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Study of the Clinical Profile of Rapidly Progressive Glomerulonephritis

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ABSTRACT

RPGN is a type of nephritic syndrome, accompanied by extensive glomerular crescent formation that, if untreated, progresses to end-stage renal disease over weeks to months. Our study aims to identify factors affecting the treatment outcome in RPGN.

Materials and Methods: A hospital based cross sectional study conducted in the Department of Nephrology at Government Medical College, Jammu for a period of one year after obtaining ethical clearance. Adults with rapidly progressive glomerulonephritis on the basis of renal biopsy were included. Routine clinical, biochemical parameters and biopsy findings were analyzed. Primary outcome identified the remission of the patient with 24-hour urine protein <500 mg/day and serum creatinine <1.4 mg/dl. Quantitative variables were compared using Mann-Whitney Test and qualitative variables were correlated using Chi-Square test/Fisher's exact test. A p value of <0.05 was considered statistically significant.

Results: In our study, about two-third of patients (63.64%) had showed no response. About one-fourth (25.45%) of patients showed partial remission and 10.91% had complete remission. Age, gender, duration of symptoms prior to diagnosis, oliguria, hematuria, hemoptysis, quantity of proteinuria were not correlated with the primary outcomes. Significant correlation was observed with variables like entry serum creatinine, entry eGFR and need of RRT on admission. More than half (90%) of our patients had secondary complications like anemia followed by infections (67%).

Conclusion: RPGN is an important cause of renal failure. Most of the patients present late to hospital. Serum creatinine and requirement of dialysis at presentation are important predictors for outcome in RPGN. Thus, to improve outcome, an early referral to nephrologist for early diagnosis and treatment is stressed.

Keywords: glomerulonephritis, RPGN, nephritic syndrome, renal biopsy

INTRODUCTION

RPGN, is a pathologic diagnosis accompanied by extensive glomerular crescent formation (i.e., > 50% of sampled glomeruli contain crescents which can be seen in a biopsy specimen).It is relatively uncommon, affecting 10 to 15% of patients with glomerulonephritis (GN), and occurs predominantly in patients 20 to 50 years. [1-3]

Classification of Rapidly Progressive Glomerulonephritis Based on Immunofluorescence Microscopy

Antiglomerular basement membrane antibody disease

Antiglomerular basement membrane (GBM) antibody disease (type 1 RPGN) is autoimmune GN and accounts for up to 10% of RPGN cases. It may arise when respiratory exposures (eg, cigarette smoke, viral URI) or some other stimulus exposes alveolar capillary collagen, triggering formation of anticollagen antibodies. The term Goodpasture syndrome refers to a combination of GN and alveolar hemorrhage in the presence of anti-GBM antibodies. GN without alveolar hemorrhage in the presence of anti-GBM antibodies is called anti-GBM glomerulonephritis.

Immunofluorescent staining of renal biopsy tissue demonstrates linear IgG deposits. [4-7]

Immune complex RPGN

Immune complex RPGN (type 2 RPGN) complicates numerous infectious and connective tissue disorders and also occurs with other primary glomerulopathies. Immunofluorescent staining demonstrates nonspecific granular immune deposits. The condition accounts for up to 40% of RPGN cases. Pathogenesis is usually unknown. [8-11]

Pauci-immune RPGN

Pauci-immune RPGN (type 3 RPGN) is distinguished by the absence of immune complex or complement deposition on immunofluorescent staining. It constitutes up to 50% of all RPGN cases. Almost all patients have elevated antineutrophil cytoplasmic antibodies (ANCAs, usually antiproteinase 3-ANCA or myeloperoxidase-ANCA) and systemic vasculitis. [12-15]

Symptoms and Signs

Manifestations are usually insidious, with weakness, fatigue, fever, nausea, vomiting, anorexia, arthralgia, and abdominal pain. Some patients present with abrupt-onset hematuria. Nephrotic syndrome is present in 10 to 30%. Hypertension is uncommon and rarely severe.

Diagnosis

Diagnosis is suggested by acute kidney injury in patients with hematuria and dysmorphic RBCs or RBC casts. Serum creatinine is almost always elevated. [16]

Urinalysis shows hematuria is always present, and RBC casts are usually present.

On CBC, anemia is usually present, and leukocytosis is common.

Serologic testing should include anti-GBM antibodies (anti-GBM antibody disease); antistreptolysin O antibodies, anti-DNA antibodies, or cryoglobulins (immune complex RPGN); and ANCA titers (pauci-immune RPGN).

Early renal biopsy is essential. The feature common to all types of RPGN is focal proliferation of glomerular epithelial cells, sometimes interspersed with numerous neutrophils, that forms a crescentic cellular mass (crescents) and that fills Bowman space in > 50% of glomeruli. [17,18]

Immunofluorescence microscopy findings differ for each type:

- In anti-GBM antibody disease (type 1), linear or ribbon-like deposition of IgG along the GBM is most prominent and is often accompanied by linear and sometimes granular deposition of C3.
- In immune complex RPGN (type 2), immunofluorescence reveals diffuse, irregular mesangial IgG and C3 deposits.
- In pauci-immune RPGN (type 3), immune staining and deposits are not detected. However, fibrin occurs within the crescents, regardless of the fluorescence pattern. [19]

Prognosis

Spontaneous remission is rare, and 80 to 90% of untreated patients progress to end-stage renal disease within 6 months. Prognosis improves with early treatment.

Death is usually due to infectious or cardiac causes, providing that a uremic death is prevented by dialysis.

Treatment

Treatment varies by disease type, although no regimens have been rigorously studied.

Corticosteroids and cyclophosphamide are usually given.

For immune complex and pauci-immune RPGN, corticosteroids (methylprednisolone 1 g IV once/day over 30 min for 3 to 5 days followed by prednisone 1 mg/kg po once/day) may reduce serum creatinine levels or delay dialysis for > 3 yr in 50% of patients

Cyclophosphamide 1.5 to 2 mg/kg po once/day is usually given and may particularly benefit ANCA-positive patients; monthly pulse regimens may cause fewer adverse effects (eg, leukopenia, infection) than oral therapy because of reduced cumulative dosing, but their role is not defined. [20]

Plasma exchange (daily 3- to 4-L exchanges for 14 days) is recommended for anti-GBM antibody disease. Plasma exchange should also be considered for immune complex and pauci-immune ANCA-associated RPGN with pulmonary hemorrhage. [21]

Kidney transplantation is effective for all types, but disease may recur in the graft; risk diminishes with time. In anti-GBM antibody disease, the anti-GBM titers should be undetectable for at least 12 months before transplantation. [22]

For patients with pauci-immune RPGN, disease activity should be quiescent for at least 6 months before transplantation; ANCA titers do not need to be suppressed.

METHODOLOGY

A cross sectional study was conducted in the Department of Nephrology at Government Medical College, Jammu for a period of one year, November 1st 2017 to 31stOctober 2018.The diagnosis of rapidly progressive glomerulonephritis was based on renal histology showing crescents in >50% of glomeruli.

Adults (> 18 years of age) with rapidly progressive glomerulonephritis and renal biopsy under light microscopy showing crescentic glomerulonephritis were included. Any other cause of rapidly progressive renal failure with <50% crescents was excluded.

After a written informed consent, a detailed history, clinical examination and treatment history will be recorded in each diagnosed case. All routine investigations including complete hemogram, urine microscopy, 24 hour urine protein, serum creatinine, blood urea, serum electrolytes, blood sugar, serum cholesterol, uric acid, X-ray chest (PA view), ECG (all leads), USG-KUB were obtained. Kidney biopsy (light microscopy) was studied.

Primary outcomes were labelled as complete, partial or no remission. Complete remission defined as24-hour urine Protein <500 mg/day and serum creatinine <1.4 mg/dl. Partial remission asdialysis independence and serum creatinine <5.8mg/dl. No response asdialysis dependency and serum creatinine > 5.8 mg/dl. Secondary outcomes included infections, hematological, endocrinological, cardiovascular, dermatological and gastrointestinal complications.

STATISTICAL ANALYSIS:

Categorical variables were presented in number and percentage (%) and continuous variables were presented as mean±SD and median. Quantitative variables were compared using Mann-Whitney Test between male and female. Qualitative variables were correlated using Chi-Square test/Fisher's exact test. Ap value of <0.05 was considered statistically significant.

RESULTS

Fifty five patients of rapidly progressive glomerulonephritis were included. There were 23 females (41.82%) and 32 males (58.18%), thus giving a male to female ratio of 1.38:1. The mean age of patients was 46.27 + 16.6 years, with a range of 20 to 75 years. Only one patient was less than 20 years. The mean age of females was 48.44+ 16.27 years and for males it was 44.72 +16.92 years. There was a slight male preponderance in all age groups except in older than 60 years. Age difference both the groups was not statistically significantly (P = 0.699).

Most of our patients presented late. The mean duration of symptoms before diagnosis was 38.58+38.14 days. Above half of our patients (54.5%) had pedal edema and oliguria. Gross hematuria was present in only 14.55% patients. About one-third of patients (32.73%) had hypertension at presentation.

TABLE 1 INCIDENCE OF EXTRA RENAL SYMPTOMS

System involved	Symptoms	No. of patients	%age
Constitutional symptoms	Fever	20	36.36.%
	Headache	0	0.0%
	Weight loss	2	3.64%
	Malaise	6	10.91%
Pulmonary manifestations	Dyspnoea	22	40.0%
	Cough	14	25.45%
	Hemoptysis	10	18.18%
Upper respiratory manifestations	Rhinorrhea	1	1.82%
	Epistaxis	0	0.0%
	Ear discharge	0	0.0%
CNS manifestations	Weakness	1	1.82%
	Wrist or foot drop	2	3.64%
	Seizures	1	1.82%
	Altered sensorium	1	1.82%
Git Manifestations	Pain abdomen	3	5.45%
	Diarrhea	0	0.0%
	Vomiting	10	18.18%
	Ugi bleed	8	14.55%
Rheumatological manifestations	Myalgia	5	9.09%
	Arthralgia	4	7.27%
	Skin rash	2	3.64%

Proteinuria is present in all the patients. More than three-fourth of the patients (80%) had nephritic presentation while in remaining eleven patients (20%) nephrotic-nephritic presentation was observed. The mean value of serum albumin was 2.96 + 0.65 gms/dl. In half of our patients (50.91%) it was in range of 3-4 gms/dl. Severe hypoalbuminemia (<2 gms) was observed in only three patients (5.45%).

Renal failure was present in all the patients at diagnosis. The mean serum creatinine was 9.45 + 4.91 mg/dl with a range from 1.51 to 25.24 mg/dl. One-third (36.36%) of the patients in our study had serum creatinine > 9.5 and one-fourth (27.27%) had in the range of 5.6 – 7.5 mg/dl. The creatinine between 1.5 – 3.5 mg/dl was seen in four patients. 85% of our patients presented with the serum creatinine of >5.5 mg/dl indicating severe renal failure at the time of diagnosis.

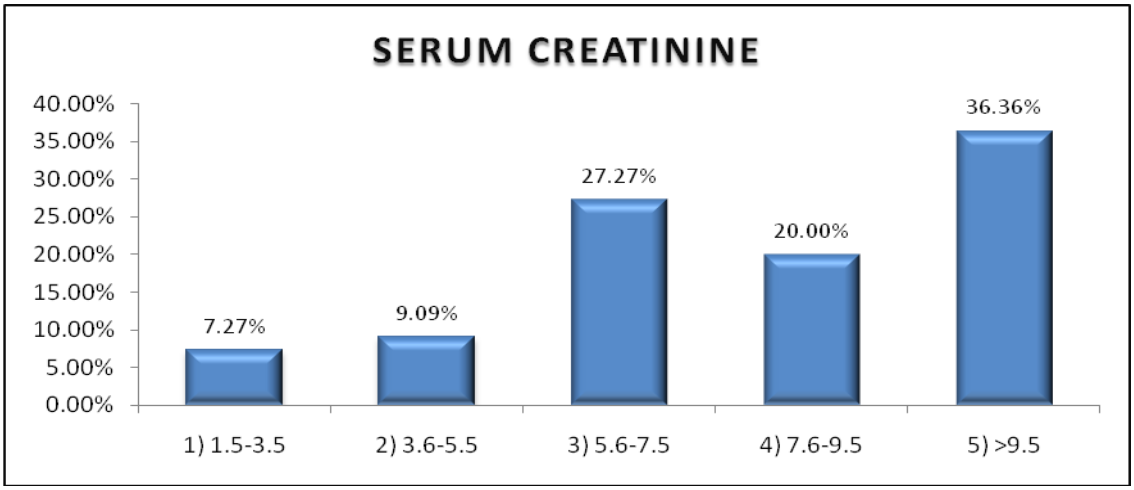

FIG. 1. DISTRIBUTION OF BASELINE SERUM CREATININE

The mean e GFR at diagnosis was 7.93 + 7.46 ml/min/1.73m^2. Almost all the patients (92.73%) presented with e GFR less than 15 ml/min/1.73m^2 in our study. Only one patient had e GFR between 45-59 ml/min/1.73m^2.

Four fifth of the patients (80%) in our study needed renal replacement therapy at diagnosis indicating severe renal failure in majority of them. Only eleven patients (20%) did not require dialysis at that time.

The mean number of sclerotic glomeruli observed in our study was 19.82% + 16.54%, suggesting an advanced stage of disease. Four fifth (80%) of patients had <35% sclerosed glomeruli, however one-fifth (20%) of them had between 36-65%.

Diagnosis of RPGN is based on >50% glomeruli showing crescents. The mean value of crescents in our study was 70.78 + 16.24%. More than half (58.18%) of the patients had 50-70% glomeruli with crescents however, in nine (16.36%) patients >90% glomeruli were found crescentic.

In our study, about two-third of patients (63.64%) had showed no response. One third of patients (36.36%) responded to treatment and had remission. About one-fourth (25.45%) of patients showed partial remission and 10.91% had complete remission. More than three-fourth (85%) of the patients showing remission were older than 40 years. Non-responders were seen maximum in age group (21-30 years). Complete remission was observed in one-tenth (10.91%) of the patients in both males and females.

Partial remission was observed in about one third of males (28.13%) and 21.74 % percent of females. No response was present in 89.57% of females and 59.38% of males.

Most of our patients presented late after onset of symptoms. No response was seen in two-third (75%) of patients, who presented after one and a half month.Oliguria (urine output <400 ml/24 hrs) was present in equal number of patients, both in responders and non-responders. P>0.05. Incidence of gross hematuria in our study is low.

TABLE 2 ASSOCIATION OF DURATION OF SYMPTOMS WITH PRIMARY OUTCOMES

Primary Outcomes	Duration of Symptoms (days)		Results			Total No.	p-value
		N No.	%	Y No.	%		
Complete	1-25.0	23	92	2	8.00	25	0.135
	26-50.0	15	88.24	2	11.76	17	
	51-75.0	3	60	2	40.0	5	
	>75	8	100	0	0.0	8	
	Total	49	89.09	6	10.91	55	
Partial	1-25.0	21	84	4	16	25	0.324
	26-50.0	10	58.82	7	41.18	17	
	51-75.0	4	80	1	20	5	
	>75	6	75	2	25	8	
	Total	41	74.55	14	25.45	55	
No Response	1-25.0	6	24	19	76	25	0.150
	26-50.0	9	52.94	8	47.06	17	
	51-75.0	3	60	2	40	5	
	>75	2	25	6	75	8	
	Total	20	36.36	35	63.64	55	

Only four patients each in the non-responders as well as responder had gross hematuria. p>0.05. Hemoptysis was present in 10 patients with partial remission and no response to treatment. Hemoptysis was not seen in complete remission cases. Four fifth (80%) of patients with remission had e GFR (CKD-EPI) <15 ml/min. All the patients with no response to treatment were admitted with e GFR <15 ml/min association of e GFR with outcomes were found significant. Complete remission (P = 0.014).

TABLE 3 ASSOCIATION OF RRT WITH PRIMARY OUTCOMES

Primary Outcomes	RRT on Presentation		Results			Total No.	p-value
		N No.	%	Y No.	%		
Complete Remission	N	7	63.64	4	36.36	11	0.011
	Y	42	95.45	2	4.55	44	
	Total	49	89.09	6	10.91	55	
Partial Remission	N	5	45.45	6	54.55	11	0.013
	Y	36	81.82	8	18.18	44	
	Total	41	74.55	14	25.45	55	
No Response	N	10	90.91	1	9.09	11	<0.0001
	Y	10	22.73	34	77.27	44	
	Total	20	36.36	35	63.64	55	

More than two third (66%) of the patients with complete remission did not require RRT on admission. Requirement for RRT was present in all except one of the patients with no response to treatment. The association was found to be highly significant. Complete remission (P = 0.011) and No response (P < 0.0001).

More than half (90%) of our patients had anemia followed by infections (67%), neutropenia seen in 47% of the patients. Diabetes, cardiac failure and cardiomyopathy was observed in 29.09% patients

TABLE 4 SECONDARY OUTCOMES

Secondary Outcomes		No.	%age
Infection	-	37	67.27
Hematological	Anemia	50	90.91
	Neutropenia	26	47.27
	Thrombocytopenia	13	23.64
Endocrinological	Diabetes Mellitus	16	29.09
Cardiovascular	Cardiac Failure	16	29.09
	Dialated Cardiomyopathy	16	29.09
Dermatological	Alopecia	2	3.64
	Rash	9	16.36
Gastrointestinal	Vomiting	22	40
	Diarrhoea	1	1.82

DISCUSSION

The mean age of patients was 46.27 + 16.6 years, with a range of 20 to 75 years. Only one patient was less than 20 years. The mean age of females was 48.44+ 16.27 years and for males it was 44.72 +16.92 years. There was a slight male preponderance in all age groups except in older than 60 years. Age difference both the groups was not statistically significantly (P = 0.699).Similar observation have been reported by others. Naidu *et al.,* from North India in their study of forty three patients with pauci-immune glomerulonephritis reported the age range of 40 to 70 years with a mean of 41.1. [23]

Most of our patients presented late. The mean duration of symptoms before diagnosis was 38.58 + 38.14 days. About half (45%) of our patients presented within 25 days and one-fifth (15%) of them reported as late as more than one and a half month of onset of symptoms. Gupta *et al.,* also in a retrospective analysis of 46 cases with crescentic glomerulonephritis done in All India Institute of Medical Science (AIIMS) observed, mean duration of illness to diagnosis of 2 months. [24]

Li *et al.,* In a single Chinese cohort of 89 patients with ANCA associated vasculitis had mean urinary protein/24 hours of 1.5 grams and reported nephrotic syndrome in 12.4% patients. [25]

Renal failure was present in all the patients at diagnosis. The mean serum creatinine was 9.45 + 4.91 mg/dl with a range from 1.51 to 25.24 mg/dl. One-third (36.36%) of the patients in our study had serum creatinine >9.5 and one-fourth (27.27%) had in the range of 5.6-7.5 mg/dl. The creatinine between 1.5-3.5 mg/dl was seen in four patients. 85% of our patients presented with the serum creatinine of >5.5 mg/dl indicating severe renal failure at the time of diagnosis. Four fifth of the patients (80%) in our study needed renal replacement therapy at diagnosis indicating severe renal failure in majority of them. Only eleven patients (20%) did not require dialysis at that time.

In our study, about two-third of patients (63.64%) had showed no response. One third of patients (36.36%) responded to treatment and had remission. About one-fourth (25.45%) of patients showed partial remission and 10.91% had complete remission.

Tang *et al.,* in their study from China enrolled 94 patients with lupus nephritis with diffuse crescentic glomerulonephritis. All the patients were under more than 6 months follow up. At the end of follow up, 12.8% were in clinical remission and 29.8% were in partial remission. [26]

In our study more than three-fourth (85%) of the patients showing remission were older than 40 years. Non-responders were seen maximum in age group (21-30 years). Complete remission was observed in one-tenth (10.91%) of the patients in both males and females. Partial remission was observed in about one third of males (28.13%) and 21.74 % percent of females. No response was present in 89.57% of

females and 59.38% of males. Most of our patients presented late after onset of symptoms. No response was seen in two-third (75%) of patients, who presented after one and a half month. Oliguria (urine output <400 ml/24 hrs) was present in equal number of patients, both in responders and non-responders. P>0.05. Incidence of gross hematuria in our study is low. Only four patients each in the non-responders as well as responders had gross hematuria. P>0.05.

Choudhury *et al.,* reported the incidence of oliguria and hematuria in 61.7% and gross hematuria in 26.4 % patients respectively but no significant association with outcomes were reported in study. [27] Sinha *et al.,* On crescentic glomerulonephritis in patients <18 years at a follow up of 34 months reported oliguria at presentation as a risk factor for renal loss. [28]

Pulmonary hemorrhage is considered as an important predictor of poor outcome in RPGN and demands more aggressive treatment. Hemoptysis was present in 10 patients with partial remission and no response to treatment. Hemoptysis was not seen in complete remission cases

Rampelli *et al.,* reported a mean 24 hours urine protein of 1.66 ± 1.7 gms/days. Proteinuria in both the pauci-immune and immune complex was accessed but no association with outcome was reported. [29]

Renal failure was present in all the patients at the time of recruitment in to the study. Majority (85%) of patients with complete remission had serum creatinine <5.5 mg/dl at entry. Whereas all the patients who had no response to treatment had creatinine > 5.5 on admission.

Ford *et al.,* In their retrospective observational study for indentifying predictors or outcome in 145 patients with ANCA associated vasculitis reported mean creatinine (biopsy) 3.38 mg/dl, (1 year) 2.18 mg/dl, e GFR at presentation once accessed and adjusted for age, gender and sclerotic pattern of glomerular injury had hazard ratio of 0.67 and was associated with significant outcome. [30]

Requirement for dialysis at presentation is one of the predictor of adverse patient and renal survival. More than two third (66%) of the patients with complete remission did not require RRT on admission. Requirement for RRT was present in all except one of the patients with no response to treatment. The association was found to be highly significant. Complete remission (P = 0.011) and No response (P < 0.0001).

CONCLUSION

This study shows that RPGN is an important cause of renal failure. Most of the patients present late to hospital. Serum creatinine and requirement of dialysis at presentation are important predictors for outcome in RPGN. Thus to improve outcome, an early referral to nephrologist for early diagnosis and treatment is stressed. Also secondary complications like anemia and infections cause morbidity in such patients and should be treated at the earliest.

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