

Oral and radiographic manifestations brought on by Sickle Cell Anemia - Clinical Case Report

Vanessa Helena Pires DINIZ¹, Arthur Diniz VIAL¹, Raquel Tolentino Dornelas ALVES^{2,*}, Flávio Ricardo MANZI¹ and Amaro Ilídio Vespasiano SILVA¹

¹ Pontifical Catholic University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.

² Faculty of Ipatinga, Fadia. Ipatinga, Minas Gerais, Brazil.

Abstract

Sickle cell anemia is a hereditary, monogenic blood disease characterized by chronic hemolytic anemia and vasoconstricting phenomena leading to acute painful crises and chronic and progressive tissue damage. It is the most common disease among hemoglobinopathies in Brazil and worldwide. The disease originated in Africa and was brought to the Americas by the forced immigration of slaves. Sickle cell patients have clinical conditions that can be intensified during dental treatment by precipitating vasoconstrictive seizures. In this clinical case, paleness was detected in the buccal buccal and the tongue despapilada. In the intra-oral radiographic examinations, areas suggestive of bone sclerosis and radiolucent areas between the root apices were found. Maxillary protrusion due to medullary expansion was found in lateral cephaladiography. In scintigraphy and magnetic resonance imaging, areas corresponding to bone infarcts and osteonecrosis were found. The dental surgeon performs an important function, being able to diagnose the oral manifestations through clinical and/or radiographic examination. By acting preventively, we can reduce the complications and improve the quality of lives of these patients.

Keywords: Sickle cell anemia; Dentistry; Radiography

1. Introduction

Sickle cell anemia is an inherited blood disease characterized by abnormal red blood cells caused by a genetic mutation of the hemoglobin A (HbA) molecule, which is renamed as hemoglobin S (HbS) [1].

Under conditions of hypoxia, dehydration, physical exhaustion, and acidosis, the mutant molecules deform and undergo polymerization, with falcization of the red blood cells; hence the "sickle cell" name. Sickling shortens the half-life of the erythrocyte. Deformed red blood cells cause difficulty in transporting oxygen to tissues and blood circulation, which causes vasoconstriction and infarction in the affected area. Vasoconstriction and infarction in turn cause ischemia, pain, necrosis and dysfunction, and chronic hemolysis resulting in jaundice and anemia [2]. In an attempt to compensate for the destruction of red blood cells, there is hyperplasia and compensatory expansion of the bone marrow, causing changes in the bone structures that can be observed radiographically [3].

Oral problems brought about by sickle cell anemia include mandibular osteomyelitis, mandibular nerve paresthesia [4], pulpal necrosis [5], facial enlargement, and gingival hyperplasia [6].

* Corresponding author: Raquel Tolentino Dornelas ALVES
Faculty of Ipatinga, Ipatinga, Minas Gerais, Brazil.

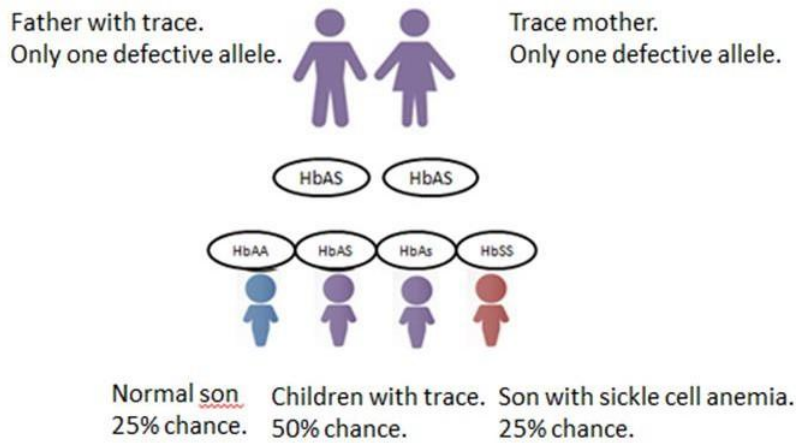


Figure 1 Genetic combination for sickle cell trait and sickle cell disease.

Source: image createdmade by the author.

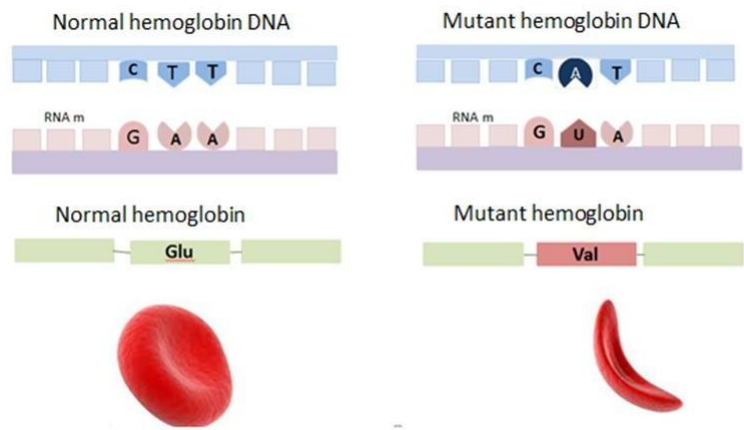


Figure 2 Image of normal hemoglobin and hemocyte mutants. Source: made by the author.

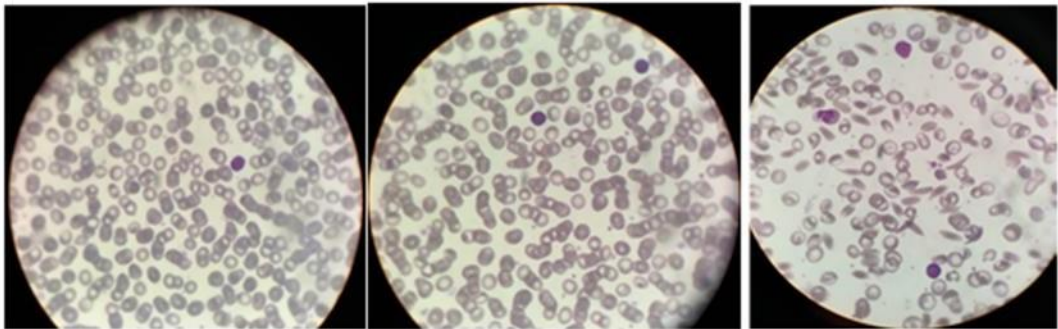


Figure 3 Microscopic image of normal red blood cells (HbAA), red blood cells with sickle cell trait (HbAS) and red blood cells with left sickle cell anemia (HbSS). From left to right.

Source: Hemominas / Hemocentro Foundation Laboratory of Belo Horizonte-MG.

1.1. Falciform anemia: Bucal and radiographic changes

People with sickle cell anemia (HbSS) are more susceptible to infections, periodontal disease, and cavities. [7]

The most common oral manifestations are: pallor of the mucosa with late eruption, hyponatalization and hypomineralization in enamel and dentin, calcifications and pulp necrosis, hypercementose and alterations of the cells of the surface of the tongue such as: smooth tongue, bleached, and despapilada [8]. Present malocclusions occur due to the protrusion of the maxilla and retrusion of the lower incisors, with presence of diastema between the maxillary central incisors [9].

Asymptomatic pulpal necrosis can be caused by a vasoconstriction of the microcirculation of the dental pulp of healthy teeth. [10]

1.2. Dental treatment

Oral complications are directly related to hemolytic anemia, bacterial infections, and vasoconstrictive seizures.

2. Clinical case report

A 47-year-old female patient with melanoderma sought dental treatment and required periapical radiography of mandibular and periapical molars from tooth 25. These radiographs were performed on the Sirona Heliodent Plus Digital Dental X-ray apparatus using the following exposure factors: 60Kv, 0.6 mA, t: 0.64s. A panoramic radiograph (77Kv, 10mA, t: 15.1s) was performed as routine examination and a lateral teleradiography (70 Kv, 12 mA, t: 1s) was performed on a Kodak 9000 digital CCD. In these tests, suggestive radiopaque areas were observed indicating bone sclerosis or neoformation of bone, as well as radiolucent areas between the apices of the teeth and the lower border of the mandible. These areas can be seen in Figures 4, 5.

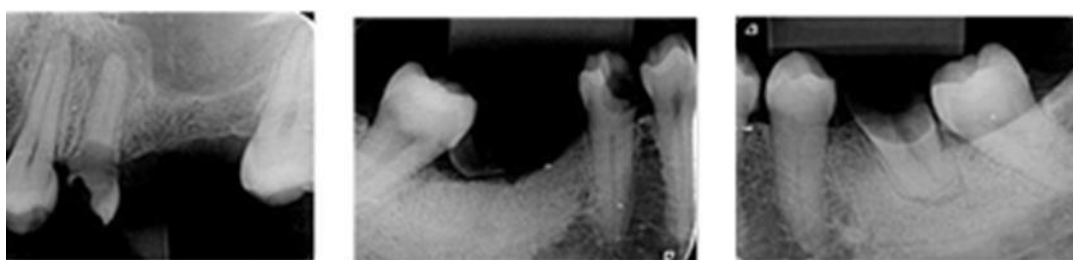


Figure 4 Periapical radiographs of tooth 25 and lower molars.



Figure 5 Panoramic radiography.

Shows the pale-faced melanoderma patient in a frontal intraoral photograph showing diastema between teeth 11 and 21 and in an inferior occlusal photo showing the tongue not papillate.



Figure 6 Patient's smile photograph, frontal intraoral image, and inferior occlusal photo showing the little papillary tongue.

In follow-up examinations of the disease, the patient underwent bone scintigraphy, radiography of the coxofemoral joint, and left shoulder magnetic resonance imaging. In the bone scintigraphy examination, images of the whole body and skull were obtained, showing diffuse heterogeneous distribution throughout the skeleton, with hypercaptation of the ^{99m}Tc radiopharmaceutical on bone ends, long bones, humeral heads, elbows, wrists, femoral heads, knees, ankles and calcaneus, corresponding to areas of bone infarcts in the phase of late remodeling. In the coxofemoral radiography, we can see aseptic osteonecrosis of the head of the left femur associated with difficulty of ambulation. The magnetic resonance imaging of the left shoulder was performed without contrast and a bone discontinuity line was observed. This discontinuity line showed a predominantly transverse pathway, with irregular contours, affecting the greater tuberosity of the humeral head. An intra-articular glenohumeral extension could represent osteonecrosis.

3. Discussion

Sickle cell anemia is a hereditary blood disease that produces sickle-shaped red blood cells (HbSS) with reduced oxygen transport capacity for tissues and organs [1], causing vasoconstriction and infarction in the affected area [2] that can be observed in Figure 3.

Sickle cell disease (SCD) includes a group of genetic disorders characterized by the predominance of hemoglobin S (Hb S). The hemoglobin S (HbS) gene may combine with other inherited hemoglobin changes, such as C, D, beta, and alpha-hemoglobin hemoglobin. The set of combinations SS, SC, SD, SE, Sbeta-thalassemia is called Sickle Cell Disease. These changes include sickle cell anemia, which is a disease in which the patient has two homozygous genes (HbSS) from asymptomatic parents, and the sickle-cell trait (HbAS) [11].

The most common manifestations of sickle cell anemia in oral mucosa are pallor of the mucosa resulting from the hemolysis of the red blood cells, which generates jaundice in the patient. Alterations of the cells of the mucosa of the tongue may occur, such as depopulated and discolored tongue [8] observed in Figure 6.

Bone changes of the jaws consist of decreased radiodensity. The medullary spaces occur as radiolucent areas between the apices of the teeth and the lower border of the mandible. Medullary hypertrophy results in loss of fine trabecular bone and formation of large medullary spaces [13], which can be observed in Figures 4 and 5.

The mandible is a less vascularized bone than the maxilla. A vasoconstrictive crisis in the mandible leads to ischemia and necrosis of the bone, presenting broad medullary spaces that become necrotic when infarcted. The oral flora can reach this area by the periodontal ligament or in a hematogenous manner. Providing a good culture medium for bacteria.

Low income also contributes to an increased risk of dental caries that should be avoided to reduce the number and severity of sickle cell crises [14], observed in Figures 4 and 5, Patients with dental losses and multiple caries.

The compensatory hyperplasia of the medullary spaces can cause expansion of the maxilla, generating an Angle Class II malocclusion which may be associated with the presence of anterior superior diastems [8].

Dental treatments[2] that are both planned and small can be completed using a local anesthetic without a vasoconstrictor. More invasive procedures increase the chances of infections, justifying the use of prophylactic antibiotic therapy. People with sickle cell disease do not have an increased risk of bleeding. Pain can be treated with paracetamol, dipyrrone or codeine. The use of salicylate leads to acidosis and interferes with platelet aggregation and therefore should be avoided. Some authors report that the use of vasoconstrictors may impede local circulation and

cause heart attack, while other authors claim that vasoconstrictors have no effect on local circulation despite hypovascularization. [8].

It is difficult to make a differential diagnosis between osteomyelitis and systemic bone necrosis. The symptoms of osteomyelitis are gingival sulcus exudate, facial edema, and lymphadenopathy. Radiography can show bone destruction. Generalized painful crises may be accompanied by mental nerve neuropathy and paresthesia of the lower lip [12].

Sickle cell disease in pregnant women with periodontitis increases the predisposition to preterm births [10].

3.1. Tests

Both isoelectric focusing electrophoresis (IEF) and high performance liquid chromatography (HPLC) can be used for the diagnosis of sickle cell disease.

3.2. Relationship with malaria

Malaria is a parasitic tropical disease that is the major cause of social and economic problems in the world [15]. Malaria is caused by the protozoan of the genus *Plasmodium*, transmitted to man by blood [16]. *Plasmodium* attacks liver and red blood cells by degrading hemoglobin to provide a source of nutrients [17]. People who have sickle cell anemia have this mechanism, making the sickle cell carrier sick with malaria.

All organs and tissues of the body are under risk of damage due to sickling. The clinical manifestations are due to vasocclusion, followed by infarction in several organs. In the bone scintigraphy examination (Figure 8), in the radiographic examination of the patient's coxofemoral joint (Figure 9) and in the MRI of the patient's left shoulder (Figure 10), we can visualize areas of bone infarcts in a late remodeling phase corresponding to areas of lower oxygenation rates in patients with sickle cell anemia.

4. Conclusion

The present report allows dental professionals to increase their knowledge about patients with sickle cell anemia, to clarify the limits of performing dental procedures on patients with sickle cell anemia, and to identify the radiographic manifestations of the damage brought about by this disease.

Compliance with ethical standards

Disclosure of conflict of interest

We authors declare that no conflict of interest is exist.

Statement of informed consent

The declared consent was obtained from all individuals included in the study.

References

- [1] Souza PHG, et al. Skeletal craniofacial changes in patients with sickle cell anemia in the city of Juiz de Fora. 2008.
- [2] Lopez FA, Campos D. Pediatrics Treaty: Brazilian Society of Pediatrics. Barueri: Manole. 2007.
- [3] Mourshed F, Tuckson CR. A study of the radiographic features of the jaws in sickle-cell anemia. Oral Surgery Oral Medicine Oral Pathology. 37(5).
- [4] Kelleher M, et al. Oral complications associated with sickle cell anemia. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontics. 82(2): 225-228.
- [5] Kaya AD, et al. Pulpal necrosis with sickle cell anemia. International Endodontic Journal, Malden. 37(9), 602-606.
- [6] Soares MRPS, et al. Sickle cell anemia: oral manifestations and multidisciplinary - clinical case report. 2013.
- [7] Sonis ST, Fazio RC, Fang L. Principles and practices of oral medicine (2nd ed.) Rio de Janeiro, Brazil: Guanabara Koogan. 1995.

- [8] Silva RBP, Ramalho AS, Cassoria RMS. Sickel cell anemia as a public health problem Brazil. *Rev Saúde Pública*. 27(1): 54-8.
- [9] Shnorhokian HI, Chapman DC, Nazif MM, Zullo TG. Cephalometric study of American black children with sickle-cell disease. *ASDC J Dent Child*. 51(6): 431-3.
- [10] Oral Health Handbook in Sickel Cell Disease. Series A. Standards and Technical Manuals. 2nd edition, Brasília – DF (sickle cell disease). 2007.
- [11] Anvisa - National Agency of Sanitary Surveillance. Manual of diagnosis and treatment of sickle diseases. Brasília, DF. 2002.
- [12] Sanger RG, Bystrom EB. Radiographic bone changes in sickle cell anemia. *J. Med*. 32(2): 32-7.
- [13] Laurence B, George D, Woods D, Shosanya A, Katz RV, Lanzkron S, et al. The association between sickle cell disease and dental caries in African Americans. *Spec Care Dent*. 26(3): 95-100.
- [14] Elisabeth Carmen Duarte – Universidade de Brasília (UnB) Leila Posenato Garcia – Instituto de Pesquisa Econômica Aplicada (Ipea) Brazil. Ministry of Health. From Chapter VI of section I of Ordinance No. 05 / GM / MS, dated September 28, 2017, published in Official Gazette no. 190, of October 3, 2017, page 360, Supplement DOU section.
- [15] Ventura AMRS, Pinto AYN, Silva RSU, et al. *Plasmodium vivax* malaria in children and adolescents - epidemiological, clinical and laboratory aspects. *J Pediatr*. 75(3): 187-94.
- [16] Wanderlye DMV, Andrade JCR, Alves MJCP. Contribution to epidemiological information on malaria in the state of São Paulo. *Rev Soc Bras de Medicina Tropical*. 20(3): 143-6.
- [17] Goldberg DE, Slater AFG. The pathway of hemoglobin degradation in Malaria Parasites. *Parasitology Today*. 8(8): 280-3.