

Oral Rehabilitation of a Patient with Sickle cell anemia - Case Report

Dr. Neeraj Trehan¹, Dr. Palvideet Kaur², Lana Elfatih Mohamed Eltahir³, Anu Mariamma Easo Kochuparampil⁴, Gurpreet Kaur⁵, Abu Saleh Mohammad Shajal⁶

¹Doctorate of Dental Medicine (DMD), Private Practitioner, USA

²BDS, MDS, Department of Pediatric and Preventive Dentistry (India), Research Assistant (USA)

³BDS (Sudan), Research Assistant (USA)

^{4,5}BDS (India), Research Assistant (Canada)

⁶BDS (Bangladesh), Research Assistant (USA)

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Abstract

Sickle cell anemia patients with dental anomalies represent a difficult challenge for the prosthetic dentist. Proper treatment planning with the medical and dental management of these patients are important prerequisites for successful results. This case report describes a 70-year-old female patient was referred to a dental clinic sickle cell anemia. It was found that the lower anterior and posterior teeth have experienced significant wear, leading to the exposure of the dentin. Furthermore, there are several areas without teeth present in both the upper and lower jaws. This case report describes a full-mouth rehabilitation of a sickle cell anemia patient with worn-down dentition including the treatment planning, medical consultation, correction of vertical dimensions of occlusion, preparation, and restoration with porcelain fused to metal crowns. It was concluded that the manifestations and degrees of severity are influenced by several factors, such as the stage at which the disease is identified, the patient's age, the frequency of hospitalizations, the need for blood transfusions, and the necessity for ongoing medication, among other considerations.

Key Words: Dentin, Metal Crown, Oral Rehabilitation, Sickle cell anemia

Introduction

Sickle cell anemia (SCA) is a genetic condition that can arise from a defect in chromosome II. The abnormal hemoglobin S (Hb S) is produced in a homozygous state due to a genetic defect, resulting in an amino acid substitution. The deoxygenation of hemoglobin S leads to the formation of sickle-shaped cells. As a result of the reduced affinity of oxygen for the abnormal hemoglobin, further deoxygenation will occur, leading to an increase in the production of sickle-shaped cells.^{1,2}

Symptoms of sickle cell disease (SCD) usually manifest in children early in life. Elevated levels of hemoglobin F offer substantial protection to infants in their early months, particularly during the first six months of life; however, the implications of this condition typically emerge shortly thereafter. Sickle cell disease presents several oral manifestations that are important to recognize. Common signs include mucosal pallor, yellowing of the tissues, radiographic abnormalities, delayed tooth eruption, and issues with enamel and dentin mineralization. Additionally, changes may occur in the superficial cells of the tongue, along with malocclusion, hypercementosis, and an unusual degree of periodontitis in children.^{3,4}

For effective dental treatment, it is essential for dental surgeons to have a solid understanding of the pathophysiology of this disease. This will allow for the development of treatment plans that also consider any

existing systemic conditions. Effective dental management for patients with sickle cell anemia necessitates a collaborative approach, emphasizing the importance of communication and teamwork before and throughout the dental treatment process, along with consultation with the patient's hematologist.^{5,6}

The Case Report

During the examination, evaluation, and treatment of worn-down teeth, a 70-year-old female patient was referred to a dental clinic that was recognized for handling medically compromised individuals. Considering the patient's medical history, it has been established that the individual has sickle cell anemia. The patient's physical condition appears to be within normal limits, although there is evidence of generalized moderate weakness.



Figure 1: Extra Oral Examination of the Patient

After a dental examination, it was found that the lower anterior and posterior teeth have experienced significant wear, leading to the exposure of the dentin. Furthermore, there are several areas without teeth present in both the upper and lower jaws. The presence of small and missing teeth, along with gingival hyperplasia, resulted in short clinical crowns and significant gaps between the teeth (Fig. 1). This observation was particularly evident in the maxillary area of the mouth.



Figure 2: Intraoral Examination

The absence of the patient's molar and the wear on the occlusal surface have led to a reduction in the vertical dimensions of occlusion (VDO) and an increase in the interocclusal rest space. (Figure 2 and 3) The patient

demonstrated effective oral hygiene practices. To ensure accurate diagnosis and effective treatment planning, as well as to maintain comprehensive patient records, intraoral photographs and x-rays were taken before the

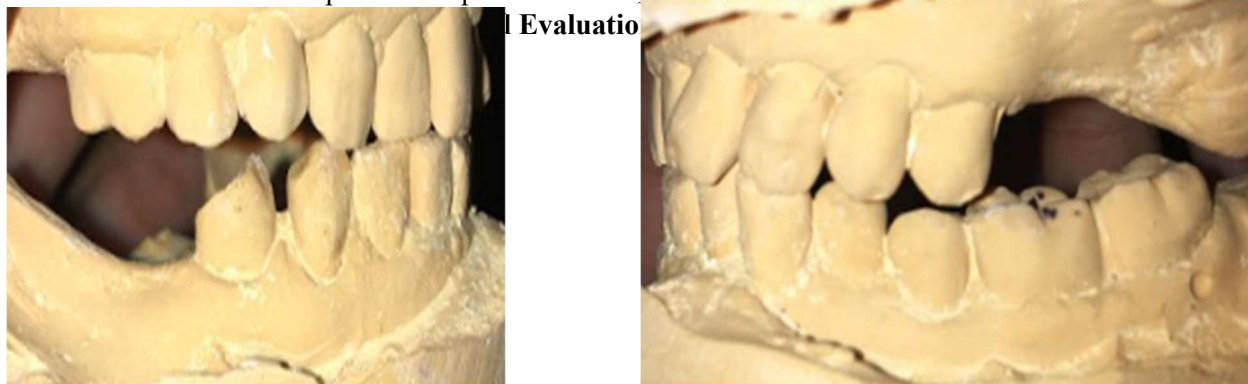


Figure 4: Occlusal Evaluation – Dynamic (Anterior Guidance)

The treatment plan was developed with careful consideration of masticatory function, the restoration of vertical dimensions, and aesthetic outcomes. A comprehensive discussion took place with the patient about the treatment plan. During this conversation, the different treatment options were clearly outlined, and the patient's consent was secured. The plan that was confirmed included a medical consultation, crown lengthening of the upper and lower molar and premolar teeth, as well as the construction of metal ceramic restorations for the missing teeth.

The hematologist, after consulting with the patient's physician, recommended that the patient receive antibiotic coverage and a blood transfusion before each dental surgery appointment. Digital impressions of the maxillary and mandibular arches were obtained following the patient's education on dental prophylaxis and oral hygiene practices. Diagnostic casts were created and subsequently mounted on a semi-adjustable articulator. This process included the use of a face bow transfer and a centric relation record, which was established using bite registration material. To assess the necessary tooth reduction for the planned metal-ceramic restorations, diagnostic preparation and wax-up were conducted on the diagnostic models.

The crown lengthening procedure was performed on a variety of teeth from both the upper and lower jaw areas, after the periodontal tissues had healed. A procedure that was performed involved preparing a tooth for metal-ceramic crowns, which included a circumferential chamfer margin configuration. To secure the provisional restorations created from the diagnostic wax-up, zinc oxide eugenol temporary cement was used. During a three-month period, the patient utilized the provisional restorations that were positioned at the new vertical dimension.

The selection of shades was made to facilitate the completion of the restorations. Crowns composed of porcelain fused to metal (PFM) and occlusal porcelain were created following the guidelines set forth by the porcelain manufacturer. (Figure 4 and 5)



Figure 5: Post-Operative Cast Models with crown



Figure 6: Radiographs of the patient

valine, another amino acid found on the 11th chromosome of sickle hemoglobin. The sickle shape occurs in erythrocytes that contain a high level of hemoglobin S, particularly in situations where there is insufficient oxygen in the blood.⁷

To accurately diagnose sickle hemoglobinopathy, it is essential to perform a thorough history and physical examination of the patient right after the diagnosis is made. Individuals diagnosed with classic sickle cell disease usually have a clear understanding of their condition and may have experienced several hospitalizations due to painful crises, episodes of significant anemia, numerous blood transfusions, and repeated occurrences of bacterial pulmonary infections. Furthermore, these individuals have encountered this condition repeatedly.^{8,9}

The dental management of the presented case was challenging, given the patient's medical condition and the significant wear on their teeth. Maintaining close communication with the patient's hematologist before each dental appointment is crucial for adequately preparing the patient for various surgical and dental interventions. When evaluating the need for blood transfusions and antibiotic coverage before specific dental procedures, it is important to consider the severity of the dental intervention in relation to the standard local anesthetic agents used. Several studies have reported management strategies that are comparable. Throughout the treatment process, we ensured that patient appointments were concise and comfortable, while also striving to minimize lengthy and complex procedures whenever feasible.¹⁰

In this case, the use of PFM crowns led to a notable retention rate, even though the clinical crowns were somewhat short. Furthermore, the use of glass-ionomer luting cement proved effective in preserving a strong bond to the tooth structure and helping to prevent tooth decay. The follow-up demonstrated a high level of patient satisfaction over duration of six years, indicating its success.

Conclusion

Sickle cell anemia is characterized by a range of clinical manifestations and varying levels of severity. The manifestations and degrees of severity are influenced by several factors, such as the stage at which the disease is identified, the patient's age, the frequency of hospitalizations, the need for blood transfusions, and the necessity for ongoing medication, among other considerations.

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