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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's 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	Hypertension, 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

(Raynaud's phenomenon, pulmonary interstitial involvement, digital ulcers, digestive alterations, presence of sclerodermic renal crisis) and activity index variables (modified Rodnan score, HAQ-DI, SHAQ-VAS).

Results: Four patients were included (75% women). The median age at the time of the AHSCT was 36.5 years (range 27–51). In all cases, the initial diagnosis was diffuse cutaneous ES, refractory to corticosteroids and at least one DMARD. Prior to autologous hematopoietic stem-cells transplantation, the clinical manifestations presented were a) severe Raynaud's phenomenon (100%) with significant joint and cutaneous involvement; b) digital ulcers (50%); c) interstitial lung disease (50%) and d) sclerodermic renal crisis (25%). In 3 of the cases (75%) the antitopoisomerase antibodies were positive. The conditioning treatment for the autologous hematopoietic stem-cells transplantation was cyclophosphamide at high doses (50 mg/kg x4 days) and anti-thymocyte globulin. In 3 patients (75%) there were slight post-transplant complications (febrile neutropenia, diarrhoea) after a median follow-up of 6.5 years (range 1–15).

The response to AHSCT is summarised in table 1. All patients showed values <1 in the Health Assessment Questionnaire on the Disability Index (HAQ-DI), in 75% with a modified Rodnan skin score (mRSS) lower than 7.

Abstract AB0718 – Table 1

PATIENT	FOLLOW-UP	MODIFIED RODNAN SKIN SCORE PRE-TRASPLANT	MODIFIED RODNAN SKIN SCORE POST TRASPLANT	HAQ-DI POST TRASPLANT	SHAQ-VAS POST TRASPLANT
Patient 1	15 years	Unknown	1/51	0,25	4/13
Patient 2	8 years	Unknown	7/51	0	3/13
Patient 3	5 years	28/51	6/51	0	0/13
Patient 4	1 year	33/51	20/51	0,50	7/13

Conclusions: Autologous hematopoietic stem-cells transplantation can be a therapeutic option in refractory and severe SS. These hopeful data must be ratified in larger studies.

Disclosure of Interest: None declared

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AB0719 CLOSE TEMPORAL ASSOCIATION BETWEEN SILICONE COSMETIC SURGERY AND SYSTEMIC SCLEROSIS ONSET

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Background: The pathogenesis of systemic sclerosis (SSc) still remains unclear, however it is increasingly thought to result from interactions between environmental factors and epigenetic features leading to the onset and progression of SSc in genetically susceptible patients.¹ Case reports of women with silicone breast implants who developed SSc have been published, but several case-control series and prospective studies in connective tissue diseases (including SSc) failed to find an increased risk of SSc associated with silicone cosmetic surgery.² However, several bias may be recognised in these studies, i.e. heterogeneous cohorts of enrolled patients not selective for SSc, non homogeneous either disease duration or disease stage at study entry. For these reason the possible effect of silicone implants as immune adjuvants is not clear.³

Objectives: Retrospective study to find out patients who developed SSc after cosmetic surgery.

Methods: The clinical files of 110 female patients with systemic sclerosis were retrospectively evaluated. Among these, four patients showing a history of silicone cosmetic surgery (3.6%) were identified, and clinical data collected.

Results: The clinical data of the four patients are below reported. 1. LS 28 year old female who underwent cosmetic breast prosthesis: two years later she complained of Raynaud's phenomenon (RP), and one more year later aggressive diffuse cutaneous SSc, along with anticentromere antibodies (ACA) positivity. 2. PJ 38 year old female who underwent cosmetic breast prosthesis: one year later she experienced RP and one more year later aggressive diffuse cutaneous SSc; anti-nuclear antibodies were positive with a speckled pattern, but specific SSc-related autoantibodies negative. 3. BS 33 year old female who underwent cosmetic breast prosthesis: two years later she complained of RP and one more year later limited cutaneous SSc with ACA positivity; SSc clinical condition partially improved and its progression stopped after breast prosthesis removal. 4. CM 58 year old female who underwent cosmetic lip silicone application: one year later she complained simultaneous onset of RP and aggressive diffuse cutaneous SSc with anti-Topoisomerase positivity; she died during follow-up.

Conclusions: This study reports a prevalence of 3.6% of silicone cosmetic surgery before SSc onset. The close temporal association between silicone implant

and disease development suggests a possible role of silicone in SSc pathogenesis. Specifically addressed clinical studies or big-data studies need to rule out this matter.

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AB0720 SYSTEMIC SCLEROSIS AND CANCER DEVELOPMENT. A SINGLE-CENTRE EXPERIENCE

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Background: Systemic Sclerosis (SSc) is an autoimmune connective tissue disease with multisystem involvement, and sometimes devastating results. In bibliography there are reports that scleroderma patients present a higher incidence of risk for cancer when compared with the general population. However, different estimates have been reported.

Objectives: The purpose of the present study was to evaluate the frequency of cancer development (CD) in a cohort of patients with SSc.

Methods: Patients that fulfilled the 2013 American College of Rheumatology/European League Against Rheumatism criteria for SSc and were followed up since 1999, were included. Date of disease onset, disease duration, autoantibodies, age, pulmonary hypertension, comorbidities and the type of CD have all been taken into account, during the period 1991–2016.

Results: Seventy-nine SSc patients have been included. 46 with limited (lcSSc) and 33 with diffuse cutaneous Systemic Sclerosis (dcSSc). Six of them, (7.6%) developed different types of cancer. Most of them were adenocarcinoma. More specifically, 2 developed pulmonary adenocarcinoma (1 with lcSSc and 1 with dcSSc), 1 follicular carcinoma of the thyroid gland (lcSSc), 1 colorectal adenocarcinoma (lcSSc), 1 B-cell lymphoma (MALT lymphoma), and 1 prostate adenocarcinoma (dcSSc). Five out of six were female patients. Mean age at the time of cancer diagnosis was 66.8-years-old, while SSc has been diagnosed at the mean age of 49.4 years. Mean time of developing any type of cancer was 15.8 years after SSc diagnosis. The diagnosis of cancer was done the last 20 months. All patients were non-smokers, had gastro-oesophageal reflux disease and pulmonary fibrosis, while 4/6 had also pulmonary hypertension and were under treatment with phosphodiesterase 5 inhibitors and bosentan. Scleroderma patients with CD have been referred to the corresponding oncology clinic for further treatment.

Conclusions: The present study on SSc and CDE provides data showing a potential association between the two entities. We found a high frequency of cancer development in patients with SSc (7.6%). Thus, a careful monitoring and screening is required when physicians follow-up scleroderma patients.

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AB0721 TRANSFER OF SYSTEMIC SCLEROSIS AFTER ALLOGENEIC BONE MARROW TRANSPLANTATION

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Background: It is accepted that donor-derived immunity is transferred with allogeneic bone marrow transplantation (BMT)¹.

Objectives: To show evidence of the transfer of systemic sclerosis by allogeneic BMT.

Methods: In this report we describe a patient with T acute lymphoblastic leukaemia who underwent BMT and developed systemic sclerosis.

Results: 34-year-old man in complete remission from a T acute lymphoblastic leukaemia treated with allogeneic BMT from his mother in february of 2012.

First seen in november 2017 for digital ulcers that appeared one year before.

He presented two necrotic ulcers: one on the second finger of the left hand and other on the third finger of the right hand (IMAGE 1). He was admitted to receive intravenous prostaglandins and complete the study.

After the BMT he developed Raynaud's phenomenon and in the examination he only presented facial and corporal telangiectasia, attributed before to chronic graft versus host disease (cGVHD).