IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 11 Ver. V (November. 2016), PP 79-81
www.iosrjournals.org

Prosthodontic Rehabilitation of Systemic Sclerosis:A Case Report

Author's Names: Dr. Litty Francis[1], Dr. S. Babukuttan Pillai[2]
[1]*Government Dental College, Alappuzha, India*
[2]*Government Dental College, Thiruvananthapuram, India*

Abstract: *Systemic Sclerosis (Scleroderma) is a symptom of group of diseases of the immune system, blood vessels and connective tissue, in which the skin, usually of the hands and feet, becomes stiff, tight and shiny. It can lead to reduced mouth opening and a consequent difficulty in performing dental procedures. The prosthodontic rehabilitation of patients with Systemic sclerosis can be challenging in terms of rendering treatment as well as patient management.*

Keywords*: Microstomia, Raynauds phenomenon,Sclerodactyly,Systemic Sclerosis, Xerostomia*

I. Introduction

Systemic sclerosis (Scleroderma, SSc) is a complex autoimmune disease with various clinical manifestations[1,2]. It is a chronic systemic disorder of unknown etiology[3] and there is excessive deposition of collagen and other connective tissue macromolecules in skin and multiple internal organs[4]. The pathognomonic features include vasculopathy of small arteries and arterioles, fibrosis of the skin and internal organs and involvement of immune system[4-6]. Based on the extent of skin involvement as well as clinical and laboratory findings, it can be differentiated into limited cutaneous systemic sclerosis (lcSSc) or diffuse cutaneous systemic sclerosis (dcSSc)[2,7]. Skin involvement in lcSSc develops gradually and remains limited to the fingers (sclerodactyly), distal extremities, and face. They have a long-standing history of Raynaud's phenomenon, limited internal organ involvement, and a better prognosis [8].dcSSc patients have a rapid onset, characteristically develop widespread skin thickening, distinctive multiple visceral organ involvement and potentially life-threatening cardiac and renal failure [2].

SSc patients have a characteristic "mauskopf" appearance with taut and shiny skin, loss of wrinkles, and occasionally an expressionless facies due to reduced mobility of the eyelids, cheeks, and mouth[2]. They may have thin lips with fine wrinkles (radial furrowing) around the mouth. The mouth opening is usually reduced, and in 80% of patients with systemic sclerosis, microstomia is seen. The involvement of tongue leads to difficulty in speaking and swallowing. Significant decrease in inter-commissural, inter-incisal and inter-vermilion border distance was observed in SScpatients [9,10] and Xerostomia was present in 70% of the patients[10,]Mucous membranes become thin and tight affecting the residual alveolar ridges and denture border extensions [11]. The limited mouth opening and sclerodactyly can lead to difficulty in impression making, performing oral hygiene measures and difficulty in insertion and removal of dentures.

Most of the case reports deal with the use of modified and/or sectional trays[12, 13]& fabrication of sectional and collapsible dentures for patients with microstomia [14]. Systemic sclerosis is a very rare, systemic disease having oral and prosthodontic implications and this case report describes an arbitrary method used for the rehabilitation of partially edentulous patients.

II. Case Report

A female patient aged 47 reported to the Department of Prosthodontics for prosthetic replacement of her missing mandibular anterior teeth. A thorough case history and clinical examination was done. The diagnosis of lcSSc was made primarily on clinical grounds using ACR-EULAR Classification Criteria for Systemic Sclerosis[15]. The skin of the fingers was thickened extending proximal to the metacarpophalangeal joints. A long history of Raynaud's phenomenon from the age of 12 and Sclerodactyly of both hands were noted [Figure 1- acro-osteolysis]. Patient also gave a history of migraine and symptoms of gastroesophageal reflux. The skin over the face was taut and shiny with reduced wrinkles [Figure 2- mauskopf appearance]. The nasal alae became atrophied, resulting in a pinched appearance of the nose. The skin over the neck showed areas of vitiligo-like hypopigmentation giving a "salt-and- pepper" appearance. Mouth opening was restricted to with a maximum opening of 25.0 mm. Patient gave a history of mild Xerostomia. Intra oral examination showed missing mandibular anteriors and premolars. The maxillary anterior teeth were protruded.

As the mouth opening was limited, the normal impression procedures were not possible. It was impossible to insert a stock mandibular tray for impression making. A putty impression using polyvinyl siloxane impression material (Zhermack Elite HD+) of the mandibular arch was made without a tray for support. The impression was then lined using polyvinyl siloxane light body impression material (Zhermack Elite HD+) to

record finer details [Figure 3 – Mandibular impression]. This was used to pour a cast in Type 3 dental stone[GypRock]. The maxillary impression was not attempted. A wax occlusal rim was made and the mandibular teeth arranged on it. The teeth arrangement was arbitrarily done keeping the incisal edges at the level of the premolar conforming to the existing arch form. The patient was recalled for trial appointment and trial denture was inserted. Minor adjustments were done during the try-in and occlusal harmony was established. For ease of insertion and removal, the denture was extended up to first molar only on both sides [Figure 4 – teeth arrangement].

Once the patient was satisfied, the trial set up was sealed, flasked and heat cured. The denture obtained was finished and polished & inserted into the patient's mouth at the next visit. Proper instructions were given regarding the insertion, removal and maintenance of the denture. The patient was advised to do passive jaw stretch exercises using stacks of tongue blades six times per session, six sessions a day. Regular recall was done and she is satisfied with the denture provided.

III. Discussion

SSc is a of connective tissue disorder occurring more commonly in women than in men[16], particularly in the age range 45–64 years[17]. The overall incidence rate in the adult population is approximately 20/million/ year and has a prevalence of 240/million[18, 19]. Prevalence and incidence appears to be greater in populations of European ancestry and lower in Asian groups[20].

The normal reparative responses of fibroblasts become amplified resulting in irreversible fibrosis. Telangiectasia are frequently seen in lcSSc and are prominent on the face, hands, lips, and oral mucosa. The healing of ischemic fingertip ulcerations leaves characteristic fixed digital "pits". The loss of soft tissue at the finger tips due to ischemia frequently associated with resorption of the terminal phalanges (acro-osteolysis)[2] and flexion contractures produce shortened claw like fingers. Varying degrees of resorption of the posterior ramus of the mandible, the coronoid process, the chin, and the condyle may be detected on panoramic radiographs affecting approximately 10–20% of SScpatients[21]. The erythrocyte sedimentation rate (ESR) is generally normal and antinuclear autoantibodies are present in almost all patients with SSc[22].

Tongue blades between the posterior teeth have been used as a non-surgical method to manage microstomia[23]. Pizzo et al.[24] showed an average increase in mouth opening of 10.7 ± 2.06 mm with jaw stretch exercises. Surgical method i.e. bilateral or unilateral commissurectomy have also been tried to improve mouth opening in SSc patients[25, 26].Patients may also have decreased ability to manipulate a tooth brush as was seen in our case. Adapted equipment may make it easier for a patient with decreased dexterity and range of motion to brush and floss one's teeth[27].

Yenisey et al.[27] described sectional maxillary and mandibular trays and a collapsed mandibular denture for the total edentulous patient with microstomia caused by scleroderma. Jivanescu et al.[28], presented the fabrication of a flexible complete denture in a female patient with scleroderma induced microstomia. A sectioned standard tray was used to make the impression. In our patient only the mandibular anterior teeth were missing and the procedures like final impression, jaw relation record were omitted for comfort of the patient. With the impression procedure used in this case, the impression and the denture fabricated was satisfactory. As the patient had only a few teeth to be replaced, there was no need for sectional trays or dentures.

IV. Conclusion

We must take into consideration the physical as well as psychological aspect of patients, as the prosthodontic management is a challenge in itself. The treatment procedures should be uncomplicated and of short duration. Restricted mouth opening often demands a deviation from the conventional treatment procedures. Customized modifications in impression procedures and prosthetic designs are often necessary in the prosthetic rehabilitation and improvement in the well-being of the patients.

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Acknowledgment: None