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## Scleroderma, myositis and related syndromes

### AB0716 AN AUTOPSY CASE OF SYSTEMIC SCLEROSIS WITH SEVERE INTESTINAL INVOLVEMENT AND LITERATURE REVIEW

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**Background:** The gastrointestinal tract (GIT) is the second most common internal organ affected by systemic sclerosis (SSc). The rate of SSc patients who develop severe GIT symptoms is lower than 10%, although various degrees of chronic intestinal pseudo-obstruction (CIPO) may occur in as many as 40% of cases (1.2).
**Objectives:** To report an autopsy case of SSc with severe intestinal involvement and review the associated literature.
**Methods:** We will present the clinical features and autopsy findings of a SSc patient and literature concerning Japanese SSc autopsy cases associated with severe intestinal involvement, found *Igaku-chuo* and *Pub-med* on Internet.
**Results:** A 69-year-old Japanese woman was diagnosed with diffuse cutaneous SSc from skin sclerosis, Raynaud's phenomenon, and mild interstitial pneumonia in January 2013. The antinuclear antibody was positive (1:160, speckled pattern), but the specific antibodies, including the anti-RNP, topoisomerase I, and centromere antibodies, were negative. In August 2015, at the age of 71, she was hospitalised for vomiting and abdominal pain. Plain abdominal radiograph showed dilation of the small bowel with air-fluid levels. Abdominal CT revealed large dilation of the small bowel in the absence of any mechanical obstruction. These findings were consistent with CIPO. Her symptoms soon improved by decompression with a long intestinal tube. But she experienced frequent relapse of CIPO. During the third hospitalisation in May 2016, an abdominal CT showed pneumatosis cystoides intestinalis (PCI) and free air in the peritoneal cavity. Medical management failed to control the CIPO. Her general conditions had gradually worsened with weight loss of 10 kg in 3 years. Home parental nutrition was initiated in January 2017. On May 2017, she developed severe pneumonia after vomiting, and her condition gradually deteriorated. She finally succumbed to her illness and an autopsy was performed. The whole alimentary tract except for the duodenum showed a thinning of the lamina propria and atrophy of the smooth muscular layers. Intimal proliferation and narrowing of arterioles were also noted. There was non-specific interstitial pneumonitis in the both lower lobes and diffuse alveolar damage in the both of upper lobes of the lungs. Vasculopathy was also seen in the lungs and heart.
The cases in the literature are summarised in table 1. Vascular damage and/or smooth muscle atrophy were presented in all cases.

**Abstract AB0716 – Table 1**. Autopsy cases of systemic sclerosis associated with severe gastrointestinal symptoms in Japan

| Case No. | Publication year | Age of death (yr) / Sex | Time from onset of * to death | | Cause of death | Comorbidity / Past history | GIT symptoms | Pathological findings of intestine | | |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | *Raynaud' phenomenon | *GIT symptoms | | | | Vascular damage[1)] | Muscle atrophy | Fibrosis |
| 1 | 1973 | 49/M | 1 yr 6mo | 5mo | respiratory failure | Silicosis, IP | Malnutrition | ○ | ○ | |
| 2 | 1976 | 32/F | 12 yr | 1y 3mo | general prostration | IP | Severe constipation | ○ | ○ | ○ |
| 3 | 2000 | 60/F | 5 yr 4mo | 4y 5mo | Sepsis | Polymyositis | CIPO, PCI | | ○ | ○ |
| 4 | 2001 | 72/F | 1 yr | 2 weeks | GIT bleeding | Hypertention, Renal failure | Bleeding | ○ | | |
| 5 | 2002 | 58/F | 3 yr 7mo | 2 weeks | Pneumonia | Dermatomyositis,IP | CIPO, PCI | | ○ | ○ |
| 6 | 2007 | 71/F | 6 mo | 2 mo | Septic shock | Renal failure/gastric and lung cancer | CIPO | ○ | | |
| 7 | this report | 72/F | 4 yr | 1y 10mo | Pneumonia | IP / breast cancer | CIPO, PCI | ○ | ○ | |

M, male; F, female; yr, years; mo, months; GIT, gastrointestinal tract; IP, interstitial pneumonia; CIPO, chronic intestinal pseudo-obstruction; PCI, pneumatosis cystoides intestinalis;
1) Intimal proliferation and narrowing of the small arteries
**Conclusions:** Vasculopathy in SSc involves small vessels, and it precedes fibrosis.[3] The triggering event of vasculopathy is unknown, but the narrowing of intestinal arterioles causing hypoxia might be responsible for dysmotility of GIT.

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### AB0717 RISK ASSOCIATION BETWEEN SCLERODERMA DISEASE CHARACTERISTICS, PERIODONTITIS AND TOOTH LOSS

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**Background:** Systemic Sclerosis (SSc (is a multi-system disorder that can have significant adverse effects upon the health of the mouth.
**Objectives:** The aim of this study was to investigate the associations between the disease characteristics of SSc, periodontal disease (PD) and tooth loss.
**Methods:** Fifty-four patients affected by SSc and 55 non-diseased controls were matched for age and gender. SSc was characterised in subtypes and with the mean duration of disease and the Modified Rodnan Skin Score [mRSS]. Patients were surveyed and examined through the evaluation of the periodontal parameters and the number of teeth.
**Results:** A logistic regression analysis showed that patients with SSc presented a higher number of missing teeth
(p=0.001) and a significant median increased odds 2.95 (95% CI 1.26 to 6.84) of PD (defined as clinical attachment loss, CAL) compared to non-diseased controls (6.83, 95% CI 1.94 to 24.36). Moreover, the less values of PD was correlated with mRSS in the total SSc group and with the mean duration of disease in patients with limited SSc
(p=0.007), even after adjusting this correlation with the presence of the major organs involvement.
**Conclusions:** This study showed that patients with SSc presented an increased odds of PD and tooth loss compared to non-diseased controls. In SSc patients, the magnitude of PD was strongly associated with the mRSS and with the mean duration of the disease. The clinicians should be aware of the potential systemic health problems related to PD.

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### AB0718 AUTOLOGOUS HEMATOPOIETIC STEM -CELL TRANSPLANTATION IN SEVERE AND REFRACTORY DIFFUSED SYSTEMIC SCLEROSIS. LONG-TERM STUDY IN A SINGLE REFERENCE CENTRE

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**Background:** Pharmacological therapy does not reverse the natural course of systemic sclerosis (SS). Autologous hematopoietic stem-cell transplantation (AHSCT) has been used in refractory autoimmune diseases, including SS. The autologous hematopoietic stem-cell transplantation is based on the ablation of the immunoreactive system with high doses of chemotherapy and a subsequent regeneration of the immune system.
**Objectives:** Study of the efficacy of autologous hematopoietic stem-cell transplantation in a cohort of patients with SS from a referral centre.
**Methods:** Descriptive study of patients with SS (ACR/$^{EULAR\ 2013}$ criteria) treated with autologous hematopoietic stem-cell transplantation in a single reference centre between 2002 and 2017. These clinical variables were reviewed

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