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A Kimmelstiel-Wilson Disease with Focal Lupus Nephritis – A Case Report

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Abstract

BACKGROUND:

A 36 years old male presented with facial puffiness and bilateral pedal edema for 1 week who is a known case for type 2 diabetes mellitus for 10 years. On evaluation we found a rare presentation of Kimmelstiel-Wilson disease with focal lupus nephritis.

INVESTIGATIONS:

Complete Hemogram and routine investigation were performed in this patient and it is revealed hypoalbuminemia and hyperlipidemia with urinary albumin 3+ and sugar 3+loss with urine protein – creatinine ratio of 2.0 and ultrasound abdomen and KUB showed normal kidney size with serum c3 -131.50 (90-150)and c4 -51.50(10-40)complements in these values.Further, renal biopsy showed sclerosed glomeruli with IgG(1+) linear positivity on glomeruli and thickened basement membrane on immunofluorescence

Histopathological examination revealed diffuse and nodular mesangial matrix expansion. PAS + Kimmelstiel Wilson nodular with micro- aneurysmal dilation, no spikes or double contour. Interstitial fibrosis and tubular atrophy were noted in 30% of core. It was diagnosed as RPS-Class 3 focal lupus nephritis.

CONCLUSION:

Kimmelstiel-Wilson syndrome is a kidney condition associated with long standing diabetes. It is evaluated by periodical nephrology follow-up for diabetes patients and adequate glycemic,blood pressure control and end stage renal disease patients are to be managed with hemodialysis.

Keywords: Kimmelstiel-Wilson, Diabetes, nodular disease, focal nephritis

INTRODUCTION

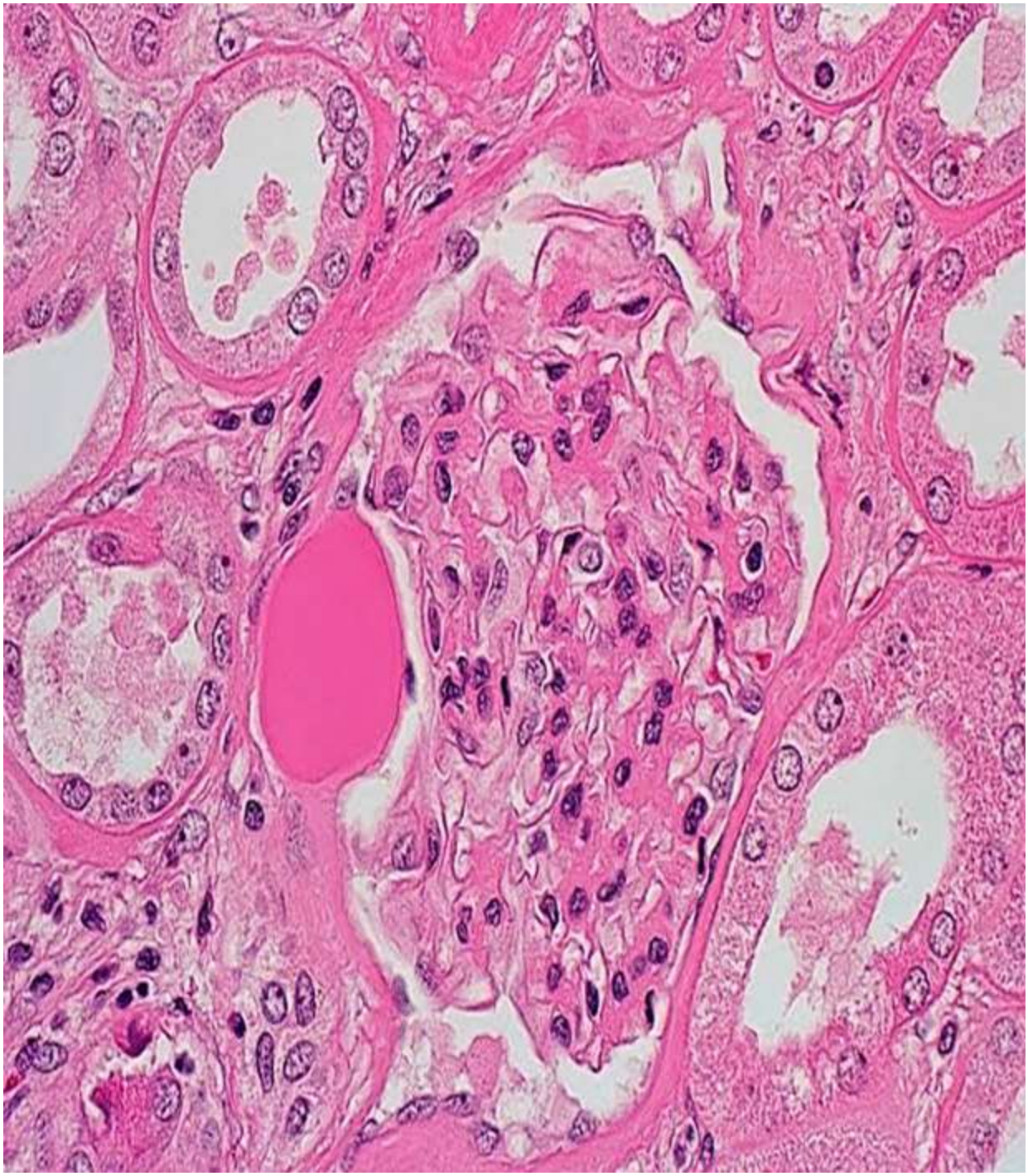
Kimmelstiel-Wilson syndrome is a kidney condition associated with long standing diabetes. It affects the network of tiny blood vessels in the glomerular, a key structure that is composed of capillary blood vessels and which is necessary for the filtration of blood.

Approximately 40% of patients with type 1 or type2 diabetes develop nephropathy, but due to the higher prevalence of Type 2 (90%) have diabetic kidney disease.

CASE HISTORY AND PRESENTATION

A 36-year-old male presented with complaints of facial puffiness in and off for 1 week and bilateral pedal edema for 10 days, No other history of breathlessness, reduced urine output. No H/o hematuria, dysuria, sore throat or use of NSAID or native medications. He is a known case of Type 2 diabetes mellitus for past 10 years and systemic hypertension for 2 years. On examination, he was conscious, oriented, no pallor, not icteric, no lymphadenopathy and bilateral pedal edema seen. Vitals were BP 190/120mm hg PR 90/min SPO2-97%RA. CVS Rs and CNS examination were normal.

motor and sensory examination revealed distal sensory loss. He was diagnosed as volume overload state and treated with anti-hypertensives and diuretics and glycemic control done. Fundus examination showed Grade 3 Hypertensive retinopathy. Urine analysis revealed proteinuria 3+and glycosuria,3+ routine hemogram showed hypoalbuminemia s. albumin-2.3mg% and hyperlipidemia total cholestrol- 456mg% S.TGL-307 mg%Further renal biopsy was done and it showed kimmelstiel-wilson disease with class 3 focal lupus nephritis as mentioned above findings and echocardiogram revealed dilated cardiomyopathy in this patient. NCS study revealed chronic sensorimotor neuropathy of lowerlimbs. Finally, he was diagnosed as Diabetic kidney disease with micro and macro vascular complications. ACE inhibitors and hemodialysis is plan of treatment.



Volume 4, Issue 4; July-August 2021; Page No 1085-1087

DISCUSSION

DEFINITION:

Kimmelstiel-wilson disease is characterized by a progressive decline in proteinuria (>300 mg/24hr) and decline in GFR, hypertension and a high risk of cardiovascular mortality and morbidity. Thickening of basement membrane is a sensitive indicator of significant nephropathy which is altered due to loss of heparan sulfate moieties.

INCIDENCE:

Microalbuminuria appears 5-10 years after the onset of diabetes.

It is recommended to test patients with type 1 disease for 5 years after diagnosis and then yearly thereafter and to test type 2 patients at the time of diagnosis and yearly thereafter.

CLINICAL FEATURES:

- The expansion of mesangium due to the accumulation of extracellular matrix correlates with the clinical manifestations.
- Nodular glomerulosclerosis represents area of marked mesangial expansion appearing a large round fibrillar mesangial zones, with extreme compression of adjacent glomerular capillaries.
- Hyperfiltration, microalbuminuria, overt nephropathy and end stage renal disease
- Microalbuminuria with urinary albumin excretion (<300 mg/24h)
- Diabetic nephropathy is due to decline in GFR and raised arterial blood pressure
- Extrarenal complications like retinopathy occurs 90% in type1 and 60% in type 2 diabetes
- Microangiopathies-stroke, carotid artery disease, coronary artery disease and peripheral vascular disease occurs two to five times common in diabetic nephropathy

DIAGNOSTIC CRITERIA:

- Renal biopsy is the gold standard with or without following evidence
- Macroalbuminuria > 300 mg/day
- Microalbuminuria < 30-300/day with retinopathy
- Microalbuminuria with diabetes for more than 10 years.

TREATMENT:

- Glycemic control
- Blood Pressure control: B.blockers used in arrhythmia 130/80 mm Hg in diabetes thiazide diuretics of GFR > 40 ml/min
- RAAS inhibition: monotherapy with ACE inhibitors or ARB
- Hemodialysis for end stage renal diseases.

CONCLUSION:

Diabetic Nephropathy is predictable on sequences, hence renal biopsy is not indicated in all cases.

40% of patient are managed conservatively.

50% of macroalbuminuria of ESRD will need RRT for survival. Hence, estimation of albuminuria should be done at the earliest for diagnosis. Once patient develops macroalbuminuria CKD is irreversible.

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Volume 4, Issue 4; July-August 2021; Page No 1085-1087