Scleromyxedema Problem in Odontological Practice

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Abstract

Scleromyxedema is a skin disorder, which is considered very rare. Its symptoms are mucin accumulation in the patients' skin, which is also called mucinosis. It causes papular and sclerodermoid bumps. Also, people affected by this disease have increased production of fibroblasts in the absence of a thyroid disorder. Another symptom includes monoclonal gammopathy, which is shown as abnormal protein existence in the person's blood. This disease has no standard treatment. All patients should have their life quality improved by any means possible. These ses are extremely challenging for most of the treatments, that are being made by a dentist, because the patient is unable to open his mouth wide enough, so that standard dental manipulations can be made. A 60-year-old patient was presented with Scleromyxedema to our clinic. He was unable to open his mouth by more than 1,7 centimeters, which is considered a very narrow mouth opening. Patients face muscles were rigid, especially – musculus orbicularis oris and musculus masseter. Any attempts to open the mouth by more than 1,7 centimeters resulted with ruptures in patients mouth corners. Intra-oral Examination revealed 2 teeth left in the lower jaw, and no teeth in the upper jaw. Patient was in need of masticatory function rehabilitation, phonetics and aesthetics improvement. Such case requires special procedures and specific adaptation, compared to a standard edentulous patients' treatment. In this case report, we review all the intra oral procedures, which should be modified accordingly to this rare case. Clinicians should be informed of such rare diseases and should be able to diagnose and treat them accordingly.

Keywords: Scleromyxedema, dental, prosthodontics, dentures, removable dentures;

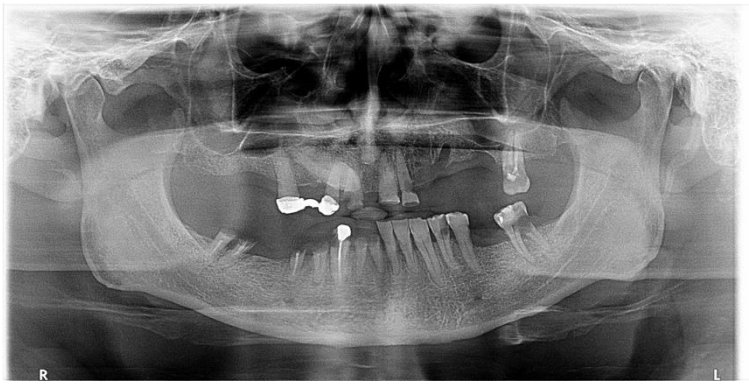
1. Introduction:

Scleromyxedema is an established disease which usually affects people, whose age is between 30 and 50 years old [1]. Pathogenesis of the disease is unknown. The main hypothesis is that circulating cytokines such as IL-1, TNF-alpha and TGF-beta, which are known to stimulate glycosaminoglycan synthesis and fibroblast proliferation in the skin, could play a role [2, 3]. Usually, the most common symptom is the

hardening of persons skin tissue. Also, papules can be observed on top of patients skin. Usually, the papulas, that we see, are firm, its color is according to the persons skin color, with some red shade on top of them. Papules size varies, but usually its around 3 millimeters in diameter. Usually, the most affected parts are knees, face and elbows, and lowered range of motion of face, fingers and extremities can be observed. Histopathological examination shows mucin deposition [4]. Patient can have symptoms of vertigo, seizures and psychosis [5]. Also, usually cardiovascular diseases are observed [6]. These types of cases make it very hard for the dentist to successfully reestablish full mouth chewing function, patients phonetic abilities and aesthetical view of his smile. In order to achieve aforementioned points, we need to access mouth rehabilitation in a different, modified way.

2. Case presentation:

The clinical case we report here is of an 60-year-old male patient, with medical history of heart diseases and scleromyxedema. Patient was using drugs: Midrol, metotrexatum, trental, potassium chloride, mildronatum and folium acid. When patient arrived at Lithuanian University of Health Sciences Maxillofacial orthopedics clinic, he reported inability to be able to move his lips freely. After clinical examination, it was concluded, that the patient needs full mouth function, aesthetics and phonetics reconstruction. Orthopantomogram (x-ray analysis) was made (Pic. 1).



Picture 1. Orthopantomogram.

We concluded, that most of the teeth were damaged, and since no other restoration may be performed, due to severely damaged teeth, we sent the patient to an oral surgeon for sanation of the mouth, so that only the healthiest teeth would remain. Most of the teeth only had their roots left. The oral surgeon removed all the infectious teeth, and only 2 teeth were left in the patients mouth: tooth number: 34 and 35 (premolars of the patients left lower jaw).

Concluding pre-prosthetic treatment, patient had no teeth in his upper jaw, and 2 teeth in his lower jaw. Patients skin had papulas all over his cheeks, forehead, chin and also on his hands (Pic. 2).

Picture 2. Papulas on patients hand.

Patients face had red blemishes, he was amimic. Patient was very limited in opening his mouth. His maximum height between upper and lower lips was: 1,9 cm (Pic. 3).

Picture 3. Maximum height between lips when the patients mouth is fully open was 1,9 centimeter.

His maximum mouth width with fully opened mouth was 4,0 cm (Pic. 4). Intraorally, patients gums were inflamed.

Picture 4. Maximum width between patients mouth corners was 4,0 cm.

Patients Orthopantomogram showed teeth number 34 and 35. Alveolar ridge resorption was clear. Two remaining teeth (number 34 and 35) were damaged by caries and they needed treatment by a general practitioner dentist. After situation evaluation, and according to patients expectations and current financial situation, we chose the most well-rounded mouth rehabilitation method – removable partial dentures (RPD) for the lower jaw, and full removable dentures for the upper jaw. In order to be able to make these prosthesis, we needed to address the main issue – patients inability to open his mouth wide enough, so that standard manipulations required to make RPD can be made.

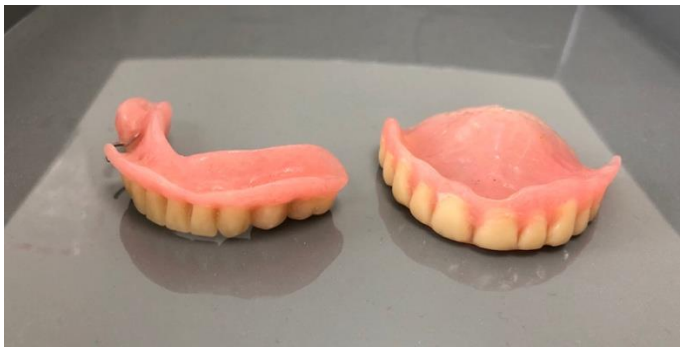
First, we needed to make impressions of the mouth in order to make individual trays. Impressions were made using polyvinylsiloxane material. Plastic trays were modified in order to fit patients mouth accordingly. Both trays were modified, and cut in pieces, because we were unable to fit them in his mouth

properly, due to rigid lips and limited mouth opening. After taking initial impressions, we made plaster casts, and formed special individual trays, to fit the patients mouth. Impressions were made with small individual trays, that we were able to fit in patients mouth (Pic. 5).

Picture 5. Small individual trays with silicone impression material, taken out of patients mouth.

Next, we took impressions with individualized trays and made plaster models, which were used to make our removable partial dentures (RPD).

When we got our final RPD, we individualized it, so that it could fit inside the narrow mouth opening. One of the sides of the partial dentures were slimmed down and reduced in width. The partial dentures were modified in order to fit sideways (Pic. 6). Only this way we could achieve good stability, phonetic rehabilitation, and aesthetic view of the patient.



Picture 6. Removable partial dentures.

3. Discussion:

Scleromyxedema is being increasingly recognized as a disease, that is widespread thru patients, that have a mean age of 59 years [7]. People, who are getting older, due to changes in their mouths structures, often need their chewing function rehabilitated with various types of treatments, including removable partial dentures [8]. It was very hard to establish a treatment plan for our patient, because these types of cases are usually being treated systematically, without any special types of treatments to rehabilitate chewing functions, and there were no similar cases that we could find in the literature. The main difficulty in treating patients with scleromyxedema is their inability to open their mouths wide enough for us, dentists, to be able to perform odontological manipulations and procedures in their mouths.

One of the most popular, and mostly accessible methods of restoring mouth chewing function is removable

partial dentures [8]. In order to make these types of dentures, we need to be able to freely make certain manipulations in the patients mouth. According to scientists Min Woo Park et al. research, patients with TMJ (temporomandibular joint ankylosis) have their mouth opening ranging from 0 to 20mm [9], whereas our patient had a mouth opening of 17mm. This is the reason why each RPD step needed to be modified. Chewing I also compromised in scleromyxedema patients. Cases report weakness of muscles, generalized weakness due to inflammatory myopathy and also fibromyalgia [10, 11]. This is why our removable partial dentures needed to be precise and as functional as possible, so that the patient would be able to masticate. Masticatory function is also very important, because it is the start of the whole gastrointestinal system. Patients, who have scleromyxedema, may develop dysphagia, and oesophageal dysmotility [12]. In that case, chewing food to small pieces is extremely important, and that is also one of the reasons, why masticatory function should be recovered as good as possible.

4. Conclusion.

Scleromyxedema is a significant disease, that affects many systems in the patients body, including stomatogenous system, which affects the patients quality of life. Early diagnosis and adaptive treatment to each individual case will improve outcomes. The earlier we make required prosthetic manipulations in the patients mouth, the earlier quality of life will be assured for the patient. This disease is associated with other organs pathologies, so it is critical, to work on these patients as a medical team.

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