CASE REPORT

Scleroderma – from the Aspect of Dental Medicine

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M odern etiopathogenic and diagnostic procedures of oral diseases are founded on molecular biology, genetic engineering, autoimmune, pathohistological, cytological and immunofluorescence methods. The autoimmune processes may include the humoral and cellular immune responses and all organ systems may be involved. The systematic and orofacial manifestations of the scleroderma with medical and pathohistological documentation indicates to interdisciplinarity cooperation in the treatment of such patients. The multidisciplinary approach in patients with scleroderma requires and suggests the most efficient prophylaxis treatment from the aspect of dental medicine. The systematic and orofacial manifestations of the scleroderma with medical and pathohistological documentation indicates to interdisciplinary cooperation in the treatment of such patients. The multidisciplinary approach in patients with scleroderma requires and suggests the most efficient prophylaxis treatment from the aspect of dental medicine. KEY WORDS: DENTAL MEDICINE, SCLERODERMA, IMMUNE RESPONSES, PROPHYLAXIS TREATMENT.

1. INTRODUCTION

Modern etiopathogenic and diagnostic procedures of oral diseases are founded on molecular biology, genetic engineering, autoimmune, pathohistological, cytological and immunofluorescence methods. The autoimmunity processes may include the humoral and cellular immune responses and all organ systems may be involved (1).

Scleroderma is a multisystem disease that affects the connective tissue and the blood vessels and causes thickening of the skin and the mucosa, smooth muscle atrophy and internal organ fibrosis (2). The prevalence of scleroderma is estimated at about 250 cases on one million people where women are much more likely to be affected by the disease than men (3:1) (3).

There are two types of scleroderma:

- Systematic scleroderma and
- Localised scleroderma.

Progressive systemic scleroderma is a multisystem disease characterised by inflammation and fibrosis of several organs. There are two subtypes of systematic sclerosis:

- Limited skin scleroderma or CREST syndrome (C-calcinosis cutis, R-Raynaud’s phenomenon, E-esophageal dysfunction, S-sclerodactyly, T- telangiectasias),
- Diffuse skin scleroderma.

The limited scleroderma develops slowly and starts with the Raynaud’s phenomenon, while the diffuse subtype is an acute scleroderma with the emergence of arthritis, hardening of the skin and a visible swelling of hands and feet.

2. CASE OVERVIEW

Patient, aged 46, was administered for the first time at the Department for Oral Medicine and Periodontology of the Faculty of Dentistry in Sarajevo on March 19, 2010.

We learned from his medical history and the accompanying documentation that the patient had been treated under the diagnosis of Sclerosis systemic progressive, Sy. Raynaud, in Germany in 2000. During the treatment, the patient underwent lab testing which indicated to positive antibodies Scl 70 highly specific for sclerodermia whose titre was > 400 U/ml (reference value < 25 U/ml) and a positive HLA B27. He was then hospitalized at the Clinic for Internal Diseases at the University Clinical Centre in Tuzla, Department of Rheumatology from December 19, 2000 - January 3, 2001, due to pain and joint swelling on both hands. The lab tests obtained during hospital treatment were all in physiological boundaries except for the number of erythrocytes (8,8) and Glucose (6,6). The X-ray of the hand indicated to a discrete stenosis of interarticular areas of the metacarpal phalanges on joints of the second and third finger on both hands. The immunofluorescence tests on fibrinogen indicated to a slight focal granular positivity (+) in the dermal collagen. The immunofluorescence tests on IgG, IgA, IgM, C3c, C1q were completely negative. The skin sample taken for biopsy was tested for lupus on a band test and did not show any morphological changes. The patient was prescribed a therapy of systemic corticosteroids (Pronison) and immunosuppressive and non-steroid anti rheumatic drug.
During the clinical examination at the Department for Oral Dentistry and Periodontology of the Faculty of Dentistry in Sarajevo, the doctors register patient’s difficulty in opening his mouth. In an attempt to smile, the patient presents the “mask face.” (Figure 1).

On the left buccal mucosa, there is a presence of an erosive lesion the size of a coin in the area of the upper carious molar.

The clinical test of the hard palate indicates to the fact that the patient is a smoker due to pronounced accessory saliva glands filled up with keratin.

The macroglossia of the tongue is evident together with the hypertrophy of the philiform papillae on the tongue’s dorsal surface. The asymmetrical distribution of the horizontal fissure indicates to lingua plicata finding. The tongue is both actively and passively completely mobile with the frenulum of the tongue in physiological boundaries. A slight tremor of the tongue is also present.

The alveolar endings are atrophic due to an early loss of a tooth. A traumatic ulcer is visible in the area of the lower fornix vestibulum.

The lower jaw contains paradigmopathic teeth no. 33, 34 and 35 and a partial prosthesis which does not satisfy the aesthetic or functional criteria. The upper jaw contains fixed prosthetic works in the region of the 13-15 and 23-24 tooth with the second level of attachment on the abutment tooth. Temporomandibular joint is in physiological boundaries without perceptible crepitations.

Generalised changes on all joints of both hands are evident, the movements in all joints of the hand and the fingers are hindered and increased level of sedimentation of the erythrocytes and hypergammaglobulinemia (4). The specific circulatory antibodies are useful to confirm the diagnosis and those include: anti-DNA antibodies, centromere antibodies, Scl 70 antibodies, anti-Sjögren Sy. A antibodies, (anti-SS-A, anti Ro), anti Sjögren Sy. B antibodies (anti-SS-B, anti-La). The biopsy of the skin and histopathological analysis confirm the deposition of thick collagen (5).

3. DISCUSSION

Scleroderma is a collagenosis which belongs into the category of autoimmune diseases. In scleroderma, the antibodies are targeted at the connective tissue and the blood vessels (1). The prevalence of scleroderma in men is between age 30 and 40 (2). In our case study, the man is 46 years old, which represents a rare prevalence rate according to data from the books (3).

However, our observation does not coincide with the findings from the US studies which indicates that the disease is more common in Afro-American rather than in the Caucasian population and that the frequency of the disease is age specific (6).

The confirmed diagnosis of progressive systemic scleroderma is Raynaud’s Syndrome with findings of positive antibodies for scleroderma whose titre was 400 U/ml (reference value is < 25 U/ml) and a positive HLA B27. Our lab tests are concordant with Burkett’s research (2) where the antinuclear an-
Antinuclear antibodies (ANA) are present in 90% of patients with scleroderma and the antinuclear and anticientromere antibodies are characteristic. In our case overview, there was the presence of the Raynaud’s phenomenon, the proximal vasospasm in fingers resulting in the change of the colour on finger pads as a response to the cold and the emotions which is one of the most common findings of the progressive scleroderma - a fact which concords with Burket’s research (2, 7). Our findings coincide with book data (8) namely the clinical type of scleroderma CREST syndrome which is characterised by the skin calcinosis. Our patient has hyperpigmentation, telangiectasis and subcutaneous calcifications. The changes registered on the bones and muscles are manifested as polyarthralgia and morning stiffness of small and large joints (9).

Oral manifestations - microstomy is developed as a result of the collagen deposit in the perioral tissues which hinders the closing of the mouth. There is also the presence of dysphagia and progressive periodontitis with a rapid loss of alveolar bone and teeth. The tongue is rigid, the speech, mastication and swallowing is difficult, which is consistent with the research (1, 2, 5). The patient underwent clinical, X-ray and OPG analysis, tooth extraction with a bad prognosis and was subjected to complete prosthetic restoration works.

Microstomy is one of the most significant consequences of scleroderma. In our case it was indicated but it did not represent a side-effect for the prosthetic rehabilitation of the patient. This is a preventive treatment procedure for microstomy in its terminal phase as well as for the dysfunction of the temporomandibular joint.

In our OPG analysis, we noticed a generalised enlargement of the periodontal ligament which corresponds to the Burket’s findings which has been confirmed in 10% of patients with scleroderma (2).

The patient presented in our case study and documented with medical documentation and the Ph findings is a confirmation of the diagnosis of the progressive scleroderma with Raynaud’s phenomenon. There is a generalised deformation of the hand joints and an amputation of the distal phalange on the right hand finger.

The symptoms included in the CREST syndrome are connected to the generalised form of the systemic (sclerosis) scleroderma. In our case overview, the limited symptoms of scleroderma that indicate to the CREST syndrome are:

- Calcinosis - calcium deposits in the skin,
- Raynaud’s phenomenon - blood vessel spasm as a response to cold or stress,
- Esophageal dysfunction - acid reflux and decrease in the mobility of the oesophagus,
- Sclerodactyly - hardening and tightening of the skin on the fingers and hands,
- Telangiectasias - dilatation of capillaries on the skin’s surface,
- which is concordant with the research (10).

4. CONCLUSION
The systematic and orofacial manifestations of the scleroderma with medical and pathohistological documentation indicates to interdisciplinary cooperation in the treatment of such patients.

The multidisciplinary approach in patients with scleroderma requires and suggests the most efficient prophylaxis treatment from the aspect of dental medicine.

REFERENCES
1. Dedić A. Autoimmune oralne bolesti: praktikum. Sarajevo: Institut za naučnoistraživački rad i razvoj KCUS; 2010. (Autoimmune oral diseases: practicum.)
2. Greenberg MS. Burketova oralna medicina: dijagnoza i liječenje. Zagreb: Medicinska naklada; 2006. (Burket’s Oral Medicine: Diagnosis and Treatment)