Original Article

Poor Outcome of Peritoneal Dialysis during Scleroderma Renal Crisis in Scleroderma Patients

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Objective: Peritoneal dialysis [PD] was a treatment but renal crisis among systemic sclerosis [SSc] patients with Scleroderma renal crisis [SRC] are uncertain. Our objectives were to determine (a) whether peritoneal dialysis [PD] was stopped at 18 months; (b) the clinical predictors thereof and (c) the proportion of patients who needed to change dialysis modality.

Materials and Methods: A historical cohort study among scleroderma renal crisis [SRC] patients followed-up at Srinagarind Hospital, Thailand, between January 1999 and December 2014.

***Results*:** 42 SRC patients were enrolled (30 females): 90% with the diffuse cutaneous SSc subset: mean age at SRC diagnosis 57.3±8.5 years. All were given Captopril, Enalapril, or Ramipril; 5 had renal recovery without dialysis and 22 (52.4%) underwent either hemodialysis [HD] (16; 72.7%) or PD (6; 27.3%). Overall 13 cases underwent continuous ambulatory peritoneal dialysis [CAPD]; 6 chose CAPD as the initial mode of dialysis and 7 changed from HD. One-third in the PD group changed mode due to peritoneal infection and/or fluid leakage. None of the PD patients were free of peritoneal dialysis within 18 months of diagnosis. Most (92.3%) died within 18 months after starting dialysis and most from SRC (75%).

Conclusion: None of the PD SRC patients were free from dialysis within 18 months of starting dialysis. One-third needed to change mode of dialysis due to complications. A minority had renal recovery without any dialysis.

Keywords: Systemic sclerosis, Scleroderma, Observational study, Peritoneal dialysis, Renal crisis

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Systemic sclerosis [SSc) is a heterogeneous autoimmune rheumatic disease of uncertain aetiology, characterized by inflammation and fibrosis of the skin and other organs, and vascular abnormalities, including Raynaud's phenomenon. It has a female predominance and typically presents between 30 and 60 years of age, without major geographical variation. SSc is classified into two subsets distinguished by the extent of skin involvement limited cutaneous systemic sclerosis [lcSSc], and diffuse cutaneous systemic sclerosis [dcSSc] with distinct differences in outcome[1]. Skin outcome in SSc can vary, including spontaneous regression[2], gradually progressive skin thickening, or rapid progressive skin thickening[3].

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Scleroderma renal crisis [SRC] is a life-threatening complication of SSc[4], defined as the onset of severe hypertension associated with a rapid increase in serum creatinine, and microangiopathic hemolytic anemia[5]. Some patients had rising serum creatinine and hemolytic anemia without hypertension[6]. Outcomes of this disorder a once-fatal complication of scleroderma have dramatically improved with the use of angiotensin-converting enzyme [ACE] inhibitors[7].

Most of the SRC patients needed dialysis for acute and long-term renal replacement therapy. Renal recovery was reported as 18 to 24 months after diagnosis of renal crisis[5,7,8]. Peritoneal dialysis [PD] is a dialysis modality for patients who need renal replacement therapy for both acute and long-term treatment. PD, however, is a rare dialysis treatment modality for SSc because of the limitation of applications due to the fibrosis of the skin and peritoneum. According to our observations of daily practice, SSc patients with SRC underwent PD as an acute or long-term renal

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replacement therapy, but there is no report on renal outcomes after PD in SSc patients. The authors, therefore, set out to determine (a) the incidence of being peritoneal dialysis free 18 months after starting PD; (b) the clinical predictors of being peritoneal dialysis free; and, (c) the proportion of the SRC patients who had to change their dialysis modality.

Materials and Methods

A retrospective study was performed among SRC patients followed up at Srinagarind Hospital, Khon Kaen University, Thailand, between January 1999 and December 2014. The population included all SRC patients who underwent or did not undergo dialysis (either PD or hemodialysis). The authors excluded any patients who underwent PD before diagnosis of SSc.

A diagnosis of systemic sclerosis [SSc] was based on criteria of the American College of Rheumatology[1]. SSc was classified as the limited or diffuse type, according to the classification by Le Roy et al[9]. The duration of disease was counted from the date of diagnosis of SRC and the date of the first symptom(s) of SSc. The duration of SRC was calculated from the date of the last follow-up minus the date of SRC diagnosis. The modified Rodnan skin score [mRSS] was assessed at 17 sites as: 0 (normal), 1 (weak), 2 (intermediate), or 3 (severe) skin tightness. Pulmonary fibrosis was defined by chest radiograph or HRCT, which would show interstitial fibrosis. The definition of stomach involvement was fulfilled when patients had either early satiety or dyspepsia. The definition of intestinal involvement was fulfilled when the patient had bloating, constipation, and/or diarrhea. Low, moderate and high dose steroid treatment was <15, 15 to 30 and >30 mg prednisolone/day, respectively.

Scleroderma renal crisis [SRC] includes: (a) the recent onset of severe hypertension (systolic blood pressure [SBP] ≥140 mmHg; diastolic blood pressure [DBP] ≥90 mmHg; a rise in SBP ≥30 mmHg and/or a rise in DBP ≥20 mmHg); (b) a rapid increase in serum creatinine (≥50% over baseline or serum creatinine >120% of upper limit of normal for local laboratory); and, (c) microangiopathic hemolytic anemia[5]. Anemia was indicated by a hemoglobin level <12 g/dL in women and <13 g/dL in men. Thrombocytopenia was defined as a platelet count <100,000 x10^3/uL. Oliguria was defined as a urine volume <400 mL/day.

Hemodialysis [HD] was used to achieve extracorporeal removal of waste products (i.e., creatinine, urea, and free water) from the blood when the kidneys are in a state of renal failure. Peritoneal dialysis [PD] or Continuous Ambulatory Peritoneal Dialysis [CAPD] is a treatment for patients with severe chronic kidney disease. The latter process uses the patient's peritoneum as a membrane across which fluids and dissolved substances (i.e., electrolytes, urea, glucose, albumin, and other small molecules) are filtered from the blood. The beginning date was the date the physician diagnosed renal crisis. Time-to-event (being peritoneal-dialysis-free) was calculated by subtracting the date of diagnosis from the end date. The end-date was the date of being peritoneal dialysis free or the last date meeting the patient in cases when the patients was lost to follow-up or the patient was still undergoing PD or CAPD.

Statistical analysis

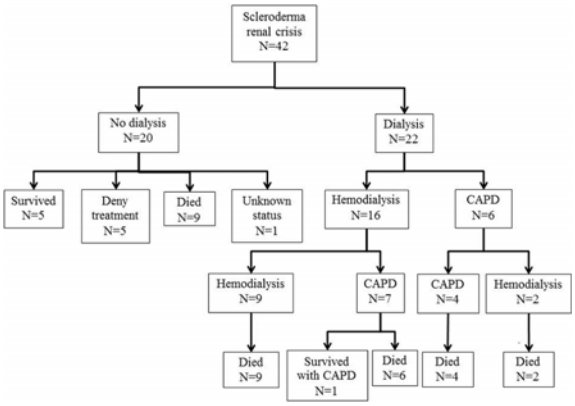
Baseline characteristics and potential factors were categorized according to the type of data (viz., dichotomous, polytomous or continuous variables). The incidence rate and median time to being peritoneal dialysis free were described, and the probability of being peritoneal dialysis free at 18 months was estimated (with 95% confidence intervals; 95% CI). Odds ratio [OR] with 95% CI was investigated to defined the predictors of being peritoneal dialysis free. All of the statistical tests were two-tailed and a *p*-value <0.05 was considered statistically significant. All analyses were performed using STATA version 11.2 (Stata Corp, College Station, TX, USA).

The present study was approved by the Human Research Ethics Committee of Khon Kaen University as per the Helsinki Declaration and the Good Clinical Practice Guidelines (HE561183).

Results

A total of 42 SRC patients were enrolled (71.4% females (n = 30)). The respective mean age at onset of SSc and SRC was 56.2±9.1 (36.9 to 72.9) and 57.3±8.5 (38 to 73.2) years. Most had the dcSSc subset (90.6%) and were anti-topoisomerase I antibody positive (95.7%). The median duration of disease at SRC diagnosis was 8.3 months (IQR 6 to 14.6).

All patients were given Captopril, Enalapril, or Ramipril for SRC treatment. Twenty cases (47.6%) did not require dialysis when SRC was initially diagnosed, while 16 (38.1%) underwent HD and 6 (14.3%) CAPD. Nine cases (40.9%) changed the mode of dialysis: 5 had renal recovery without dialysis and 5 refused dialysis. All patients in the CAPD group died within 18 months of their diagnosis with SRC and 1 in the HD group survived with CAPD (Figure 1). None



CAPD = Continuous Ambulatory Peritoneal Dialysis

Figure 1. Flow chart of scleroderma renal crisis patients.

were taken off dialysis within 18 months of onset of SRC. All of those who denied treatment died within 1 month after diagnosis SRC. The indication and mode of dialysis depended on the decision of attending nephrologists.

Clinical comparisons between patients undergoing vs. not undergoing dialysis at the onset of SRC are presented in Table 1. Oliguria, hyperCKaemia, and pulmonary fibrosis were associated with requiring dialysis (OR 2.27 (95% CI 1.46 to 3.54), 2.43 (95% CI 1.06 to 5.55), and 1.86 (1.14 to 3.02), respectively). Tendon friction rub had a negative association with requiring dialysis (OR 0.36 (95% CI 0.13 to 0.97). Due to the low number of the patients in each group, it was not possible to do a multivariate analysis.

Patients who had oliguria, pulmonary fibrosis, gastroesophageal reflux disease [GERD], and hand deformity preferred HD over CAPD (p-value <0.001, 0.03, 0.02 and 0.04, respectively) (Table 1).

Overall, 13 cases underwent CAPD; chosen as the initial mode of dialysis (at onset of SRC) in 6 cases and an alternative mode in 7. Half of the patients who underwent CAPD at SRC onset were the dcSSc subset and had the anti-topoisomerase I antibody. All of the patients had normal urine flow albeit they had uremia before starting CAPD. One-third in the CAPD group changed mode of dialysis due to peritoneal infection and/or fluid leakage. None of the PD patients were peritoneal dialysis-free within 18 months of SRC diagnosis. All of these patients died within 18 months of starting dialysis (5 or 83.3% due to SRC and 1 or 16.7% due to an unknown cause). Seven of the 16 cases that underwent HD at onset of SRC changed mode of dialysis to CAPD because of financial limitations. One patient experienced inadequate PD so intermittent HD was employed. Most of the patients (6 of 7) who changed mode to CAPD died within 4 months after onset of SRC. The clinical characteristics of the patients including those who underwent CAPD at onset of SRC, and those changed mode from HD are presented in Table 2.

Discussion

The authors described the clinical and demographic characteristics of 42 SSc patients who developed SRC with the objective of analyzing the incidence of being peritoneal-dialysis-free at 18 months after starting PD. The clinical predictors of being peritoneal-dialysis-free and the proportion of patients who had to change dialysis modality were of particular interest. None of the patients, however, achieved being peritoneal-dialysis-free within the specified time and most died within 1 year of SRC diagnosis.

Angiotensin converting enzyme inhibitor [ACEI] is the treatment of choice for SRC despite the lack of a randomized controlled trial[7]. Dialysis is the treatment of choice when serum creatinine rises notwithstanding ACEI treatment. The literature indicates that (a) 20 to 50% of patients with end-stage renal disease getting ACEI need dialysis[10-12] while (b) around 55% could stop dialysis within 2 to 18 months after starting treatment[12] and (c) not all of the SRC patients need dialysis[13]. Around 35.7% (15 cases) of our SRC patients had stable serum creatinine and normal urine flow while taking ACEI, so they did not need dialysis initially; however, only 12% had stable renal function 18 months after onset of SRC and the remainder died thereafter during follow-up. SRC continues to have a poor outcome among SSc patients. Patients who do not initially require dialysis should have their urine flow and renal function closely monitored in order to avoid any delay in dialysis so as to lessen the risk of morbidity and mortality.

Around 64.3% of our SRC patients (including the patient who refused treatment) required dialysis and this rate of needing dialysis is higher than previously reported[14,15]. Oliguria, hyperCKaemia, and pulmonary fibrosis were associated with dialysis requirement in our SRC patients. In general, oliguria and uremia are the principle indications for starting dialysis in patients with acute kidney injury[16]. In the present study, patients with oliguria were started on dialysis. HyperCKaemia was also associated with dialysis requirement, perhaps because of renal toxicity related to myoglobin[17]; thus, renal recovery would

Table 1. Clinical comparisons between patients undergoing and not undergoing dialysis at the onset of SRC and clinical comparison according to mode of dialysis

Clinical	Dialysis				Mode of dialysis		
	No n = 14 (%)	Yes n = 22 (%)	OR (95% CI)	*p*-value	HD n = 16 (%)	CAPD n = 6 (%)	*p*-value
Age at onset >60 years	5 of 13 (38.5)	7 of 19 (36.8)	0.97 (0.54 to 1.77)	0.93	6 of 14 (42.9)	1 of 5 (20)	0.36
Age at SRC >60 years	5 (35.7)	8 of 21 (38.1)	1.04 (0.60 to 1.81)	0.89	7 (43.7)	1 of 5 (20)	0.34
Female	12 (85.7)	14 (63.6)	0.67 (0.42 to 1.08)	0.15	9 (56.3)	5 (83.3)	0.24
Disease duration of SRC >1 years	10 of 13 (76.9)	9 of 20 (45)	0.60 (0.35 to 1.04)	0.07	9 of 14 (64.3)	2 (33.3)	0.20
Diffuse cutaneous subset	12 of 12 (100)	14 0f 17 (82.4)	0.54 (0.38 to 0.77)	0.12	11 of 13 (84.6)	3 of 4 (75)	0.66
Anti-topoisomerase I positive	11 of 11 (100)	10 of 11 (90.9)	0.48 (0.30 to 0.75)	0.31	7 of 8 (87.5)	3 of 3 (100)	0.52
Raynaud phenomenon	10 of 13 (76.9)	16 of 21 (76.2)	0.98 (0.53 to 1.82)	0.96	13 (81.3)	3 of 5 (60)	0.33
Fingertip ulcer	3 (21.4)	1 of 20 (5)	0.39 (0.07 to 2.20)	0.14	0	1 of 5 (20)	0.08
Hand deformity	5 of 13 (38.5)	12 of 20 (60)	1.41 (0.79 to 2.52)	0.23	11 of 15 (73.3)	1 of 5 (20)	0.04*
Weakness	6 of 13 (46.2)	13 of 20 (65)	1.37 (0.75 to 2.51)	0.28	10 of 15 (66.7)	3 of 5 (60)	0.79
Tendon friction rub	8 of 13 (61.5)	3 of 19 (15.8)	0.36 (0.13 to 0.97)	0.01*	3 of 14 (21.4)	0	0.26
Dysphagia	7 of 13 (53.9)	17 of 21 (81)	1.77 (0.79 to 3.95)	0.09	14 (87.5)	3 of 5 (60)	0.17
GERD	5 of 13 (38.5)	9 of 20 (45)	1.11 (0.64 to 1.92)	0.71	9 of 15 (60)	0	0.02*
Stomach symptoms	4 of 13 (30.8)	6 of 20 (30)	0.96 (0.17 to 6.04)	0.96	6 of 15 (40)	0	0.09
Intestinal symptoms	1 of 13 (7.7)	6 of 19 (31.6)	1.65 (1.02 to 2.67)	0.11	4 of 14 (28.6)	2 of 5 (40)	0.64
Pulmonary fibrosis	1 of 13 (7.7)	8 of 19 (42.1)	1.86 (1.14 to 3.02)	0.03*	8 of 14 (57.1)	0	0.03*
Pulmonary hypertension	0	2 of 20 (10)	1.72 (1.28 to 2.32)	0.24	2 of 15 (13.3)	0	0.39
Pericardial effusion	6 of 13 (46.2)	14 of 21 (66.7)	1.40 (0.77 to 2.54)	0.24	12 (75)	2 of 5 (40)	0.15
CPK >200 U/L	4 of 12 (33.3)	17 of 21 (81)	2.43 (1.06 to 5.55)	0.01*	12 of 15 (80)	5 (83.3)	0.86
Anemia	14 (100)	22 (100)	NA	NA	16 (100)	6 (100)	NA
Thrombocytopenia	3 (21.4)	12 (54.6)	1.68 (1.00 to 2.81)	0.05	7 (43.8)	5 (83.3)	0.10
Hypertension	11 (78.6)	16 (72.7)	0.89 (0.51 to 1.55)	0.69	11 (68.8)	5 (83.3)	0.49
Oliguria	0	11 (50)	2.27 (1.46 to 3.54)	0.001*	11 (68.8)	0	<0.001*

SRC = scleroderma renal crisis; GERD = gastroesophageal reflux disease; CPK = creatine phosphokinase; NA = no analysis; HD = hemodialysis; CAPD = continuous ambulatory peritoneal dialysis

Table 2. Clinical characteristics of patients who underwent CAPD at SRC onset and after changing mode from HD

Characteristics at diagnosis of SRC	CAPD at SRC onset						CAPD after changing mode from HD						
	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8	Case 9	Case 10	Case 11	Case 12	Case 13
Age, years	38	67	55	48	54	52	47.5	64	20	67.5	70	49	47
Sex	Female	Male	Female	Female	Female	Female	Female	Female	Male	Female	Male	Male	Female
Cutaneous subset	Limited	Diffuse	Diffuse	Diffuse	ND	Limited	ND	Diffuse	ND	Diffuse	Limited	Diffuse	Diffuse
Anti-topoisomerase I antibody	Positive	ND	Positive	Positive	ND	ND	Positive	Positive	Positive	Positive	ND	ND	Positive
Modified Rodnan skin score	13	ND	19	ND	ND	4	ND	28	46	34	ND	37	ND
Raynaud phenomenon	No	ND	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	Yes	No	Yes
Fingertip ulcer	No	ND	Yes	No	No	No	No	No	No	No	ND	No	No
Hand deformity	Yes	ND	No	No	No	No	Yes	Yes	Yes	Yes	ND	Yes	Yes
Weakness	Yes	ND	No	Yes	Yes	No	Yes	Yes	Yes	No	ND	Yes	Yes
Tendon friction rub	No	ND	No	No	No	No	Yes	Yes	No	No	ND	No	No
Dysphagia	No	ND	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
GERD	No	ND	No	No	No	No	Yes	No	Yes	Yes	Yes	No	Yes
Stomach symptoms	No	ND	No	No	No	No	No	Yes	Yes	No	No	No	Yes
Intestinal symptoms	Yes	ND	No	No	No	Yes	No	No	No	Yes	Yes	No	ND
Pulmonary fibrosis	No	ND	No	No	No	No	Yes	Yes	No	Yes	Yes	Yes	ND
Pulmonary hypertension	No	ND	No	No	No	No	No	No	No	No	No	No	No
Pericardial effusion	No	ND	Yes	Yes	No	No	Yes	Yes	Yes	Yes	No	Yes	Yes
CPK > 200 U/L	Yes	Yes	Yes	Yes	Yes	No	Yes	Yes	Yes	No	ND	Yes	Yes
Treatment													
Prednisolone (mg/d)	45	10	50	No	30	10	30	10	40	20	No	60	30
- Indication	Myositis	Edematous skin	Myositis	-	Myositis skin	Edematous	Myositis	Edematous	Myositis skin	Myocarditis	-	Myositis	Myositis

ND = no data; CAPD = continuous ambulatory peritoneal dialysis; SRC = scleroderma renal crisis; GERD = gastroesophageal reflux disease, CPK creatine phosphokinase

Table 2. Cont.

Characteristics at diagnosis of SRC	CAPD at SRC onset						CAPD after changing mode from HD						
	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8	Case 9	Case 10	Case 11	Case 12	Case 13
Immunosuppressive drug	No	No	Cyclophosphamide	No	No	No	No	No	Cyclophosphamide	No	No	No	No
- Indication	-	-	Alveolitis	-	-	-	-	-	Alveolitis	-	-	-	-
BUN/Creatinine (mg/dl)	99.2/5.2	81.4/9.3	82.1/5.5	147.9/5.2	108.1/5.7	74.2/8.3	129.7/8.9	112.4/5.4	92.6/5	98.9/8.1	57.2/6.2	108/5.6	79.8/5.9
Anemia	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Thrombocytopenia	Yes	Yes	Yes	No	Yes	Yes	No	No	No	No	Yes	Yes	Yes
Hypertension	Yes	Yes	Yes	No	Yes	Yes	Yes	No	No	Yes	Yes	Yes	Yes
Oliguria	No	No	No	No	No	No	Yes	No	Yes	Yes	Yes	Yes	Yes
Indication for dialysis	Uremia	Uremia	Uremia	Uremia	Uremia	Uremia	Uremia	Uremia	Uremia	Uremia	Oliguria	Uremia and oliguria	Uremia and oliguria
Complication of CAPD	None	Infected CAPD	Infected CAPD	Infected CAPD	Infected CAPD	Infected CAPD and CAPD leakage	None	None	Inadequate dialysis	Infected CAPD	None	None	None
Change mode of dialysis to hemodialysis	No	Yes	No	No	No	Yes	Changed from HD	Changed from HD	Changed from HD	Changed from HD	Changed from HD	Changed from HD	Changed from HD
Cause of death	SRC	SRC	Unknown	SRC	SRC	SRC	SRC	Alive	SRC	Sepsis	SRC	Sepsis	SRC
Duration of SRC at death (months)	13.6	5	9.2	3.8	3.5	2.3	2.4	-	4	3	2.7	1.9	2.4

ND = no data; CAPD = continuous ambulatory peritoneal dialysis; SRC = scleroderma renal crisis; GERD = gastroesophageal reflux disease, CPK creatine phosphokinase

not be possible in patients with SRC. None of our patients underwent kidney biopsy, so the authors cannot provide the nature of the mechanism of acute kidney injury in those patients. It is intriguing that tendon friction rub had a negative association with dialysis among SRC patients despite its indicating a poor prognosis in SSc[18] and its being associated with SRC[19]. The authors cannot explain this associated condition.

Mode of dialysis depends on the overall patient condition. PD is a dialysis modality for patients who are not fluid overloaded since PD does not remove fluid immediately or quickly. So, it is not surprising that all of our SRC patients who still had normal urine flow chose PD as their initial dialysis modality.

CAPD seems to be associated with poor outcomes in Thai SSc with SRC. According to our study, most of the patients who underwent CAPD had complications related to the procedure. Infection was the most common complication, which required the mode of dialysis be changed. Inappropriate local peritoneal exit-site care or complicated procedures for PD might lead to bacterial contamination and cause infection. The authors did not suggest performing PD in SSc patients because of the potential for poor outcomes and associated complications. In cases where PD is chosen such as limitations to venous access or cardiopulmonary compromise sterile technique should be applied to avoid infection.

According to its pathogenesis, fibrosis is the main pathology in SSc although peritoneal fibrosis has never been reported. Most of our SRC patients who underwent CAPD had adequate dialysis but 1 case experienced an infected CAPD and experienced leakage. The patient with CAPD leakage had the limited cutaneous SSc subset and no skin tightness of the abdominal skin, so the leakage was not associated with severity of skin tightness. Other possible causes of the CAPD leakage in the patient could have been peritoneal fibrosis, peritonitis, or technical error during tube placement. The authors did not, however, perform peritoneal assessment or peritoneal biopsy to discern the cause of leakage, so the authors cannot conclude its nature.

According to our results, 7 of 16 patients had their mode of dialysis changed from HD to CAPD because of financial reason. The changing mode of dialysis could be regarded as a confounding factor that might be a proxy for a poor outcome of CAPD in SRC patients; however, we are limited in our ability to evaluate and compare the effectiveness and adequacy of HD and CAPD in our SRC patients. Our objective, moreover, was not to compare the outcome of the mode of dialysis in SSc patients, so we cannot provide any detail on dialysis adequacy for either HD or CAPD in patients who changed their mode from HD to CAPD. The effectiveness of each mode of dialysis should be further investigated vis-a-vis what is most appropriate for SRC.

Almost 70% of overall CAPD patients (9 of 13) died from SRC despite early dialysis and ACEI treatment. This might be associated with the ongoing SRC disease process in patients commonly diagnosed with the dcSSc subset and a short duration of disease. According to the nature of disease, the prevalence of SRC is more common in dcSSc than in lcSSc and it frequently occurs in the first 4 years after disease onset[4,20]. As a consequence, SRC could be already be underway in our SSc patients, leading to a major cause of death among this group of patients.

Most of our patients received steroid therapy during diagnosis of SRC; 5 of 6 of those who had an infected CAPD took steroid therapy, 1 of whom died from sepsis. Since steroid use can suppress immune function, it may also increase the risk of peritoneal infection among SSc patients who underwent peritoneal dialysis. There is a report of peritoneal infection among systemic lupus erythematosus patients receiving steroid therapy while undergoing peritoneal dialysis[21]; however, there have been no similar reports on SSc patients. Since steroid (an equivalent dose of prednisolone 15 mg/d) is associated with the development of SRC[11], it is uncertain that steroid use can decrease the rate of renal recovery and/or increase mortality among SRC patients. The effect of steroid use on the prognosis of SRC in SSc needs further investigation.

The limitations of the present study were (a) its retrospective data collection (b) the relatively small number of patients limiting the power of the multivariate analysis (c) the lack of a gold standard to define SRC and a diagnosis that relied upon an expert opinion and (d) the lack of autopsy proof on the precise cause of death. Although none of our patients were peritoneal-dialysis-free 18 months after being diagnosed with SRC, this is the first study to determine the incidence of being peritoneal-dialysis-free 18 months after starting PD. The preliminary data have some value for evaluating patients in Thailand with SRC; in order to determine the outcome of treatment with PD. Ultimately, the data can be used for devising better care of SSc patients with SRC.

Conclusion

None of the PD SRC patients were free from dialysis 18 months after starting dialysis and one-third needed to change the mode of dialysis due to complications. Few patients experienced renal recovery without some kind of dialysis.

What is already known on this topic?

SRC is a serious complication in SSc. The clinical triad was rapid rising of serum creatinine, microangiopathic hemolytic anemia and high blood pressure. ACE inhibitors lead to dramatic clinical improvement and renal recovery was around 18 to 24 months after diagnosis of SRC, however; most of the patients needed dialysis despite of early diagnosis and treatment. PD is a modality treatment for SSc but it has a limitation of application related to the fibrosis of the skin and peritoneum. Renal outcome in SRC after PD is uncertain.

What this study adds?

Almost of SRC patients were diffuse cutaneous SSc. Renal outcome of SRC after PD was poor. None of the PD SRC patients could stop the dialysis within 18 months after starting dialysis. One-third of the PD SRC patients had to change mode of dialysis due to peritoneal infection and/or fluid leakage.

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Author contributions

All of the authors have read and prepared the manuscript.

Potential conflicts of interest

The authors have no conflicts of interest.

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