A CHALLENGING CASE OF LUPUS-LIKE IMMUNE-COMPLEX MEDIATED GLOMERULONEPHRITIS ASSOCIATED WITH SINUSOIDAL OBSTRUCTIVE SYNDROME

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Introduction: Immune-complex mediated disease presenting with massive deposits, full house immunofluorescence and varied glomerular patterns is usually associated with SLE. We are presenting an interesting unusual case of a patient with an undetermined autoimmune disease presented as sinusoidal obstructive syndrome and immune-complex mediated glomerulonephritis that varied from a membranous pattern with cryo-type organized deposits to a proliferative pattern with massive subendothelial deposits in the absence of systemic manifestations of SLE.

Methods: A 53 years old gentleman with a history of nephrotic syndrome in the context of an unspecified autoimmune disorder. Patient has history of sinusoidal obstructive syndrome (etiology unknown) and underwent TIPS procedure complicated by TIPS syndrome. Two years later, he was diagnosed with membranous nephropathy/PLA2R negative (biopsy proven). He was unsuccessfully treated with steroids and MMF, and then started on Rituximab with a good response to proteinuria and serum albumin levels. However, the patient is recently complaining of worsening nephrotic range proteinuria, a clinical work up including renal biopsy has been conducted to reveal the cause of his worsening proteinuria and reach the optimal medical management options.

Results: The pertinent lab results show low albumin, high cholesterol, and low C3 and C4 serum level. Renal biopsy shows immune complex mediated glomerulonephritis with massive subendothelial, mesangial and few subepithelial immune deposits and in a background of increased global and segmental glomerular scarring. Lupus work up have always been negative for double stain DNA.

Conclusions: This patient shows an immune-complex mediated glomerulonephritis associated with an unidentified autoimmune disease that raised the question of primary unique renal SLE. No conflict of interest

CORTICOSTEROID SPARING AGENTS IN FREQUENT RELAPSING AND STEROID DEPENDENT NEPHROTIC SYNDROME IN CHILDREN: A SINGLE CENTER RETROSPECTIVE STUDY

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Introduction: Children with frequently relapsing nephrotic syndrome (FRNS) or Steroid-dependent nephrotic syndrome (SDNS) may be prescribed non-corticosteroid immunosuppressive agents whenever there is a failure to maintain remission with low-dose alternate-day prednisone and/or significant adverse effects of prednisone develop. A wide variety of immunosuppressive agents have been used in these patients to reduce the number of relapses and maintain remission.

Methods: A retrospective study was conducted on all steroid-sensitive nephrotic syndrome (SSNS) children (1-11 years) who received any type of second line agents (e.g. CNI, MMF, cyclophosphamide, and Rituximab) over a period of 9 years from January 2010 to January 2019 in pediatric nephrology unit in Prince Sultan Military Medical City, Riyadh.

Results: The study included 24 patients. Their age at diagnosis ranged between 1 and 11 years with a mean of 3.8 years and standard deviation of ± 2.6 years. During the first year of steroid therapy, relapse occurred among 87% of patients; of them, the number of relapses being 4 or more in 21.7%. Regarding indication for the second line of treatment, SDNS was the most frequent reported (60.9%), followed by FRNS (30.4%). Concerning agents used in the second line, MMF ranked first (58.4%), followed by Cyclophosphamide (33.3%). Number of relapses after starting steroid sparing agent was more than once among 41.7% of patients. Duration of remission after starting steroid sparing agent
ranged between 2 and 72 months (14.2±14.1). Overall response to the second line of treatment was observed among majority of patients (91.7%). Renal biopsy was performed in 45.8% of patients. Concerning side effects of steroid sparing agents, electrolytes disturbances and hypertension were reported by two (8.3%) and one (4.2%) patients respectively. Cystatin C remission was significantly longer among patients treated with cyclosporine (48±33.9 months) compared to other lines of treatment, p<0.001. On the other hand, hypertension was only reported among patients treated with cyclosporine, p=0.003.

Conclusions: The overall response of children with SDNS and FRNS to the second line agents was significant, with favorable longer remission free period with cyclosporine use with no major side effects. Our results affected by the retrospective design of the study, as well as the small sample size. Therefore larger scale study with prospective design is highly encouraged.

No conflict of interest

POS-449
ADULT ONSET MINIMAL CHANGE DISEASE: CLINICAL CHARACTERISTICS, TREATMENT AND PROGNOSIS

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Introduction: Minimal change disease (MCD) is an uncommon glomerulopathy in adults, accounting for almost 15 to 20% of nephrotic syndrome (NS) unlike children in whom MCD is the most common cause of NS accounting for almost 80% of cases. Most MCD patients respond well to steroids and have a preserved renal function. However, MCD remains a therapeutic challenge for the nephrologist especially for patients who respond well to steroids and have a preserved renal function. However, the second line agents was significantly longer among patients treated with cyclosporine compared to other lines of treatment, p<0.001. On the other hand, hypertension was only reported among patients treated with cyclosporine, p=0.003.

Conclusions: The overall response of children with SDNS and FRNS to the second line agents was significant, with favorable longer remission free period with cyclosporine use with no major side effects. Our results affected by the retrospective design of the study, as well as the small sample size. Therefore larger scale study with prospective design is highly encouraged.

No conflict of interest

POS-450
FIBROBLAST GROWTH FACTOR AS A BIOMARKER OF FOCAL SEGMENTAL GLOMERULOSCLEROSIS IN HIV-POSITIVE AND HIV-NEGATIVE CHILDREN PAID FOR SUBMISSION 2020-A-WCN21-0093 (100981327899)

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Introduction: HIV infection can lead to the development of HIV-associated nephropathy (HIVAN) with the majority of patients progressing to end-stage kidney disease. Previous studies have recognised basic fibroblast growth factor (bFGF) as a biomarker for HIVAN, since significant levels of bFGF low-affinity receptors have been found in the kidneys of HIV-infected children.

The aim of this study was to assess the association between bFGF and kidney disease in the development of focal segmental glomerulosclerosis (FSGS) in HIV-positive and negative children.

Methods: The study group consisted of 31 children; HIVAN (n=11) and idiopathic FSGS (n=20). The control group consisted of both HIV-positive (n=20) and HIV-negative (n=20) children with no kidney disease. Serum samples from all patients in both the study and control groups were analysed for bFGF. ALL HIV positive patients were stable on HAART for over 6 months before being recruited. Estimated glomerular filtration rate (eGFR) was calculated using the modified Swartz formula.

Results: Serum was obtained and stored at -80°C, before bFGF levels were quantified, as mean fluorescence intensity (MFI), using the Bio-Plex Pro Human Cytokine assay (Bio-Rad laboratories Inc., USA) on a Bio-Plex MAGPIX Multiplex system (Bio-Rad Laboratories Inc., USA) using Bio-Plex Manager version 4.1 software.

Data were entered into SPSS version 24 (IBM Corp., USA) and GraphPad Prism version 5 (GraphPad Software, USA) for analysis. A p-value <0.05 was considered as statistically significant. A descriptive statistical analysis of the data was conducted prior to inferential analysis. Independent samples t-test and ANOVA were used to determine if high levels of bFGFs in FSGS compared with controls were responsible for the development of glomerulosclerosis. 12 tests were used to assess any associations between categorical variables.

Results: The concentration of bFGF was higher, in comparison with idiopathic FSGS children, in HIVAN children (p=0.0167). There was also a significant elevation of serum bFGF levels in children with HIVAN when compared with HIV-positive (p=0.0288) and HIV-negative (p=0.0043) control groups.

There was no statistically significant difference in bFGF concentration between children with idiopathic FSGS and those within the positive and negative control groups.

Conclusions: This study demonstrated statistically significant differences between bFGF levels in children with HIVAN and controls.

No conflict of interest