ESRD, Case Report.*

I. INTRODUCTION

Immunoglobulin A nephropathy (IgAN), also known as Berger's disease, is one of the leading causes of chronic kidney failure and end-stage renal disease ESRD [1]. It is a disease manifested by depositions of IgA in kidneys leading to inflammation in the glomeruli of the kidneys which progress to cause kidney damage [2]. An important finding in patients with IgAN is the presence of glomerular and circulating immune complexes. The progression of renal damage resulting from IgAN is usually slow and develops over several months to years. Cases of rapidly progressing renal failure caused by IgAN are relatively rare.

II. CASE PRESENTATION

A 38-year-old Asian male having no known medical history presented with scrotal swelling and symmetrical limb swelling for two weeks. During the same time course, the patient had had few episodes of nausea and vomiting and was experiencing unusual fatigue. The patient denied the presence of fever, chills, skin rashes, changes in the characteristics of urine, or travel history. However, he had a history of a recurrent sore throat for the last two months. He was not using any OTC medications and was not a smoker with no history of tobacco use or alcohol consumption. There was no known medical history in the patient's family except for the hypertensive mother.

The physical examination of the patient during the admission to the hospital was significant for a high blood pressure reading of 190/95 mmHg while all other vital signs were within the normal values. He was presented with 3+ symmetrical lower extremity edema which was extending up to the thigh. There was no renal bruit, pharyngeal exudate, or anomalous skin rash.

The laboratory test results are presented in tables (1-3) below. The dipstick urinalysis was found to be significant for large blood, >900 mg/dl of protein, and 10-15 red blood cells (RBCs) on microscopy. Urine studies showed a protein-to-creatinine ratio of 5.26 g/dL, i.e., nephrotic range proteinuria. As a result of this indicator, several tests were done to figure out the etiology for the nephrotic syndrome. The patient's thrombophilia profile was within the normal limit; however, cholesterol level was found to be elevated (table. III) and the patient had hypoalbuminemia. (table. I). Hypogammaglobulinemia was indicated by serum and urine protein electrophoresis with no abnormal protein spike. Both Anti-nuclear antibody (ANA) and anti-glomerular basement membrane antibody (anti-GBM) tests were negative and anti-streptolysin O (ASO) titers were unremarkable. Hepatitis and HIV serology was negative. Cytoplasmic-antineutrophil cytoplasmic antibody (C-ANCA) and perinuclear antineutrophil cytoplasmic antibody P-ANCA antibodies were also negative. The value of Glycosylated hemoglobin (HbA1C) level was 3.1% and is considered normal.

When hospitalized, the patient was given nicardipine drip to bring back blood pressure to normal, and later he was switched to oral anti-hypertensive medication including isosorbide dinitrate, carvedilol, hydralazine, and diltiazem. Urine output during the whole hospitalization period remained less than 500 ml/day, i.e., he was oliguric. Furosemide was also administered to the patient but the urine output was not improved; however, edema of the extremity improved slightly. The consultant nephrologist suspected the presence of IgAN based on the physical and clinical presentation of the patient. The patient was scheduled for a renal biopsy to identify the cause of the nephrotic syndrome and to confirm/exclude IgAN. Intermitted hemodialysis (HD) was also started because creatinine level raised to 11.4 mg/dl with blood urea nitrogen (BUN) to 90 mg/dl. Edema significantly improved with HD and uremia was also resolved. Erythropoietin stimulating agents (ESA) along with intravenous iron were used to treat anemia presented by a hemoglobin level of 8.1 g/dl.

Renal biopsy findings revealed that the specimen had 19 globally sclerotic glomeruli, four additional glomeruli per

level of the section had segmental sclerosis, and three partial cellular crescents. Endocapillary hypercellularity was present as well. In the mesangium, the immunofluorescence assays confirmed the presence of 3+ diffuse fine granular staining for IgA and 2+ staining for IgM and C3. Based on the biopsy findings and clinical presentation, the diagnosis was made as rapidly progressive IgAN (2009 Oxford classification M1, E1, S1, T2) [4].

The medication chart on discharge comprised a beta-blocker, angiotensin-receptor-blocker, nondihydropyridine calcium-channel blocker, and hydralazine to control high blood pressure. High-intensity statin therapy was given for the management of hyperlipidemia. Given the ESRD, the patient was scheduled for hemodialysis thrice a week and he was suggested to meet a transplant nephrologist for the evaluation of possibilities of kidney transplantation in the future.

TABLE I. SERUM CHEMISTRY

Test name	Test result	Reference range
Sodium (Na)	137 mmol/L	135-145 mmol/L
Potassium (K)	4.1 mmol/L	3.4-4.8 mmol/L
Chloride (Cl)	105 mmol/L	96-106 mmol/L
Bicarbonate (CHO3)	27 mmol/L	21-28 mmol/L
Blood urea nitrogen (BUN)	54 mg/dL	10-20 mg/dL
Creatinine (Cr)	8.7 mg/dL	0.6-1.2 mg/dL
Albumin	2.4 g/dL	3.5-5.5 g/dL
Total protein	5.1 g/dL	6.4-8.3 g/dL

TABLE II. COMPLETE BLOOD PICTURE (CBP)

Test name	Test result	Reference range
Hemoglobin (Hb)	8.1 g/dL	12-16 g/dL
Platelets (Thrombocytes)	171 × 10^3/µl	150-450 × 10^3/µl
White blood cells (Leukocytes)	7.8 × 10^3/µl	4.5-10.5 × 10^3/µl
Red blood cells (Erythrocytes)	5.1 × 10^6/µl	4.7- 6.1 × 10^6/µl
Mean corpuscular volume (MCV)	98.1 fL	78-102 fL

TABLE III. LIPID PROFILE

Test name	Test result	Reference range
Cholesterol	215 mg/dL	<200 mg/dL
Low density lipid (LDL)	114 mg/dL	<130 mg/dL
Triglycerides (TG)	106 mg/dL	<150 mg/dL
High density lipid (HDL)	55 mg/dL	>45 mg/dL

III. DISCUSSION

IgA nephropathy has been described as being the top cause of glomerulonephritis and one of the leading causes of chronic kidney failure and end-stage renal disease ESRD [1,2,4]. Though it is a fact that IgA deposition with a predominance of Lambda light chains takes place in the glomerular mesangium, the pathophysiology of IgAN is not fully understood [5,6]. Patients with IgAN usually present in two scenarios. One is presenting with gross hematuria after an upper respiratory tract infection (URTI). The other form is presenting with hematuria along with a varying degree of proteinuria [7]. Nephrotic range proteinuria, thrombotic microangiopathy, and malignant hypertension might be present in severe and rapidly progressing cases of IgAN [8]. Performing renal biopsy is crucial in staging and classifying the disease according to Oxford Classification. It is also beneficial in determining the prognosis of the disease at the time of biopsy [4,8]. Other clinical findings and laboratory results that can help in determining the severity of the disease include high serum creatinine (Cr), reduction in glomerular filtration rate (GFR), persistent proteinuria, and hypertension (blood pressure > 140/90 mmHg) [9]. The presence of pathological findings of crescents, tubular atrophy, interstitial fibrosis, glomerular and/or segmental sclerosis, and interstitial cellular infiltrates on renal biopsy is an indicator of worsening state of the disease and high risk of developing ESRD as seen in this case [10].

Management of "IgA nephropathy with crescentic glomerulonephritis has not been studied in randomized controlled trials and there have been only some observational studies for the treatment and management of it" [11]. The use of "pulse intravenous (IV) methylprednisolone to be followed by oral prednisone, IV cyclophosphamide, or plasmapheresis" was suggested by the observational studies [11]. However, the patient, in this case, had already developed ESRD by the time of diagnosis and hence was not given any trial of these therapies. Other treatment options in IgAN include "the use of angiotensin-converting enzyme inhibitors (ACE-I) or angiotensin receptor blockers (ARB)" to achieve better blood pressure control and proteinuria [12]. Regular assessment of serum Cr, GFR, urinary sediment, and proteinuria can be performed to monitor disease severity.

IV. CONCLUSION

IgAN affects and damages the kidneys and can progress to ERSD. Though the progression to ESRD occurs over a long period, rapidly progressive IgAN can lead to ESRD over a relatively short time course. This case report presents a typical case of rapidly progressing IgAN in which the patient had already developed ESRD by the time of diagnosis. Early diagnosis and treatment are of key importance in the prevention of progressive forms of IgAN nephropathy.

ETHICS AND CONSENT

Ethical approval is not required for an anonymous case report. Written informed consent was obtained from the patient for publication of this case report.

CONFLICT OF INTEREST

The author declares no conflict of interest.

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