Journal of Korean Medical Science
Vol. 11, No. 3, 244-249 June 1996

Anorectal Dysfunction in Systemic Sclerosis

Kyung-Chul Kim, Hyo-Jin Park, Soo-Kon Lee, Jun-Pyo Chung, Kwan-Sik Lee, Chae-Yoon Chon, In-Suh Park

Department of Internal Medicine, Yonsei University College of Medicine, Seoul, Korea

This study was aimed to evaluate the anorectal dysfunction in systemic sclerosis(SSc) and propose the clinical significance of anorectal manometry in patients with SSc. Seven patients with SSc were evaluated with manometry for anorectal function and an additional 11 normal subjects were collected as a control group. The study group underwent esophageal manometry as well and the correlation between the degree of anorectal and esophageal dysfunction was evaluated. Patients showed a lower tolerance for balloon distention of the rectum than controls(minimal sensory volume and urgency volume, $P<0.05$). The resting and squeezing pressure of the anal sphincter and the functional length of the anal canal showed no significant difference in these two groups. Rectoanal inhibitory reflex was absent in one(14%) and diminished in two(29%) of seven patients with SSc. SSc patients also showed abnormal esophageal manometry findings, notably decreased LES pressure and body amplitude of distal 2/3 esophagus. The comparision between manometric profiles of anorectum and esophagus showed no significant correlation by statistical analysis. In conclusion, our data could suggest that anorectal function may be impaired in patients with SSc which could reflect the involvement of the anorectum by the disease, and that anorectal manometric studies can be useful to detect such dysfunction in patients with SSc, even before clinical symptoms.

Key Words : *Systemic sclerosis, Anorectal dysfunction, Anorectal manometry*

INTRODUCTION

Systemic sclerosis(SSc) is a multisystem disorder of unknown cause characterized by fibrosis of the skin, blood vessels, and visceral organs, including the gastrointestinal tract, lungs, heart, and kidneys (LeRoy et al., 1988). The gastrointestinal tract is affected in about 50 to 80% of patients with SSc (Cohen et al., 1980 ; Abu-Shakara et al., 1994 ; Sjogren, 1994). Dysphagia associated with motor abnormalities of the distal esophagus is the most common clinical gastrointestinal manifestation (Hurwitz et al., 1976 ; Maddern et al., 1984). Although esophageal manifestations are widely recognized, those due to involvement of the lower gastrointestinal tract are not. Such manifestations were thought to be present mainly in long-standing cases (Poirier and Rankin, 1972) and there were few published reports concerning them until recently. Recent reports suggest that the anorectal function is affected rather early and frequently in SSc patients (Hamel-Roy et al., 1985 ; Chiou et al., 1989 ; Basilisco et al., 1992 ; Leighton et al., 1993 ; Engel et al., 1994).

Address for correspondence : *Hyo-Jin Park, M.D., Department of Internal Medicine, Yong Dong Severance Hospital, Yonsei University College of Medicine, 146-92, Dogok-Dong, Kangnam-Ku, Seoul 135-270, Korea.*
Yong Dong P.O. Box 1217, Seoul, Korea.
Tel : (02) 3450-3314, Fax : (02) 561-3887

The purpose of this study is to investigate the anorectal dysfunction in SSc by evaluating anorectal manometry in patients with this disease, correlate these manometric abnormalities with those of the esophagus, and propose the clinical significance of anorectal manometry in SSc patients.

MATERIALS AND METHODS

Patients

Seven patients with SSc and 11 normal subjects were studied in the department of internal medicine, Severance Hospital, Yonsei University College of Medicine from April, 1994 to January, 1995. The patients were recruited from rheumatology department after informed consent was given. The control group consisted of 11 patients who came to the hospital for physical check up and had no anorectal syptoms or pathology. The clinical features of the patients were obtained through a careful review of their medical records and individual interview.

Anorectal manometry

All SSc and control patients underwent manometric study with a continuous water perfusion system. The manometric tube consists of two polyethylene catheters with two side holes which were 2.5 cm apart. A low-compliance pump (Arndorfer Specialities Inc., Greendale, Wis., USA) perfused distilled water through the catheter at a rate of 0.6 ml/min. A rectal distending balloon was attached to the distal end of the catheter which was connected to an external transducer. Pressures transmitted to the transducer were recorded on a four-channel polygraph(Synetics, Stockholm, Sweden). Patients were positioned in the left lateral position and the catheter was inserted in the anus of subjects who received no enemas, no laxatives, premedication, or other preparation except defecation before the examination.

Anal pressure was measured by the station pull-through technique. Rectoanal inhibitory reflex (RAIR) was assessed by measuring anal pressure during inflation of the rectal balloon with 50ml of air. If there was no response by 50 ml of air inflation, negative RAIR was considered. In addition to the functional length (FL) of the anal canal, the volume of balloon inflation to elicit the first sensation of defecation (minimal sensory volume, MSV), urgency volume(UV), maximal tolerated volume (MTV) were measured.

Esophageal manometry

All SSc patients underwent esophageal manometry study with the standard low-compliance system (Arndorfer Specialities Inc., Greendale, Wis., USA). Using the station pull-through technique manometric parameters were measured and they were interpretated with the standard criteria.

Statistical analysis

Comparisons between two unpaired means were made using the Mann-Whitney U test. Pearson correlation analysis was used to test whether there was a correlation between the manometric profiles of the anorectum and esophagus. Data were expressed as mean±standard deviation and p values less than 0.05 were considered to be significant.

RESULTS

Clinical Data

The seven patients with SSc were all female and their average age was 36.3 yrs(range, 22-56 yr). These patients were compared with 11 female controls with a mean age of 45.4 yrs (range, 19-61 yr). The duration of disease varied from 1 to 6 yr (mean, 2.7 yr). Half of the patients had symptoms related to gatrointestinal tract and those with upper gastrointestinal symptoms had also lower gastrointestinal symptoms (Table 1). Their specific symptoms are summarized in Table 2.

Table 1. Clinical features of patients with systemic sclerosis

Patient No	Age (Yr)	Sex	Duration (Yr)	UGI Sx	LGI Sx
1	33	F	3	−	−
2	22	F	3	+	+
3	46	F	1	−	−
4	30	F	1	+	+
5	36	F	6	+	+
6	56	F	1	−	−
7	31	F	4	+	+
Mean	36.3		2.7		

UGI Sx : Upper gatrointestinal symptoms
LGI Sx : Lower gastrointestinal symptoms

Table 2. Gastrointestinal symptoms of patients with systemic sclerosis

	No of cases(%)
LGI Symptoms	
Incomplete evacuation	4 (57%)
Constipation	2 (29%)
Fecal incontinence	1 (14%)
Tenesmus	1 (14%)
UGI Symptoms	
Nausea	3 (43%)
Chest pain	2 (29%)
Heartburn	1 (14%)
Regurgitation	1 (14%)
Belching	1 (14%)

Anorectal function studies

Manometric findings of the anorectum are summarized in Table 3. The patients with SSc were less able than the controls to tolerate distention of the rectum with an air-filled balloon. The mean of minimal sensory volume was 35.0±4.5ml, 48.2±6.8ml ($p<0.05$), urgency volume was 87.5±29.5ml, 118.2±29.5ml ($p<0.05$), maximal tolerated volume was 143.3±32.8ml, 171.4±39.5ml, in SSc patients and controls respectively (Fig. 1). The resting and squeezing pressure of the anal sphincter and the functional length of the anal canal showed no significant difference in these two groups (Fig. 2). Rectoanal inhibitory reflex was decreased in two (29%) and absent in one (14%) of seven patients with SSc. The manometric results of individual patients are summarized in Table 4.

Esophageal manometry and its relation to anorectal manometry

SSc patients also showed abnormal esophageal manometry findings, notably decreased LES pressure and body amplitude of distal 2/3 esophagus. The comparision between manometric profiles of anorectum and esophagus showed no significant correlation by statistical analysis (Table 5).

Table 3. Anorectal manometric results in patients with systemic sclerosis and controls

	SSc (n=7)	Control (n=11)	P-value
Minimal Sensory Volume (ml)	35.0 ± 4.5	48.2 ± 6.8	$p < 0.05$
Urgency Volume (ml)	87.5 ± 29.5	118.2 ± 29.5	$p < 0.05$
Maximal Tolerated Volum (ml)	143.3 ± 32.8	171.4 ± 39.5	NS
Anal Canal Length (cm)	3.3 ± 0.4	3.6 ± 0.5	NS
Anal Sphincter			
Resting Pr (mmHg)	76.6 ± 19.0	79.3 ± 15.5	NS
Squeezing Pr (mmHg)	90.3 ± 64.9	103.8 ± 47.0	NS
RAIR (%)	46.6 ± 28.6	56.4 ± 12.2	NS

RAIR : Rectoanal inhibitory reflex
NS : Not significant

Table 4. Clinical and anorectal manometric data of individual patients with systemic sclerosis

Patient No	LGI Sx	Rectal volume (ml)			AS Pr (mmHg)		RAIR (%)
		MSV	UV	MTV	Resting	Squeezing	
1	–	40	75	115	100	70	31
2	IE, Tenesmus	30	70	140	74	121	31
3	–	35	95	165	81	222	64
4	IE, Constipation	35	90	150	103	79	51
5	IE, FI	30	55	100	59	53	61
6	–	35	87	143	60	27	88
7	IE, Constipation	40	140	190	59	56	0

LGI Sx, Lower gastrointestinal symptom
AS Pr, Anal sphincter pressure ; RAIR, Rectoanal inhibitory reflex
MSV, Minimal sensory volume ; UV, Urgency volume ; MTV, Maximal tolerated volume
IE, Incomplete evacuation ; FI, Fecal incontinence

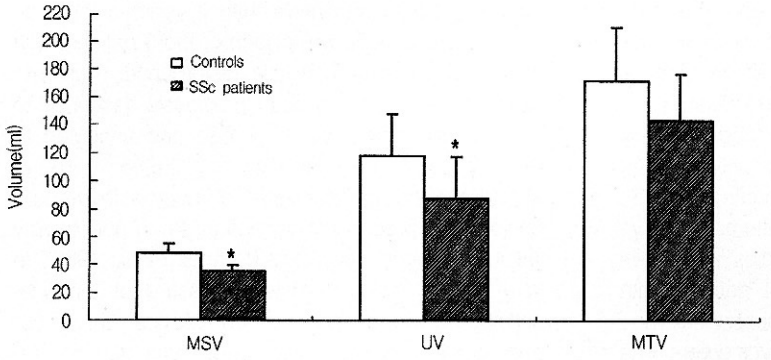

Fig. 1. Rectal sensation of balloon inflation in patients with systemic sclerosis (SSc) and in controls.
MSV : minimal sensory volume, UV : urgency volume, MTV : maximal tolerated volume. * : $p<0.05$

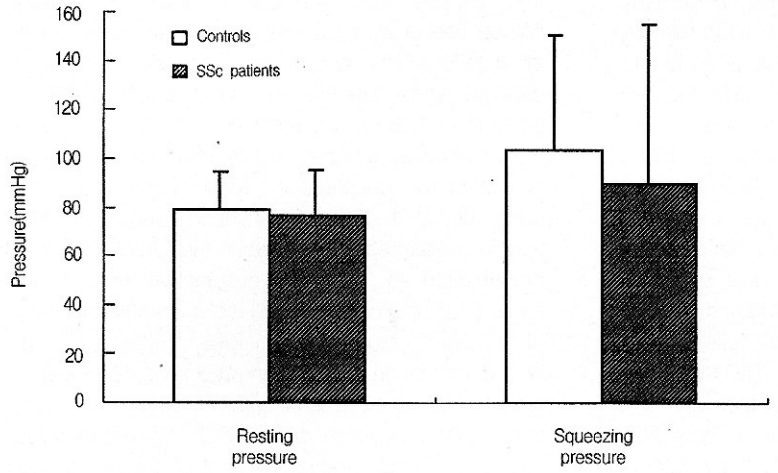

Fig. 2. Resting and squeezing pressure of anal sphincter in patients with systemic sclerosis (SSc) and in controls. There was no significant difference in these two groups.

Table 5. Anorectal and esophageal manometric results in patients with systemic sclerosis

Patient No	Anal sphincter		RAIR (%)	LES Pr (mmHg)	Contraction amplitude of distal 2/3 esophagus (mmHg)
	Resting Pr (mmHg)	Squeezing Pr (mmHg)			
1	100	74	31	8	26.3
2	74	121	31	10	7.7
3	81	222	64	11	5.8
4	103	79	51	10	22.1
5	59	53	61	13	24.3
6	60	27	88	28	20.7
7	59	56	0	24	16.7

RAIR : Rectoanal inhibitory reflex
LES Pr : Lower esophageal sphincter pressure

DISCUSSION

Replacement of the esophageal and gastrointestinal muscularis propria by fibrous tissue is well recognized in SSc (D'Angelo et al., 1969; Cohen et al., 1980). Clinically symptomatic and significant gastrointestinal involvement occurs in approximately 50% of all patients with SSc(Cohen et al., 1980; Sjogren, 1994). In addition, many patients with SSc who do not have gastrointestinal symptoms will have subclinical involvement. Careful studies have revealed that as many as 75-90% of patients with SSc have abnormalities noted on esophageal motility testing, although in many, the involvement was not clinically significant (Turner et al., 1973; Akesson and Wolheim, 1989). Involvement of the anorectum was the next most frequent, occuring in 50-70% (Hamel-Roy et al., 1985). Physiologic abnormalities of the anorectum may be found in SSc even before clinical symptoms and recent studies have focused on anorectal physiological findings in patients with SSc (Hamel-Roy et al., 1985; Chiou et al., 1989; Basilisco et al., 1992; Leighton et al., 1993; Engel et al., 1994).

The physiological findings previously described in patients with SSc include reduced anal sphincter resting pressure (Whitehead et al., 1989; Engel et al., 1994), impaired rectoanal inhibitory reflex (RAIR) (Hamel-Roy et al., 1985; Pemberton, 1990; Leighton et al., 1993) and reduced rectal compliance and capacity(Whitehead et al., 1989). In our patients with SSc, abnormal manometric findings (either low resting pressure of anal sphincter, impaired rectal capacity, and/or weak RAIR) in 5(71%) of 7 patients. Both neuropathic and myopathic processes appear to be involved in the pathogenesis of gastrointestinal manifestations in SSc. It has been suggested that gastrointestinal hypomotility initially is based on a neural defect, whereas in late disease dysfunction is the result of smooth muscle atrophy and fibrosis (Sonnex et al., 1986; Greydanus and Camilleri, 1989).

An absent or diminished RAIR was described to be the most common manometric abnormality, accounting for 71 and 50 percent of patients of SSc in two recent studies (Hamel-Roy et al., 1985; Leighton et al., 1993). In our study, the reflex was absent or blunt in 3 (43%) of 7 patients. Considering the duration of the disease and comparing with the previous reports in which the duration was not mentioned in detail, our study revealed that 3(75%) of 4 patients of SSc with more than 2 years of duration had impaired RAIR, while none of the 3 patients with less than 2 years had this disturbance. This impairment of reflex has been previously described to occur in the early stages of SSc and attributed to neural dysfunction(Hamel-Roy et al., 1985; Chiou et al., 1989). The pathogenesis of neuropathy is still unclear, but probably is secondary to an abnormality within the myenteric plexus (Sonnex et al., 1986). In later stages reduced resting pressure of the anal sphincter and reduced rectal compliance can occcur and these changes have been attributed to the infiltrative process affecting the muscularis propria of the rectum, that is muscular dysfunction resulting from collageneous replacement of smooth muscle (Whitehead et al., 1989; Basilisco et al., 1992; Engel et al., 1994). Reduced resting pressure and rectal capacity were observed in 3(43%) and 2(29%) of 7 patients of SSc in our study.

Anorectal dysfunction in SSc may result in fecal impaction or constipation, fecal incontinece and rectal prolapse. Fecal incontinece has been previously described as probably the most common presentation of anorectal dysfunction and to be related to weakness of anal sphincters(Hamel-Roy et al., 1985; Leighton et al., 1993; Engel et al., 1994). Constipation or fecal impaction is also associated with impaired anal sphincter function(Cohen et al., 1980; Basilisco et al., 1992). Weakness of the submucosa and chronic straining of the rectal wall and internal anal sphincter may contribute to the development of rectal prolapse (Leighton et al., 1993). Pressure of a rectal prolapse further decreases anal sphincter pressure and can contribute to incontinence(Leighton et al., 1993). In our patients of SSc, 4 of 7 patients had symptoms referrable to anorectal involvement All the four patients complained of fecal impaction or incomplete evacuation, two of them also had symptoms of constipation, but only one of them complained of fecal incontinence.

In the disease course of SSc, the changes detected by anorectal manometry occur much earlier than the deterioration of anorectal function clinically. Consistent with a previous report(Chiou et al., 1989), three of our 7 patients of SSc had no symptoms concerning anorectal involvement, but abnormal manometric findings already had been discovered in

two of them. On the other hand, we observed one patient who complained of constipation and yet her manometric findings revealed normal anorectal function. The physiologic cause of constipation in this patient may be due to colonic dysfunction rather than anorectal dysfunction. Careful manometric studies in a previous report (Hamel-Roy et al., 1985) have documented that patients with abnormal esophageal manometry almost always also have abnormal anorectal motility. Consistent with this report, five of our 7 patients with SSc had disturbed esophageal motility and coexisting anorectal motility disturbances were observed in 4 of them. In the series of Hamel-Roy et al.(1985), there was a correlation between the amplitude of the lower esophageal sphincter relaxation and the amplitude of the rectoanal inhibitory reflex in response to rectal distention. This finding was not consistent with our results which showed no significant correlation between the degrees of esophageal and anorectal manometric abnormalities.

Our data suggest that manometric abnormalities of the anorectum correlate well with the duration of the disease, but poorly with the symptoms. None of the 3 patients of SSc with less than 2 years of duration had any manometric abnormalities, while all of the 4 patients with more than 2 years had at least one abnormality of manometric profiles. Most of the previous reports did not mention in detail the correlation between the severity of manometric profiles and that of symptoms except one(Chiou et al., 1989) in which a correlation between reduced resting pressure of the anal canal and severity of fecal incontinece was suggested.

In conclusion, these findings suggest that anorectal function may be impaired in patients with SSc which can reflect the involvement of the anorectum in the disease, and that anorectal manometric studies can be useful to detect such dysfunction in patients with SSc, even before clinical symptoms.

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