



A CASE REPORT OF KIMMELSTIEL-WILSON SYNDROME

General Medicine

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ABSTRACT

This is a rare case of Kimmelsteil Wilson Syndrome presenting with swelling all over the body since 8 months. Patient was a known case of diabetes mellitus type 1 for 10 years on regular insulin & based on reports nephrotic syndrome is suspected. Patient had high grade proteinuria, hypoalbuminemia and 24 hour urine collection confirms nephrotic proteinuria. A renal biopsy was performed and histopathological findings showed diffuse and focal nodular glomerulosclerosis with thickened basement membrane of tubules, evidence of atherosclerosis and mesangial nodules.

KEYWORDS

K-W Syndrome (Kimmelsteil-Wilson Syndrome), Diabetic nephropathy

INTRODUCTION

Kimmelsteil Wilson syndrome is a variety of nephrotic syndrome arising from type 1 diabetes mellitus. Diabetes mellitus is most frequent cause of chronic kidney failure in both developed and developing countries. It is also known as nodular diabetic glomerulosclerosis or intercapillary glomerulonephritis diabetic nephropathy. The syndrome was first described by a British physician Clifford Wilson (1906-1997) and American physician Paul Kimmelsteil (1900-1970) in 1936.

It is characterized by

- 1) histological lesion is nodular glomerulosclerosis,
- 2) it develops in a long lasting diabetes mellitus (approx. > 10 years)
- 3) commonly starts with proteinuria,
- 4) In this requirement of insulin becomes less with development of Kimmelsteil Wilson Syndrome,
- 5) later, hypertension may be present,
- 6) end stage renal disease can be expected to occur within 5-6 years.

Case Report

A 42 years old male patient with type 1 diabetes mellitus since 10 years on regular insulin presenting with bilateral swelling on both lower extremities & all over the body for 8 months and breathlessness for 3 days. The swelling had gradual worsened over last 2 weeks. There was no history of fever, chills, cough and common cold, chest pain, arthralgia & skin rash. He had no history of epistaxis, hemoptysis, hematuria/burning micturition or darkening of urine.

Past history had known case of diabetes mellitus type 1 for 10 years. No significant family and personal history of patient.

ON ADMISSION

Temperature : normal

Pulse Rate: 86/min

Blood Pressure : 170/90 mmHg

Random Blood Sugar : 193 mg/dl

Urine acetone : Negative

SpO₂ : 93 % on RA

Respiratory System Examination : bilateral LZ crepitations +

Cardiovascular System : NAD

There was 3+ pitting edema on both lower extremities & signs of anaemia mild pallor visible.

Investigation

BLOOD INVESTIGATION	
Hemoglobin	8.5 mg/dl (M : 13-17 mg/dl)
Total leucocyte count	3800 cells/cumm (4000-10000 cells/cumm)
Platelet count	2.35 lacs/cumm (1.5-4.5 lacs/cumm)
Hematocrit	26.0 % (M : 45 + 5%)
HbA1c	11.6
CRP	>0.6 mg/dl

ESR	16 mm in 1 hr
RBS	193 mg/dl
Creatinine	3.2 mg/dl (0.7-1.4 mg/dl)
Urea	38 mg/dl (15-45 mg/dl)
S. albumin	1.8 gm/dl (3.5-5 mg/dl)
S. sodium	136 mg/dl
S. potassium	4.5 mg/dl
SGPT	95 IU/L
Total cholesterol	101 mg/dl
Triglyceride	86 mg/dl
PT/INR/aPTT	14.5/1.14/24.7
URINE FINDINGS	Turbid, albumin ++, Sugar +, bile salts & bile pigments absent, pus cells 3 to 4/hpf & 7 to 10/hpf RBC
SPECIAL INVESTIGATION	
Urine microalbumin	734 mg/24 hr
24 hr urine microalbumin/ creatinine ratio	21780 mg/g
S. ANA	Negative

USG (abdo+KUB) : liver, spleen, GB and pancreas – NAD
 RIGHT KIDNEY : 9.3 × 4.3 cm & LEFT KIDNEY : 8.1 × 4.1 cm size with bilateral increase cortical echogenicity, CMD partially maintained possibility of RPD (Renal parenchymal disease).

Renal Biopsy:

Histopathological findings : section shows diffuse and focal nodular glomerulosclerosis with thickened basement membrane of tubules, evidence of atherosclerosis & mesangial nodules (Kimmelsteil Wilson nodules). Overall features suggestive of diabetic nephropathy with mesangial nodules & glomerulonephritis. In this case stage IV diabetic nephropathy detected as per pathologic classification of diabetic nephropathy.

Treatment Given:

As per history, clinical examination and supported by investigation we consider it a case of diabetic nephropathy in diabetes mellitus. As per investigation and biopsy report nephrologist opinion taken and started IV human albumin & steroids and then after needed hemodialysis for volume overload. Treated with diuretics and 4 cycles of hemodialysis patient relief better and stable on volume overload. Patient followed up on nephrologist and physician had advised twice weekly hemodialysis and medication of diabetes on regular insulin which was decrease in dose requirements that required in previous years and antihypertensive drugs.

DISCUSSION

Kimmelsteil-Wilson syndrome is a kidney condition associated with long-standing diabetes. It affects the network of tiny blood vessels (the microvasculature) in the glomerulus, a key structure in the kidney that is composed of capillary blood vessels and which is critically necessary for the filtration of the blood. Features of Kimmelsteil-

Wilson syndrome include the nephrotic syndrome with excessive filtration of protein into the urine (proteinuria), high blood pressure (hypertension), and progressively impaired kidney function. When severe, Kimmelstiel-Wilson syndrome leads to kidney failure, end-stage renal disease, and the need for chronic kidney dialysis or a kidney transplant.

Pathologic changes characteristic of diabetic nephropathy can be artificially divided into 5 stages. At the onset of diabetes mellitus, a patient's renal histology shows no abnormalities.

Stage I: Within 2 to 3 years, subtle glomerular basement thickening.

Stage II: After 3 to 8 years, further glomerular basement thickening and mild mesangial matrix widening.

Stage III: Incipient diabetic nephropathy 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 (i.e. glomerular scar formation).

Stage IV: Overt diabetic nephropathy, associated with macro albuminuria (> 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 tubulo interstitium (tubular atrophy and interstitial fibrosis) is noted.

Stage V: End-stage renal failure from diabetes mellitus occurs after 20 to 30 years and is characterized by glomerular capillary closure and sclerotic and hyalinized glomeruli (i.e. scarred kidneys).

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- [4] Kimmelstiel-Wilson Syndrome .Author Prof. Hayk S. Arakelyan. Full Professor in Medicine, Doctor of Medical Sciences, Ph.D , Grand Ph.D .Senior Expert of Interactive Clinical Pharmacology , Drug Safety, Treatment Tactics, General Medicine and Clinical Research.