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C1Q NEPHROPATHY in children with Nephrotic Syndrome - Treatment Strategies and Outcomes

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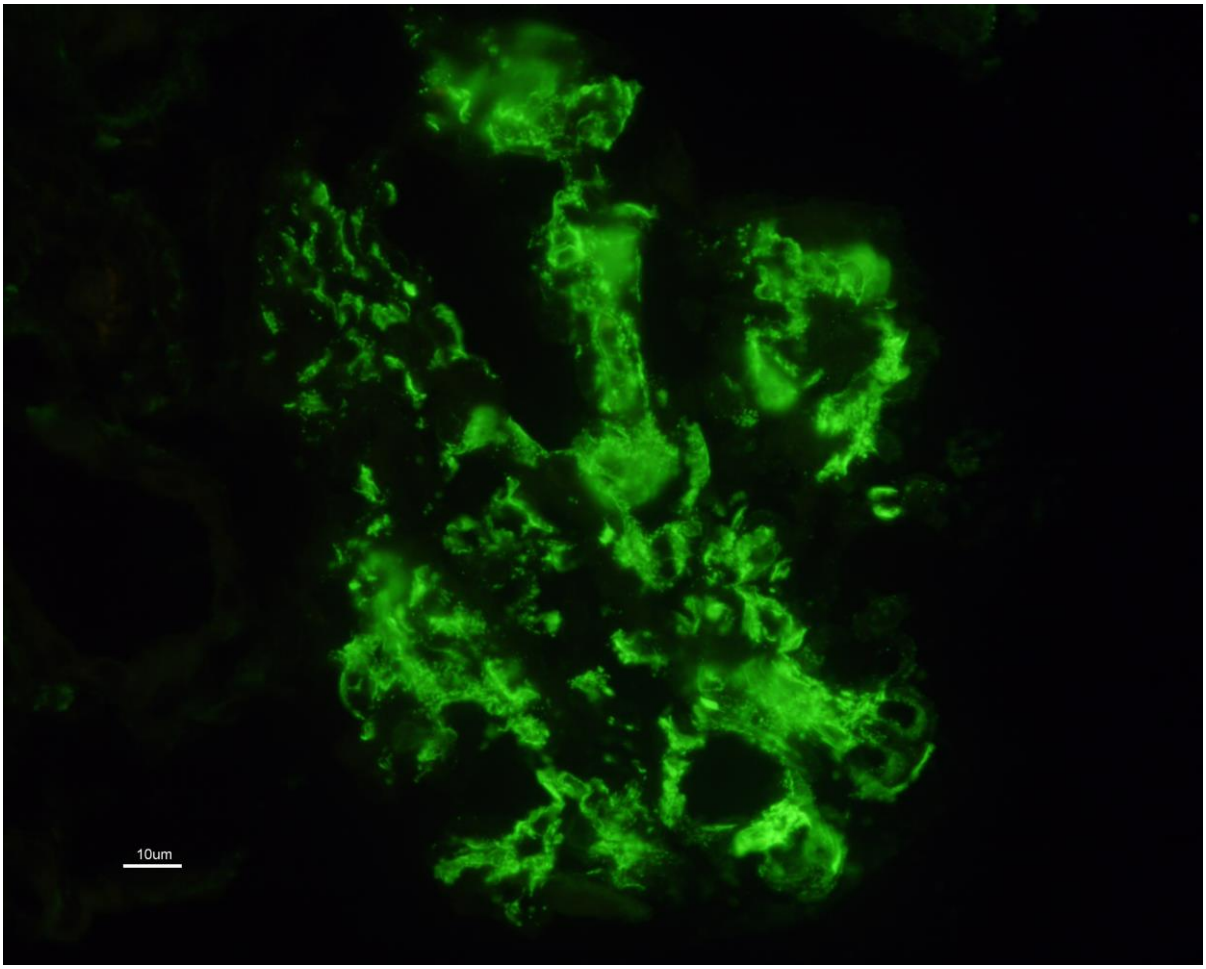
Objectives: There is a paucity of clinical data on C1Q Nephropathy in children South East Asia and India. This is the first detailed analysis conducted to elucidate the prevalence, clinicopathological profile and response to different immunosuppressives in children with C1q nephropathy (C1qN) in Asian children.

Methods: Detailed demographic profile, clinical features, urine and blood chemistries, kidney biopsy (LM/IF/EM) and response to different immunosuppressives analysed between August 2015 to October 2020 in children with steroid dependent/resistant nephrotic syndrome with C1qN.

Results: C1qN was diagnosed in 16 (14.13%) among 113 children who underwent biopsy for Steroid dependent/resistant NS. Mean age is 44 months (Range18- 99 months) and M:F ratio 12 (75%) and 4 (25%) respectively and mean follow up is 3.5 years. 8 (50 %) have coexistent minimal change (MCNS) pattern, 7 (43.7%) have FSGS and one (6.2%) have diffuse mesangial hypercellularity. 13 children had complete follow up of which 8 (61.5 %) and 4 (30.7%) cases presented as steroid dependent and primary steroid resistance while 1 (7.6%) had Joint pain with rashes respectively. At presentation 7 (53.8 %) had hypertension, 12(92.3%) had nephrotic range proteinuria and 6 cases (46.1%) had hematuria. 9 of 12 cases achieved complete remission with Calcineurin Inhibitor therapy and 2 non responders ,1 partial responder and 1 responded to Mycophenolate. Of 6 FSGS cases, 4 had complete remission, 1 had Partial Remission and 1 was in non-Remission. Of 6 cases with MCNS,5 had complete remission and 1 in non-remission. Renal Functions remained normal in all except one case who had progression to CKD stage 3

Conclusions: 1 out 7 children with difficult nephrotic syndrome can have underlying C1Q nephropathy. Calcineurin inhibitors are most beneficial to attain and maintain remission. Renal functions remain normal in majority. Among C1qN ,MCNS and FSGS patterns are seen equally and respond almost similarly to CNIs

Immunoflourescence deposits of C1q in C1q Nephropathy



Detailed Case descriptions along with response to different immunosuppressives

Distribution of treatment given and response

Case No.	Light Microscopy	Therapy	Duration of Follow-Up (Months)	Serum Creatinine (mg/dL)	Urine PCR	Outcome
1	Minimal change	Tacrolimus	51	0.3	0.3	CR
2	Minimal change	Tacrolimus	29	0.3	0.23	CR
3	FSGS	Mycophenolate, tacrolimus	25	0.4	0.3	CR
4	FSGS	Tacrolimus, cyclosporine	22	0.3	0.2	CR
5	Minimal change	Mycophenolate	12	0.4	0.2	CR
6	Diffuse mesangial hypercellularity	Cyclosporine, mycophenolate, rituximab, tacrolimus	11	0.3	0.38	NR
7	FSGS	Tacrolimus	18	1	1.4	PR CKD 3
8	Minimal change	Cyclosporine	7	0.2	0.34	CR
9	FSGS	Mycophenolate, tacrolimus	7	0.2	0.23	CR
10	FSGS	Mycophenolate, cyclosporine	6	0.2	0.18	CR
11	MCNS	Cyclosporine	5	0.3	0.2	CR
12	MCNS	Cyclosporine	6	0.2	0.18	CR
13	FSGS	Tacrolimus	4	0.3	3	NR

FSGS = focal segmental glomerulosclerosis, MCNS = minimal-change nephrotic syndrome, CR= Complete remission; NR = No remission, PR = Partial remission