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agranulocytes and anti-centromere with titers > 1/1000. A posi-
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SUMMARY
Endoanal sonography is now an effective tool for the assessment of ano-rectal pathologies. We present a case of rectal prolapse in a patient with progressive systemic sclerosis, with low resting anal pressure, no rectoanal inhibitory reflex in manometry, and a thin, heterogeneous, difficult to delineate, internal sphincter on endoanal ultrasound. We also provide a review of the literature on ano-rectal involvement in progressive systemic sclerosis.

RÉSUMÉ
Aspect échoendoscopique du sphincter anal interne au cours de la sclérodermie
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L’échoendoscopie est un examen indispensable pour évaluer certaines pathologies ano-rectales. Nous rapportons le cas d’une malade atteinte de sclérodermie systémique avec un prolapsus rectal symtomatique. La manométrie objectivait un pression anale de repos basse et une absence de réflexe anorectal inhibiteur. L’échoendoscopie endoanale mettait en évidence un aspect fin, hétérogène et difficile à délimiter du sphincter anal interne. Nous avons réalisé une analyse détaillée de la littérature de l’atteinte ano-rectale au cours de la sclérodermie systémique.

Introduction
Progressive systemic sclerosis (PSS) is a systemic disease associated with alterations in the microvasculature, the auto-nomic nervous system, and the immune system, leading to fibro-sis. Nearly 90% of patients have some degree of gastrointestinal (GI) involvement, and approximately one-half of them are symp-tomatic. Severe GI problems, including malabsorption and intesti-nal pseudo-obstruction, are much less common; affecting fewer than 10% of patients during the first nine years of this illness [1]. The first visceral manifestation to be described was esophageal disease, which remains the most common source of GI symptoms in scleroderma. It has since been demonstrated that any part of the GI tract (from mouth to anus) may be involved. Nevertheless, anal incontinence is less well recognized and its incidence is unknown in large series [2]. We present a case of PSS associated with anal incontinence and rectal prolapse, and further discus-anorectal involvement in PSS.

Case report
A 63 year old woman was diagnosed with CREST syndrome in 1995. She had had Raynaud’s syndrome since 1992, as well as chronic heartburn with no dysphagia. Upper GI endoscopy revealed a grade B esophagitis (Los Angeles classification). Esophageal manometry revealed aperistalsis of the lower two thirds and a hypotonic lower sphincter typi-cal of PSS. There were also cutaneous plantar calcifications, dorsal telangiectasias, and sclerodactyly. Autoimmune work up only found positive antinuclear antibodies and anti-centromere with titers > 1/1000. A posi-tive Shimer test confirmed associated Sjogren syndrome. There was no
evidence of cardio-pulmonary, articular or renal involvement. She was treated with proton pump inhibitors, calcichine, calcium channel blockers, and artificial tears.
In 2002, she complained of a two year history of progressive pass-sive anal incontinence to mucus (1-2 episodes per week), flatus (once every ten days) and more rarely liquid stools. However, she wore no pro-tection and reported a fair well being despite her symptoms. She also complained of worsening dyschesia. She had one hard stool every two to three days, with straining and feeling of incomplete evacuation, not fully relieved by perianal digital maneuvers. There was no diarrhea, no abdominal pain or uro-genital symptoms. She had no other medical history except one uncomplicated vaginal delivery.
Proctologic examination found a dilated anus, a fair voluntary contrac-tion and no clinical anismus. Lower rectal mucosa was normal on rectoscopy with no evidence of solitary rectal ulcer. A complete rectal prolapose was externally visible on straining in the squatting position. Gynecological examination was normal. Colonoscopy was normal. Ano-rectal manometry was performed using a perfused catheter with a station-ary pullthrough technique. It revealed low resting anal pressure (25 mmHg), a squeeze pressure of normal amplitude (90 mmHg) and duration (32 seconds), a normal anal canal length, and a normal rectal sen-sitivity. Rectal distension volumes eliciting a first perception, an urge to defecate, and maximally tolerable were respectively 20 mL, 80 mL, and 240 mL of air. Progressive inflation of the intra-rectal balloon (10-60 mL of air) failed to elicit any rectoanal inhibitory reflex (RAIR). There was no manometric evidence for anismus. Endoanal ultrasound (figure 1) using a Bruel and Kjaer transducer found a thin, heteroge-neous, difficult to delineate, internal sphincter, a normal external anal sphincter and a normal puborectalis muscle. Rectal mucosa presented a normal ultrasound structure. Fistulography showed a rectal prolapse of 25 mm length during straining originating from the lower rectum, asso-ciated with a mild enterocoele. There was no evidence of rectoceles or significant perineal descent. Rectal evacuation of contrast media was complete. There was no evidence of paradoxical contraction of the anal sphincter or the puborectalis during evacuation. Pudendal nerve terminal motor latencies were normal and external anal sphincter electromyogra-phy showed no evidence of anismus or neuropathy.
The patient declined surgical repair of the rectal prolapse (rectopexy) and was offered a trial of dietary fibers and glycerine suppositories. This led to a significant improvement of her dyschesia. Moreover, she had no soiling episodes of liquid stools but still reported occasional incontinence to mucus or flatus.
Discussion

GI manifestations of PSS have been recently reviewed [3]. D’Angelo et al. were the first to publish a case report suggesting PSS as a causative factor of rectal prolapse [4]. Over the past few years, endoscopic ultrasound has been shown to be the most accurate method to assess the anatomic integrity of the anal sphincter (figure 2). Engel et al. [5] were the first to provide endoanal ultrasound evidence for internal anal sphincter fibrotic involvement in two scleroderma patients presenting with anal incontinence, low resting pressure and absent RAIR. The internal anal sphincter was thin and hyperechoic in both patients, with an intact external sphincter, as in our patient (figure 1). Anorectal function abnormalities in PSS were reported in 1985, by Hamel-Roy et al. [6]. The RAIR was of lesser amplitude than normal in 74% of patients with PSS and was absent in 13%. They concluded that anorectal motility was abnormal as frequently as esophageal motility, with a significant correlation between the amplitude of lower esophageal sphincter relaxation and the amplitude of the RAIR in response to rectal distension with 30-50 mL of air [6]. This finding can be easily understood because of the functional and histological similarities between lower esophageal and internal anal sphincters, when they undergo the fibrous transformation characteristic of PSS. In early phases of the disease, fibrous connective tissue proliferation in the rectal submucosa and muscularis mucosa leads to reduced rectal compliance, diminished or absent RAIR, as well as low resting pressure. In later phases, smooth muscle ischemic atrophy results in anal incontinence. In their series of seventeen scleroderma patients evaluated with anorectal manometry, Chiou et al. [7] found a significant decrement in resting pressure in the study group compared to age-matched normal subjects. Twelve (71%) of their 17 patients had absent RAIR, and paradoxical contraction of the anal sphincter during rectal balloon inflation was noted in ten patients. Nine patients had different degrees of anal incontinence and abnormal manometric profiles were found in six of eight asymptomatic patients. Therefore, only two patients (12%) had neither symptoms nor manometric evidence of anorectal involvement. The difference in the functional length of the anal canal, the volume of the first defecating sensation, the voluntary component, the difference between squeeze pressure and resting pressure, were also found to be statistically significant. Paradoxical contraction of the anal sphincter can be found in patients who still have normal resting pressure, if the autonomic neuropathy rather than muscle fibrosis is the main insult in early PSS [8]. In their series of eight scleroderma patients, Leighton et al. [9] found abnormal resting pressure and reduced rectal capacity in all patients, with four patients having associated second degree complete rectal prolapse, as well as lower anal pressures. Absent RAIR was also found in half of the studied group. The fibrous infiltrative process involving rectal muscularis propria as well as weakness of the submucosa during straining accounts for rectal prolapse in scleroderma patients, as well as for abnormal rectosigmoid motility such described by Whitehead et al. [10]. Rectal prolapse worsens anal incontinence by reducing rectal compliance and contributing to low resting pressure. It should therefore be corrected surgically either by a transanal approach or anterior rectal resection of the prolapsed mucosa, thus enhancing temporarily the patients continence and quality of life. Improvement in continence lasted one week, six months, and seven years in the three patients operated for rectal prolapse by Leighton et al. [9], but incontinence recurred due to progressive fibrosis of the anal sphincter.

We think that our patient has anorectal involvement of PSS because of low resting anal pressure, absent RAIR and, above all, an abnormal appearance of the internal anal sphincter at ultrasound. Admittedly, the low resting anal pressure and the absent RAIR may be explained by a non specific rectal prolapse [11, 12]. On the other hand, this unusual ultrasound aspect may be a specific feature of progressive systemic sclerosis as reported by Engel et al. [5]. These authors described two patients with a very thin internal anal sphincter on endosonography (identical to our case). This atrophy was assumed to have been caused by fibrous replacement of the sphincter that could be partly ischaemic in origin [5]. Moreover, several authors have demonstrated that internal anal sphincter thickness was greater in patients with non specific rectal prolapse than in controls [13-15]. So the debate is not closed…

ABBREVIATIONS:
PSS : progressive systemic sclerosis
GI : gastrointestinal
RAIR : rectoanal inhibitory reflex

Fig. 1 – Abnormal internal anal sphincter in progressive systemic sclerosis (arrows) (Bruel and Kjaer transducer).

Sphincter interne anormal chez une patiente atteinte de clérodermie systémique (Sonde d’échographie de Bruel et Kjaer).

Fig. 2 – Normal middle anal canal, showing the internal and external anal sphincters (Bruel and Kjaer transducer).

Canal anal moyen d’aspect normal objectivant les sphincters internes et externes (Sonde d’échographie de Bruel et Kjaer).
Conclusion

Although it has not been extensively studied, anorectal involvement is well established in the spectrum of GI manifestations of PSS. Gastroenterologists should therefore suspect this manifestation, especially if there is esophageal involvement. Manometry and endoanal ultrasound should be used. A low resting anal pressure, an absent RAIR and, above all, an atrophic internal anal sphincter on endoanal ultrasound suggests a PSS involvement. Rectal prolapse should also be looked for since it is a surgically correctable factor of anal incontinence.

REFERENCES