Dental Health in Sickle Cell Disease

Salma M. AlDallal Haematology Laboratory Specialist, Amiri Hospital, Kuwait
Mobile: +965-90981981 E-mail: dr.s.aldallal@outlook.com

INTRODUCTION

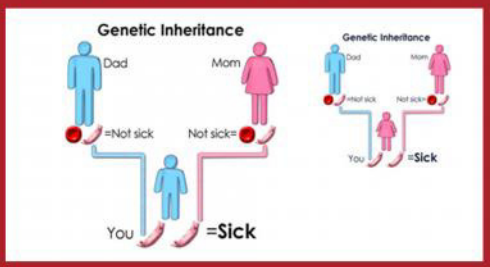
What is Sickle Cell Disease (SCD)?

- SCD is a genetic disorder caused due to replacement of glutamic acid by valine in position 6 at the N-terminus of the beta-globin chain as shown in Fig. 1. Patients with SCD exhibit two copies of the sickle cell gene, inheriting a copy from each parent; hence, both parents must have the sickle cell trait or gene to pass on to their children. Fig.2 shows the genetic inheritance pattern of the disease.

Fig.1. Replacement of Glutamate by Valine amino acid.

- The replacement results in formation of hemoglobin S (HbS).

Fig.2: The genetic inheritance pattern of sickle cell disease.



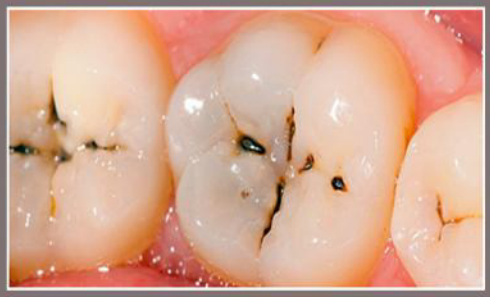
SCD Manifestation:

- Usually manifests early in childhood.
- Common oral manifestations are mucosal pallor, radiographic abnormalities, yellow tissue coloration, delay in tooth eruption, enamel disorders, dentin mineralization disorder, changes in the tongue's superficial cells, malocclusion, and a degree of periodontitis.

Dental Health and SCD:

Dental caries are the most common dental diseases in SCD patients (Fig. 3). These are teeth disorders characterized by dentin demineralization and enamel and cementum destruction.[1] For treating dental problems, dental surgeons should have an in-depth understanding of the pathophysiology of SCD. This will enable them to determine treatment plans so that they can also take systemic conditions into consideration.[1,2]

Fig.3. Dental caries in the molar teeth.



METHODS

- Blood investigations: A blood test is done to check for HbS.
- Hemoglobin electrophoresis: It is done to identify the variant of abnormal hemoglobin.

RESULTS & DISCUSSION

Blood investigations revealed the blood picture characteristic of sickle cell anemia (Fig. 4). Hemoglobin electrophoresis revealed HbS and HbF (Fig. 5).

Figure 4: Blood film of a patient with SCD. The black arrows show sickle-shaped cells (S).[5]

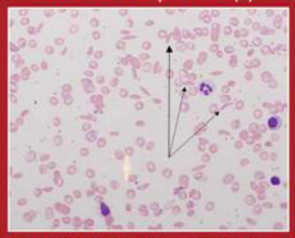
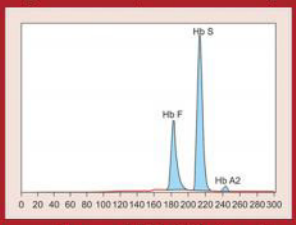


Figure 5: Hemoglobin electrophoresis showing HbF and HbS.



In SCA, the abnormal hemoglobin sticks together when it gives up its oxygen to the tissues. Sickle-shaped cells do not move easily through blood. They become hard and sticky that results in clump formation. The clumps block blood flow in the blood vessels. The clinical manifestations of the disease are firstnoted between 6 months and 3 years. Microvascular occlusions are thought to be responsible for most of the clinical manifestations of SCD.[3] Apart from this, SCD is associated with a number of dental complications as shown in Table 1.

Table 1: Reported dental complications of sickle cell disease.[1]

Dental Complications of Sickle Cell Disease
Dental Caries
Dental Erosions
Infractions
Hypodontia
Malocclusions
Pulp Necrosis
Abnormal Trabecular spacing
Infection

In most cases, the SCD patients are aware of their diagnosis and have experienced multiple hospitalizations due to episodes of severe anemia, painful crises, multiple blood transfusions and so on.[4] When treating patients with SCA, the dental surgeon should institute aggressive preventive dental care including: diet control, oral hygiene instructions, tooth brushing, flossing instructions and fluoride gel application.[1]

CONCLUSION

SCD is presented with a wide variety of clinical symptoms, and varied degrees of severity that can be determined based on the phase during which the disease is diagnosed, the patient's age, past hospitalization history, drug use pattern, and blood transfusions.

It is highly critical that the dental professionals should be aware of the oral manifestations and physiopathology of the disease. Additionally, they should cautiously obtain the patient's clinical history and collect information about specific features so that they can build up a plan for any dental treatment as per the patient's limitations and requirements.

Dental care and dental hygiene among the SCD patients is a neglected area that requires further studies. The strength of this study is to emphasize on the importance of dental complications in SCD, which often worsens other clinical features of the disease including vaso-occlusive crises and systemic infections.

Literature Cited

1. Ballas S.K., Lobo C.L.D.C., Cavalcanti W.E. (2014).Dental Complications of Sickle Cell Disease. J Interdiscipl Med Dent Sci. 2:152. doi:10.4172/2376-032X.1000152.
2. Acharya S. (2015). Oral and Dental Considerations in Management of Sickle Cell Anemia. Int J Clin Pediatr Dent. 8(2): 141–144.
3. Hebbel R.P., Osarogiagbon R., Kaul D. (2004). The endothelial biology of sickle cell disease: inflammation and a chronic vasculopathy. Microcirculation. 11(2):129-51.
4. Smith J.A., Ann N.Y. (1989). The natural history of sickle cell disease. Acad Sci. 565:104.
5. Piccin A., Fleming P., Eakins E., McGovern E., Smith O.P.,Mc Mahon C. (2008). Sickle cell disease and dental treatment.J. Ir. Dent. Assoc. 54:75–79.