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Research Paper

KIMMELSTIEL-WILSON SYNDROME: A CASE REPORT.

Medical Science

KEYWORDS : : Kimmelstiel Wilson, Glomerulosclerosis, Diabetic nephropathy.

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ABSTRACT

We present a rare case of Kimmelstiel Wilson syndrome initially presenting with swelling in the lower extremities since 6 months. Based on the findings of edema, high grade proteinuria, hypoalbuminemia, and lipiduria, nephrotic syndrome is suspected. A 24-hour urine collection confirms nephrotic proteinuria (5.25 g of protein). A renal biopsy was performed. Histopathological examination of the biopsy tissue showed mesangial nodules (ie, Kimmelstiel-Wilson nodules), diffuse thickening of capillary loops, and glomerulosclerosis.

I. Introduction

Diabetes mellitus (DM) is the most frequent cause of chronic kidney failure in both developed and developing countries [1]. Diabetic nephropathy, also known as Kimmelstiel- Wilson syndrome or nodular diabetic glomerulosclerosis or intercapillary glomerulonephritis, is a clinical syndrome characterized by albuminuria (>300 mg/day or >200 mcg/min) confirmed on at least two occasions 3-6 months apart, permanent and irreversible decrease in glomerular filtration rate (GFR) , and arterial hypertension [2]. The syndrome was first described by a British physician Clifford Wilson (1906-1997) and American physician Paul Kimmelstiel (1900-1970) in 1936 [3].

II. Case Report

A 45 year old female with history of type 2 diabetes mellitus and hypertension since 10 years came to MGM OPD with complaint of increasing swelling in her lower extremities. The swelling worsened over the past 6 weeks. There was no history of fever, chills, arthralgias, joint swelling, or skin rash. She reported no visual changes, epistaxis, hemoptysis, or cough. She had no symptoms of flank pain, hematuria, dysuria, or darkening of the urine. Her medications included oral hypoglycemic drugs and antihypertensives. Family history of diabetes mellitus and hypertension was present. The patient's blood pressure was elevated at 157/95 mm Hg. There was 3+ pitting edema in the lower extremities. No skin rash, petechiae, or purpura was seen. A neurologic examination revealed no focal lesion, motor or sensory changes.

The patient was thoroughly investigated and the findings are as follows:

Serum chemistries

Albumin 2.9 g/dL, Chloride 105 mEq/L,Creatinine 3.83 mg/dL, Potassium, 4.3 mEq/L, Sodium 134 mEq/L, Total cholesterol 261 mg/dL,Urea nitrogen (blood) 43.7 mg/dL, FBS 230 mg/dl

Hematologic findings

Hemoglobin 10.5 g/dL, Leukocyte count 9560/mm3 , Platelet count, 1.76 lakh/mm3, Hematocrit 38.3% , Hemoglobin A1c 7.3%

Urinary findings:

Erythrocytes 10/hpf, Protein- 5.25 g/dL, Hyaline casts-few

Based on the findings of edema, high grade proteinuria, hypoalbuminemia, and hyperlipidemia, nephrotic syndrome was suspected. A 24-hour urine collection confirmed nephrotic proteinuria (5.25 g of protein). Analysis of serum and urine protein and immunoelectrophoresis revealed no immunoglobulins or free light chains. A renal biopsy was performed. Light microscopic evaluation of the kidney tissue was notable for mesangial nodules (ie, Kimmelstiel-Wilson nodules), diffuse thickening of capillary loops, and glomerulosclerosis.

Microscopic findings:

Histopathological examination of H & E stained section studied show few renal glomeruli. There is enlargement of glomeruli with focal thickening of glomerular basement membrane as well as focal mesangial sclerosis & expansion. Few neutrophils are also seen in the mesangial matrix. Interstitium shows mild inflammatory cell infiltrate comprising of neutrophils, eosinophils & lymphocytes. Cloudy change is seen in proximal convoluted tubules.Focal mesangial sclerosis showed PAS positive. Histopathological features suggestive of Diabetic Nephropathy (Nodular Glomerulosclerosis) with Glomerulonephritis.

III. Discussion

Pathologic changes characteristic of diabetic nephropathy can be artificially divided into 5stages. At the onset of diabetes mellitus, a patient's renal histology shows no abnormalities. Within 2 to 3 years (stage I), subtle glomerular basement thickening occurs. After 3 to 8 years (stage II), further glomerular basement thickening and mild mesangial matrix widening develop. Incipient diabetic nephropathy (stage III) is noted at 8 to 15 years and is clinically evident with the onset of microalbuminuria (30–300 mg/24 hours, or > 20 µg/min). Renal histology reveals further glomerular capillary basement membrane thickening, mesangial widening, and intracapillary glomerulosclerosis (ie, glomerular scar formation). Overt diabetic nephropathy (stage IV), associated with macroalbuminuria (> 300 mg/24 hours) and renal dysfunction, occurs after 15 to 29 years. As seen in the case patient, advanced glomerular basement membrane thickening and mesangial widening, Kimmelstiel-Wilson nodules, arteriolar hyalinization, and glomerulosclerosis are noted on renal biopsy. In addition, altered structure of blood vessels (hyalinosis) and tubulointerstitium (tubular atrophy and interstitial fibrosis) is noted. End-stage renal failure (stage V) from diabetes mellitus occurs after 20 to 30 years and is characterized by glomerular capillary closure and sclerotic and hyalinized glomeruli (ie, scarred kidneys).[4]

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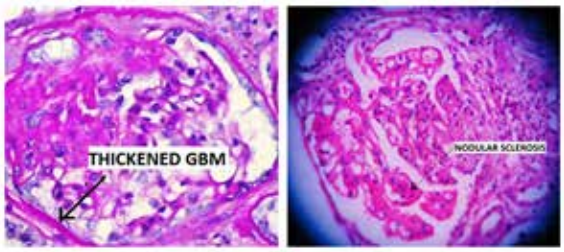
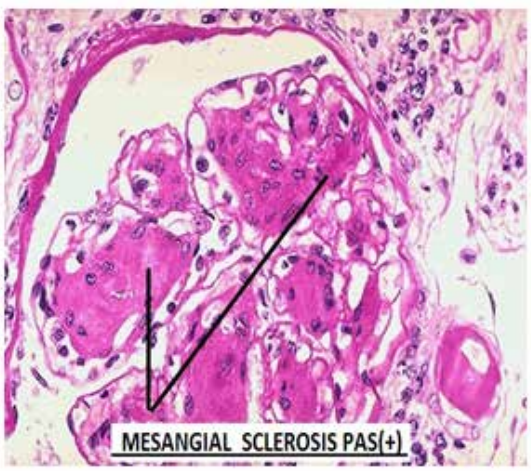
IV. Figures





V. Conclusion

The purpose of this article to stress again the frequency of this grave complication of diabetes, more especially because it affords opportunity to emphasize the degree to which the smallest divisions of the vascular tree may be involved in grossly pathological processes in diabetic subjects, also to attempt to clarify the position of intercapillary glomerulosclerosis and the Kimmelstiel-Wilson syndrome.

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