

	31/3	4/4	23/4	8/5	26/5	1/7
WBC	8400	8000	17400	14400	8600	6600
N/L	70/25	80/15	86/12	72/26	68/10	70/25
Hg	11.4	12.5	11.9	11.3	10.2	10
PLT	326	375	281	301	175	152
UREA	22	26	134	70	93	38
Cr	0.7	0.7	2.3	3.3	5.8	1.6
Na	139	140	136	134	135	133
K	3.5	3.9	3.2	2.7	3	205
Glu	68	68	37		64	
U.Acid		6.1	14.8		4.3	
ESR	30	60	84	31		
Protien	3.9	3.6	3.1			5.4
Albumin	1.9	1.5	1.4	1.8	2	3.2
Chol	216	279	203			
Tg	140	176	217			
P	5.7	5.7	3.5		8.8	
PT	100%	100	64%		56%	
INR	1	1	1.3			
PTT	32	32				
CA	8.2	8.2				
Bleeding Time						1
Coagulation Time						3
Urinalysis						
WBC	30-40	5-7				
RBC	15-18	5-2				
24h protein	3200mg					
24h Volume	1600ml					
TSH	4.22					
C3	69.9					
C4	20.1					
CK	40					
GGT	17					
SGPT	15					
SGOT	19					
ABG						
PH	7.41	7.38	7.31			
PO2	97	97	212			
PCO2	22.5	21.4	24.5			
HCO3	14.3	12.7	12.6			

Results: successful and well prepared nephrectomy with daily dialysis session overcomes any serious state and complication

patient now on dialysis schedule 2 times/week and preparing for kidney transplantation

Conclusions: Kidney biopsy may give a diagnosis that physician may not consider through clinical and lab assessment, and it is a necessary because the right diagnosis and the right treatment may save the patient life.

SUN-455

POEMS SYNDROME PRESENTING WITH ACUTE KIDNEY FAILURE: A CASE REPORT

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Introduction: Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome is a rare paraneoplastic syndrome with multiple systemic manifestations. Substantial kidney involvement is rarely related to this disease.



Methods: We report the case of a 65-year-old female patient who presented with acute anuric kidney failure with the association of edema, gradual onset weakness of all limbs and skin changes (hyperpigmentation and skin thickening).

Results: Nerve conduction study showed severe sensorimotor polyneuropathy. Serum immunofixation showed lambda light chain restricted monoclonal gammopathy. Hormonal assay showed adrenal and thyroidal endocrinopathy.

Conclusions: Following the diagnosis of POEMS syndrome, the patient was given methylprednisolone and dexamethasone without clinical improvement. She died five months after the diagnosis by cardiac rhythm disorders.

SUN-456

MONOGENIC NEPHROTIC SYNDROME IN NIGERIAN CHILDREN



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Introduction: The genetic architecture of idiopathic nephrotic syndrome (NS) in African children is unknown despite evidence pointing to racial differences in the prevalence and clinical course of NS. The objective was to determine the frequency of single gene mutations in Nigerian children with idiopathic NS.

Methods: Genomic DNA from 89 children with NS from 89 families was screened for mutations in 40 NS genes by targeted sequencing of custom amplicons (TSCA) followed by confirmatory direct sequencing. Both strands of the exons were sequenced and the sequences were analysed with the Sequencher Program. We defined pathogenicity using the American College of Medical Genetics variant classification schema. Nephrotic syndrome and response to corticosteroid were defined using the KDIGO guidelines.

Results: The study involved 89 (males: 66.3%) children with NS including 11 (12.5%) children with steroid resistant NS. The median age at diagnosis of NS and enrolment in the study was 5.0 (1.3-14.8) and 8.0 (2.1-16.0) years, respectively. We identified pathogenic mutations in two genes (*INF2* and *TRPC6*) in 2 of 89 (2.2%) children with NS. Interestingly, both children have steroid sensitive (SSNS) course.

Conclusions: We found single gene mutations in <5% of Nigerian children with NS and highlight the rarity of commonly described NS genes in Nigerian children. There is need for larger multi-site studies to verify the results of this study.

SUN-457

ELDERLY ONSET RAPIDLY PROGRESSIVE RENAL DYSFUNCTION WITH KIDNEY ENLARGEMENT AND TUBULOINTERSTITIAL INJURY MIGHT BE A NEW DISEASE ENTITY OF CILIOPATHY



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Introduction: Autosomal dominant tubulointerstitial kidney disease (ADTKD) is a group of uncommon genetic disorders characterized by slowly progressive decline in kidney function and autosomal dominant inheritance. ADTKD is usually described as not showing enlarged kidneys. However, there have been several reports of rapidly progressive renal dysfunction in elderly patients with bilateral kidney enlargement and ADTKD-like histology in Japan. Here we reported three cases with enlarged kidneys who have undergone dialysis with rapid progression and performed analysis of gene panel for target sequence of inherited kidney diseases.

Methods: CASE PRESENTATION: [Cases 1] Eighty-four-year-old Japanese woman without family history of kidney diseases was noted to