MONOCYTES TO LYMPHOCYTES RATIO IS CORRELATED WITH DISEASE ACTIVITY IN BEHÇET’S DISEASE

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Background: Behçet’s disease (BD) is a complex, inflammatory multisystem disorder. Since the lack of universally recognised pathognomonic laboratory test, the diagnosis relies heavily on clinical findings. Currently, the Monocytes to lymphocytes to ratio (MLR), Neutrophils to Lymphocytes ratio (NLR), Platelets to Lymphocytes ratio (PLR) and Red blood cell Distribution Width(RDW) have been demonstrated as a assessment of disease severity in many rheumatism diseases. Nevertheless, to our knowledge, only a few studies have investigated NLR, PLR, RDW in patients with BD.

Methods: A total of 37 patients with BD fulfilling the criteria of the International Study Group for BD and 37 age and gender-matched healthy controls were enrolled in the study retrospectively. MLR, NLR, PLR, RDW, C-reactive protein (CRP) level and Erythrocyte Sedimentation Rate(ESR) level were evaluated. The correlation between the variables were tested with Pearson correlation. Area Under Curve(AUC) value, sensitivity, specificity, and the optimal cut-off values were determined using Receiver Operating characteristic Curves (ROC). According to the optimal cut off value, BD patients were divided into low-value group (<the optimal cut off value) and high value group (>the optimal cut off value). The patient’s clinical characteristics between the two groups were compared.

Results: The MLR, NLR, PLR and RDW were (0.37±0.24), (2.91±1.95), (155.09±55.08) and (13.83±7.17) in BD group, while (0.18±0.04), (1.45±0.46), (115.66±28.01) and (13.07±1.19) in control group, the difference was significant (P all<0.05). MLR, NLR and PLR were all correlated positively with ESR(r=0.363, P<0.05; r=0.611, P<0.05; r=0.496, P<0.05) and CRP(r=0.713, P<0.05; r=0.785, P<0.05; r=0.765, P<0.05; r=0.394, P<0.05). RDW was not correlated with ESR and CRP. ROC curve results showed that the AUC of MLR, NLR, PLR and RDW for BD were 0.841(CI95%: 0.748–0.935), 0.815(CI95%: 0.712–0.918), 0.720(CI95%: 0.699–0.840), 0.635(CI95%: 0.505–0.765), MLR yielded a highest AUC. In addition, the optimal cut off value of MLR for BD was 0.23, with the specificity of 73.0% and sensitivity of 83.8%. In 37 BD patients, 14 belong to low MLR group, 23 belong to high MLR group. The comparison results show that high MLR value group have higher CRP level and higher incidence of genital ulceration(P<0.05).

Conclusions: MLR was elevated in BD patients as compared to control group, having a close relationship with disease activity.

REFERENCES:

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Disclosure of Interest: None declared


GIANT CELL ARTERITIS IS COMORBID WITH TUBERCULOSIS

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Background: Giant cell arteritis (GCA) is a medium- and large-vessel vasculitis with an onset age after 50 years, whereas Takayasu arteritis (TA) is a rare large-vessel vasculitis with an onset age younger than 40 years. The association between TA and tuberculosis (TB) was suggested. However, the association between GCA and TB was rarely reported.

Objectives: To understand the association between TA and TB

Methods: Clinical data between November 1998 and October 2017 at PUMCH, Beijing, China, were retrospectively reviewed. Ninety-one patients diagnosed with GCA were included in the study. Precise clinical data were collected and analysed.

Results: A total of 20 patients (22.0%) had a history of active tuberculosis and received anti-tuberculosis therapy. On comparing the clinical features of the patients with TB and those without TB, obvious weight loss (p<0.011), lower percentage of dyslipidemia (p<0.042), higher percentage of anti-phospholipid (p<0.010), and lower white blood cells (p<0.006) were noted in the TB group.

Abstract AB0715 – Table 1. Clinical features and comorbid diseases of the patients with TB and without TB

<table>
<thead>
<tr>
<th></th>
<th>GCA with TB</th>
<th>GCA without TB</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (year, diagnosis)</td>
<td>65.10±6.39</td>
<td>65.38±7.51</td>
<td>0.886</td>
</tr>
<tr>
<td>Scap tenderness or pain</td>
<td>2 (10)</td>
<td>22 (31.0)</td>
<td>0.060</td>
</tr>
<tr>
<td>Tenderness and abnormal pulsation of temporal artery</td>
<td>6 (30)</td>
<td>13 (18.3)</td>
<td>0.256</td>
</tr>
<tr>
<td>Visual loss</td>
<td>8 (40)</td>
<td>25 (35.2)</td>
<td>0.694</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>3 (15)</td>
<td>20 (28.2)</td>
<td>0.231</td>
</tr>
<tr>
<td>Jaw claudication</td>
<td>6 (30)</td>
<td>20 (28.2)</td>
<td>0.873</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>11 (55)</td>
<td>36 (50.7)</td>
<td>0.734</td>
</tr>
<tr>
<td>GI symptoms</td>
<td>5 (15)</td>
<td>11 (15.5)</td>
<td>0.221</td>
</tr>
<tr>
<td>Weight loss</td>
<td>16 (80)</td>
<td>34 (47.9)</td>
<td>0.011</td>
</tr>
</tbody>
</table>

Conclusions: This study demonstrated that the percentage of TB history in patients with GCA was higher that than in the general population. The definite association between TB and GCA remains unknown. Hence, further studies are required to elucidate the mechanisms underlying TB in the pathogenesis of GCA.

Clinicians should recognise the possibility of comorbid TB in patients with active GCA, and take appropriate measures to control weight loss and lower white blood cell count.

REFERENCES:

Acknowledgements: We thank all the physicians from department of internal medicine of PUMCH participated in the caring of this patients.
Scleroderma, myositis and related syndromes

AB0716 AN AUTOPIST CASE OF SYSTEMIC SCLEROSIS WITH SEVERE GASTROINTESTINAL 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 intestinal 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 dilated 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 pneumatoasis cystoides intestinales (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 was surveyed and examined through the evaluation of the periodontal parameters and the number of teeth.

Conclusions: 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.

Disclosure of Interest: None declared

REFERENCE:

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. The triggering event of vasculopathy is unknown, but the narrowing of intestinal arterioles causing hypoxia might be responsible for dysmotility of GIT.

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 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.

Disclosure of Interest: None declared

REFERENCE:

Table 1

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Publication year</th>
<th>Age of death</th>
<th>Sex</th>
<th>Year of death</th>
<th>Cause of death</th>
<th>Comorbidities</th>
<th>GIT symptoms</th>
<th>Pathological findings of vascular damage</th>
<th>Intimal proliferation</th>
<th>Narrowing of small arteries</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1973</td>
<td>49</td>
<td>M</td>
<td>1984</td>
<td>Stomach bleeding</td>
<td>BP, MP</td>
<td>Odynophagia</td>
<td>Occlusion</td>
<td>Small vessels</td>
<td>1</td>
</tr>
<tr>
<td>2</td>
<td>1976</td>
<td>25</td>
<td>F</td>
<td>1981</td>
<td>Stomach bleeding</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>1</td>
</tr>
<tr>
<td>3</td>
<td>1980</td>
<td>65</td>
<td>M</td>
<td>1984</td>
<td>Stomach bleeding</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>1984</td>
<td>70</td>
<td>M</td>
<td>1990</td>
<td>Stomach bleeding</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>1</td>
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<tr>
<td>5</td>
<td>1985</td>
<td>60</td>
<td>F</td>
<td>1990</td>
<td>Stomach bleeding</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>1</td>
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<tr>
<td>6</td>
<td>1990</td>
<td>65</td>
<td>F</td>
<td>1991</td>
<td>Stomach bleeding</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>1</td>
</tr>
<tr>
<td>7</td>
<td>1992</td>
<td>60</td>
<td>M</td>
<td>1994</td>
<td>Stomach bleeding</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
<td>Odynophagia</td>
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</tbody>
</table>

M, male; F, female; yr, years; mo, months; GIT, gastrointestinal tract; IP, interstitial pneumonia; CIPO, chronic intestinal pseudo-obstruction; PCI, pneumatoasis cystoides intestinales; 1) Intimal proliferation and narrowing of the small arteries

Conclusions: Vascularopathy in SSc involves small vessels, and it precedes fibrosis. The triggering event of vascularopathy is unknown, but the narrowing of intestinal arterioles causing hypoxia might be responsible for dysmotility of GIT.

REFERENCES:
Abstract AB0718 – Table 1

<table>
<thead>
<tr>
<th>PATIENT</th>
<th>FOLLOW-UP</th>
<th>MODIFIED RODNAN SKIN SCORE PRE-TRANSPLANT</th>
<th>MODIFIED RODNAN SKIN SCORE POST-TRANSPLANT</th>
<th>HAQ-DI POST-TRANSPLANT</th>
<th>SHAQ-VAS POST-TRANSPLANT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>15 years</td>
<td>1/51</td>
<td>0.25</td>
<td>4/13</td>
<td></td>
</tr>
<tr>
<td>Patient 2</td>
<td>8 years</td>
<td>Unknown</td>
<td>7/51</td>
<td>0</td>
<td>3/13</td>
</tr>
<tr>
<td>Patient 3</td>
<td>5 years</td>
<td>28/51</td>
<td>6/51</td>
<td>0</td>
<td>0/13</td>
</tr>
<tr>
<td>Patient 4</td>
<td>1 year</td>
<td>33/51</td>
<td>20/51</td>
<td>0.5/0</td>
<td>7/13</td>
</tr>
</tbody>
</table>

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


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 How- ever, several biases 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 before and after disease onset were identified in larger studies.

Results: The clinical data of the four patients are below reported. 1. LS 28 year old female who underwent cosmetic breast prostheses: two years later she complained of Raynaud’s phenomenon and in the examination he explained of Raynaud’s phenomenon and in the examination he explained.2. PJ 38 year old female who underwent cosmetic breast prostheses; one year later she experienced RP and one more year later aggressive diffuse cutaneous SSc, along with anticientromere antibodies (ACA) positivity. 3. PJ 38 year old female who underwent cosmetic breast prostheses: 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 of Raynaud’s phenomenon and in the examination he explained of Raynaud’s phenomenon and in the examination he explained. 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) intestinal ulcer disease (50%) and d) scleroderma 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 x 4 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.

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


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. Data 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 (IScSc), 1 B-cell lymphoma (MALT lymphoma), and 1 prostate adenocarcinoma (IScSc). 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.

Disclosure of Interest: None declared


AB0721

TRANSFER OF SYSTEMIC SCLEROSIS AFTER ALLOGENEIC BONE MARROW TRANSPLANTATION


Background: It is accepted that donor-derived immunity is transferred with allogeneic BMT.

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).