Oral manifestations of Systemic Sclerosis and Correlation with anti-Topoisomerase I Antibodies (SCL-70)

Ismet H. Bajraktari¹, Avni Kryeziu¹, Fadil Sherifi², Halit Bajraktari³, Ali Lahu¹, Genc Bajraktari⁴

¹Clinic of Rheumatology, University Clinical Centre of Kosovo, Prishtina, Kosovo
²Clinic of Gastroenterology, University Clinical Centre of Kosovo, Prishtina, Kosovo
³Ambulance for Internal Medicine and Rheumatology, "Promedica", Prishtina, Kosovo
⁴National Institute for Public Health of Kosovo, Prishtina, Kosovo

Corresponding author: Avni Kryeziu- MD specialist for Internal Medicine and Rheumatology, Clinic of Rheumatology, UCCK, Prishtina, email:drakryeziu12@hotmail.com

ABSTRACT

Introduction: Progressive systemic sclerosis (PSS) is a chronic autoimmune illness. Clinical oral manifestations in Scleroderma are very frequent. Aim: To explore the oral manifestations, frequent and rare, to investigate whether there are differences between gender and the observed correlation of changes in relation to Antibodies Anti-Topoisomerase I. Material and methods: in the study were included 75 patients (65 females and 10 males), their mean age was 45.2±10, duration of illness was around 5.1±12 years diagnosed according to the ACR criteria and treated in the period 2010-2013. Results: 98.7% of our patients were ANA positive, whereas 49.3% of them were Anti-SCL-70 positive. Patients in 91% of cases had one or more oral manifestations of disease. The most frequent oral manifestations are: small mouth (n = 39), the lingua short frenulum (n = 21), Xerostomia (n = 24) and paradontopathia (n = 16), while more rare are: Telangiectasia (n = 14), decreased interincisal distance (n = 9), missing teeth (n = 9), absorption of dental alveoli (n = 5) and Neuralgia n. trigeminus (n = 3). Oral symptoms have been frequent in patients with Scleroderma, SCL-70 positive but not statistically significant difference. Conclusions: Oral changes have high frequency in patients with Scleroderma and these changes provide high discomfort of the mouth and lower quality of life. Oral health care to patients with Scleroderma is very important and it affects a lot in reducing the level of disease and increase the quality of life.

Key words: Progressive systemic sclerosis, Oral manifestations, Small mouth and Anti SCL-70

1. INTRODUCTION

Progressive systemic sclerosis (PSS) is a chronic autoimmune illness which is characterized with three major manifestations:

• Process of fibrosis in the skin and internal organs,
• Non-inflammatory vasculopathy of small blood vessels, and
• Specific autoantibodies which are not pathogenic, but are in correlation with activity of illness (1, 2).

In a PSS, a typical patient has developed Reynaud phenomenon, skin edema and sclerosis in the hands, face, trunk, lungs are affected with subsequent pulmonary hypertension and cardiomyopathy, heart and kidneys are affected which result in renal hypertensive crisis. These clinical manifestations might present as sole symptoms or combined together (3).

Clinical manifestations in the gastrointestinal tract (GIT) are very frequent manifestations after cutaneous manifestations which are the most frequent ones, and often attention is not paid to the latter, because cutaneous manifestations are not perceived as manifestations per se, even though have direct impact in the quality of life of patients (4). Damages in GIT are present among large number of patients with PSS, but it is still unknown why certain parts of digestive tube are more affected in comparison to the other parts (5, 6). However, certain clinical manifestations in the GIT are so frequent that are considered as dominant manifestations of PSS (Oral and esophagus changes) (7). Changes caused by PSS span from mouth to anus. Changes in the mouth are in the shape of microstomia which is caused due to skin fibrosis on the face that gives the look of “the bird’s face” which is one of the most distinctive features of patients with PSS (8). Other changes on the mouth are shortening of tongue frenulum, changes on the gum and uvula, fibrosis of salivary glands associated with dry mouth (syndrome sicca
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The research has calculated the following statistical parameters: frequency, median age, and Chi-Square test, correlation of oral clinical manifestations and anti-Topoisomerase I.

Electronic inquiry of search engine Google and in PubMed didn’t yield data on similar research conducted in Kosovo and Balkan Peninsula.

4. RESULTS

In clinical research laboratory and radiological, clinical oral manifestations of patients with PSS have found: of 75 patients with PSS, 91% had one or more clinical manifestations oral.

2. AIM AND OBJECTIVE

The research aims to find common oral manifestations and rare, to note that there are differences between the patients’ gender and noticed the correlation of these changes in relation to antibodies against topoisomerase I.

3. MATERIAL AND METHODS

Our study is prospective, and based on descriptive, research and analytical method we have explored PSS oral manifestations among patients who were diagnosed with PSS based on the revised ACR criteria from 2010 to 2013 (16). In all patients are required Anti-nuclear antibodies (ANA), Anti-centromere antibodies (ACA) and antibodies against Topoisomerase I (Anti SCL-70.) 75 patients: 65 patients were females, respectively 10 males, with median age 45.2 ± 10.1 Patients were with active illness ranging from 1.5–8 years, with a median of 5.1±12 years, whereas the illness has been evaluated in accordance with EUSTAR criteria (European Scleroderma Trial and Research) (17).

Evaluation score regarding affection with systemic sclerosis is done in accordance with EUSTAR as well: affection of cardiovascular system is scored with 2 points, skin – 2 points, erythrocyte sedimentation >30 in the first hour -1.5 points, Rodman’s modified cutaneous test > 20 – 1 point, decrease of value of complement – 1 point, distal necrosis of hand fingers – 0.5 points, and DLCO (diffusing capacity of the lung for carbon monoxide) lower than 80%–1 point. Patients are all residents of Kosovo and the study is not included 7 patients diagnosed with PSS who were not residents of Kosovo. All patients were informed and have provided consent to be part of our study. During the examination of patients with SSP is marked each clinical manifestation of mouth, for each was conducted X-ray and CT and special cases was also conducted consultations with Maxillofacial Surgeon Specialist or Oral Surgeon.

Most frequent clinical manifestation on the mouth was small mouth (microstomia) found in 52.0% of patients, followed by dry mouth 32.0%, shorter frenulum linguae 28.0% and parodontopathy in 21.3% of patients. The following symptoms were most frequently seen in males in comparison to females, but with no significant statistical difference (P=0.05), (Table 2).

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Rare oral manifestations are presented more men than women.

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Negative anti SCL - 70</th>
<th>Positive anti SCL - 70</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>%</td>
</tr>
<tr>
<td>Total</td>
<td>38</td>
<td>100</td>
</tr>
<tr>
<td>Small mouth</td>
<td>15</td>
<td>39.5</td>
</tr>
<tr>
<td>Shorter frenulum linguæ</td>
<td>9</td>
<td>23.7</td>
</tr>
<tr>
<td>Dry mouth</td>
<td>11</td>
<td>28.9</td>
</tr>
<tr>
<td>Parodontopathy</td>
<td>6</td>
<td>15.8</td>
</tr>
</tbody>
</table>

X²-test, P-value X²=0.446, P=0.930

Table 4. Clinical symptoms in mouth among patients with scleroderma and Anti SCL-70

Oral symptoms are more frequent among patients positive to SCL-70, but in X²-test we didn’t found any significant correlation (P>0.05), (Table 4).

5. DISCUSSION

Our patients with PSS underwent examination related to oral clinical manifestation. 91% of patients were having one or more oral manifestations of PSS. There are numerous researches which were dealing with oral manifestations of PSS. Canadian authors in their paper The Canadian Oral Health systemic sclerosis study III have analyzed 163 patients; 90% were female, mean age 56 ± 11 years, disease duration 14 ± 8 years and their results were: Sjögren's syndrome, decreased interincisal distance, missing teeth and parodontopathia (17).

Results from our study are almost identical with the findings from the working of Canadian authors. Marmary author has researched Scleroderma oral manifestations in 41 patients and found that reduced mouth opening, mandibular condyle damaging, and coronoid process are the most frequent damages of Scleroderma (18). In other research Robert E Wodd analyzed 31 female patients with progressive systemic sclerosis and findings for: Xerostomia, erosion of the mandible and parodontopathy are similar to the results from our study (19). In research: Analysis of the oral manifestations of systemic sclerosis is researched group of 32 patients with Scleroderma and other research except analyzed dry mouth where our results are very close to their results (20). Author Scardina in their publication: Early diagnosis of progressive systemic sclerosis: the role of orofacial phenomena analyzed 34 patients; Women were 30, males were 4, and findings that Xerostomia, reduced mouth opening and Neuralgia n. trigeminus are the most common damages (21). Vincent C with co-author in his research with 30 patients found that: Telangiectasia perioral, reduced opening the mouth, xerostomia, mandibular absorption, Neuralgia n. trigeminus are the most common injuries. Our results are identical with its findings except Telangiectasia which we presented our research on low frequency (we youngest patients and shorter the duration of the disease) (22). Author Michaels Van Amelsfort found data matches the data from the research our reduced opening, mouth and dental disturbance (23). Study published in 2003 in the journal Arthritis and Rheumatism stated that level of anti-topoisomerase I (anti SCL – 70) has a positive correlation with the activity of illness in the PSS (15).

6. CONCLUSION

• In the research were included 75 patients – 65 females and 10 males.
• Median age of patients was (45.2±10.1), while duration of illness was 5 years.
• Our patients with PSS in 98.7% of cases were ANA positive, whereas anti SCL – 70 were positive 49.3% of cases.
• Patients in 91% of cases had one or more oral manifestations of disease.
• Most frequent oral manifestations of patients with PSS are: small mouth, frenulum short lingua, dry mouth and Parodontopathy.
• Rare oral manifestations are: Telangiectasia, decreased interincisal distance, missing teeth, alveolar absorption and Neuralgia n. trigeminus.
• Presence of oral manifestations symptoms didn’t give significant difference between genders.
• Oral manifestations are present more often in patients who were positive in the Anti SCL-70 but no statistical difference significant.
• Additional scientific research needed to certify the reasons why oral pathology does not affect equally all patients with Progressive Systemic Sclerosis.
• Oral manifestations are due to discomfort in the mouth and reduce the quality of life.
• Oral health care for patients with PSS is very important because it affects a lot in reducing the overall morbidity of this disease and improves quality of life.

CONFLICT OF INTERESTS: NONE DECLARED.

REFERENCES

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