Methods: Patients with NS onset between age 1-60 years and diagnosed with SRNS between January 2010 and October 2015 (n=76) were included. All patients underwent renal biopsy and were started on either cyclosporine (CsA) at 4-5 mg/kg/day targeting whole blood trough levels of 80 - 120 ng/ml or tacrolimus (TAC) at 0.12-0.15 mg/kg/day targeting trough levels of 5-10 ng/ml. All patients were given steroids at a dose of 0.1-0.15mg/kg/day during therapy with CNIs. In patients who achieved complete remission (CR), TAC dose was reduced to achieve trough levels of 3-6ng/ml. All patients received maximum tolerable doses of ACEI/ARBs. CNI therapy was continued for at least 6 months in all patients. CNIs were stopped at 6 months in patients who failed to achieve either CR/PR. In patients who had remission at 6 months, CNIs were continued for a minimum of 12 months. Of the total 76 patients, 30 were newly started on CNIs and had serum uPAR done at baseline and at 6 months and β3 integrin staining on renal biopsy was done.

Results: Patients with age of onset of NS ≤ 12 years of age were classified as childhood-onset NS (n=36) and those with age of onset >12 years of age (n=40) were classified as adult-onset NS. The commonest histology was focal segmental glomerular sclerosis-not otherwise specified (FSGS NOS) (n=45, 59.2%). Minimal change disease (MCD) was the second most common lesion (n=22, 28.9%). In our study, 86.8% of patients received TAC and 13.2% of patients received CsA. The CR at 6 months was 53% and 45% in childhood onset and adult onset SRNS respectively. The CR at 12 months was 66% and 68% in childhood onset and adult onset SRNS respectively. CNI resistance was 20%. AKI was the commonest adverse effect, seen in 16%, followed by infections (6.6%). suPAR levels at baseline did not differentiate between SRNS and SSNS or between FSGS and non-FSGS. The suPAR levels at 6 months of CNI therapy did not correlate with response or resistance to therapy. β3-integrin was negative(n=10) or trace to 1+ (n=12) in our patients with SRNS; their expression did not correlate with histology or response to CNIs.

Conclusions: CNI therapy was continued for at least 6 months in all patients. CNIs were stopped at 6 months in patients who failed to achieve either CR/PR. In patients who had remission at 6 months, CNIs were continued for a minimum of 12 months. Of the total 76 patients, 30 were newly started on CNIs and had serum uPAR done at baseline and at 6 months and β3 integrin staining on renal biopsy was done.

Trace to negative staining for beta 3 integrin antibody in normal kidney

Trace to 1+ positivity in FSGS case

Granular staining (3+) for beta 3 integrin antibody in a case of lupus nephritis-V

MON-020

MULTIPLE POTENTIAL AETIOLOGIES OF MEMBRANOPROLIFERATIVE GLOMERULONEPHRITIS IN A COMPLEX PATIENT

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Introduction: Membranoproliferative glomerulonephritis has a broad possible range of causes. The syndrome of proliferative glomerulonephritis with monoclonal immunoglobulin deposits (PGNMD) as part of the monoclonal gammopathy of renal significance (MGRS) spectrum is not yet clearly defined; also unclear is
the ability for low viral load, untreated hepatitis B to cause proliferative glomerulonephritis (GN) or the significance of anti-Ro in absence of other biochemical, histological or clinical classical lupus manifestations. 

Methods: We present a case of a sixty-three-year-old female who presented with acute on chronic renal dysfunction; she was treatment naive hepatitis B positive with low viral titre, anti-Ro antibody positive and had known IgM kappa monocular gammapathy with stable titres. Importantly cryoglobulins and the rest of her autoimmune panel was negative.

Results: Kidney biopsy light microscopy showed proliferative GN with membranoproliferative pattern. There was moderately severe arteriolar sclerosis and arteriolarosclerosis with few vessels showing intimal fibrinoid necrosis with hyperplasia of tunica media (onion skinning). Six out of twenty glomeruli were obsolescent and there were two fibrocellular crescents. Immunofluorescence showed +4 granular mesangial and peripheral capillary wall reaction with IgM and kappa, 3+ granular mesangial and peripheral capillary wall reaction noted with IgG, lambda and C3 only. Electron microscopy showed variable foot process effacement and a few cryoglobulin-like cylindrical organised deposits. The patient experienced improvement in renal function with mycophenolate and prednisone, however there was debate of the use of chemotherapy agents such as rituximab or traditional myeloma therapy. We are planning to do a repeat biopsy in the future to evaluate regression of renal lesions.

Conclusions: This case demonstrates the increasing evidence that many patients with proliferative glomerulonephritis do not fit neatly in to one pathophysiological box and thus treatment must be tailored to the individual patient.

MON-021

IMMUNOHISTOLOGICAL SPECTRUM OF CRESCENTIC GLOMERULONEPHRITIS, A SINGLE CENTER STUDY IN BANGLADESH

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Introduction: Crescentic glomerulonephritis (GN) is defined as presence of crescents in >50% of glomeruli sampled for renal biopsy. This is a critical diagnosis with clinical implications that ideally is communicated to the referring physician immediately. There is limited data on the etiology, clinical and histopathological spectrum of crescentic glomerulonephritis (CrGN) in Bangladeshi population.

Methods: All the crescentic glomerulonephritis diagnosed histologically at Armed Forces Institute of Pathology, Dhaka irrespective of age and sex are included in the study. The study duration was from January 2017 to June 2018. The renal biopsy samples were undergone H&E, PAS, Masson trichrome and Methanamine silver staining. For each case direct immunofluorescence study was carried out for IgG, IgM, IgA, C3, Clq, Kappa and Lambda. Depending on clinical, biochemical, serological, histological and immunofluorescence evaluation crescentic glomerulonephritis was classified.

Results: During stipulated period 21 crescentic glomerulonephritis were diagnosed which comprises 2.56% out of total 819 non transplant renal core biopsy patients. Mean age was 27.38 +14.928 the largest age group (8/21) being paediatric age group (<16 yrs). The male female ratio was 0.75. The most common presentation was rapidly progressive glomerulonephritis (RPGN) (7/21), followed by generalized body swelling (6/21). On urine examination 19/21 patients had haematuria and 7/21 patients had massive proteinuria (>3.0 Gm/24 hrs). The mean serum creatinine level was 5.65 mg/dl (1.3 to 18.85 mg/dl). Serum C3 & C4 level was found low in 6/18 and 2/15 cases respectively. ANA was positive in 2/21 cases and among those 1 was anti dsDNA positive and labeled as systemic lupus erythematosus (SLE). Among 21 cases 01 could not be classified due to lack of glomerulus during direct immunofluorescence (DIF) study. Pauciimmune crescentic glomerulonephritis (PI-CGN) was 10 (50%). Immune complex mediated crescentic glomerulonephritis (IC-CGN) was 9 (45%) and anti glomerular basement membrane disease (anti GBM disease) was 01(5%). Among the PI-CGN, 5 (50%) were ANCA positive and 03 (30%) were ANCA negative, for other 02 PI-CGN cases ANCA report was not available. Among ANCA positive PI-CGN 01 had associated ANA positivity. Among IC-CGN, 02 were crescentic IgA, 01 was crescentic lupus nephritis, 01 was isolated C3 glomerulonephritis, 03 were membranoproliferative glomerulonephritis with diffuse crescent formation and rest 02 were IC-CGN of unknown reason. The glomerular crescents formation was 80.33% in PI-CGN while it was 61.82% among IC-CGN cases and 66.67% in anti GBM glomerulonephritis. Among IC-CGN, 4/9 (44.44%) cases needed dialysis on admission while 5/10 (50%) PI-CGN cases were managed during admission by dialysis. The only anti GBM disease was tried to be managed by frequent dialysis but failed and plasmapheresis was advised but could not be instituted who ultimately succumbed to death following severe haemoptysis.

Conclusions: In our study commonest cause of crescentic glomerulonephritis was pauci-immune crescentic glomerulonephritis followed by immune complex mediated glomerulonephritis. While one case of anti GBM glomerulonephritis was found that was possibly a case of Goodpasture syndrome.

MON-022

STERIOD RESISTANCE IN CHILDHOOD NEPHROTIC SYNDROME IN MAPUTO, MOZAMBIQUE

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Introduction: First line treatment for childhood nephrotic syndrome, characterized by proteinuria, hypoalbuminemia and edema, is corticosteroid therapy. An association between African ethnicity and steroid resistance has been reported, but published data from sub-Saharan Africa is limited, and no data available to date from Mozambique. The aim of this study was to determine the clinical profile, to assess the level of steroid resistance, and to determine the benefit of corticosteroid treatment in childhood nephrotic syndrome at Hospital Central De Maputo, Mozambique.

Methods: A Retrospective evaluation of patient charts and logbooks was performed to collect demographic and clinical data for a cohort of children (<15 years) diagnosed with nephrotic syndrome, between 2010 and 2015, and followed for at least 6 months, to better ascertain the disease burden and characterize the outcomes and clinical management of nephrotic syndrome in Mozambique. Patients were characterized as sensitive and resistant based upon response to corticosteroid therapy.

Results: Of 27 included patients, 17 (62.9%) were steroid sensitive and 10 (37%) were steroid resistant. 16(59.2%) were boys and 11 (40.7%) were girls, with age group of <5years corresponding to 12 (44.4%) and >5years 15 (55.5%). Steroid resistance (60%) was noted higher in age group <5 years, as compared to 40% in >5years. No cases of plasmadium malaria were isolated in our series. HIV status was negative in 24 of these patients, and positive in one, with two patients who were not tested.

Conclusions: Incidence of corticosteroid resistance is higher than those reported in Asian or Western countries. Based on these findings, improved access to second line treatment is needed. Considering the care of children with nephrotic syndrome is greatly compromised by very limited resources and limited laboratory facilities, more than half of these patients may benefit from the judicious use of corticosteroids.

MON-023

HYPOKALEMIC NEPHROPATHY IN NEPHROTIC SYNDROME PATIENT

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