

Oral Considerations in the Management of Sickle Cell Disease: A Case Report

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Abstract

The phenomenon of erythrocyte sickling observed in sickle cell anaemia is responsible for ischaemia and tissue infarction compromising several organs and systems including the mouth and face. This brief paper reports the case of a 17-year-old female with a complicated sickle cell anaemia, hypertension and paraplegia (after an ischaemic stroke at the age of six years). Oral examination revealed the absence of tooth 12, fractures of teeth 11, 21 and 22 (from trauma), active caries lesions in the enamel of teeth 36, 37 and 46, mucosal pallor, and a “smooth tongue”. Oral radiographs revealed bone rarefaction and trabecular bone coarsening. Dental surgeons and physicians should be aware of the general and oral abnormalities that can be present in individuals with sickle cell anaemia to allow for preventive measures and implementation of effective treatment options.

Key Words: Sickle Cell Anaemia, Haemoglobin Diseases, Oral Manifestations, Dentistry, Disease Prevention

Introduction

Sickle cell anaemia is a genetic disease caused by replacement of glutamic acid by valine in position 6 at the N-terminus of the beta-chain of globin, thus resulting in haemoglobin S [1-2]. Under conditions of hypoxia, erythrocytes that predominantly contain haemoglobin S take on a shape resembling a sickle [1-4]. This sickling is reversible through increased oxygen levels, although constant changes in shape result in cell membrane lesions that make the cells rigid, preventing them from returning to their normal state [5-6]. Sickle cell anaemia is diagnosed through a neonatal screening test, in which drops of blood are drawn from the newborn's heel to permit haemoglobin electrophoresis or chromatography to be performed [7].

The reduction in oxygen-transport capacity results in circulatory difficulties, including vaso-occlusive conditions, which diminishes the lifespan of the red blood cells to approximately 20 days [8-9]. In addition, sickled erythrocytes may occlude the microvasculature because they adhere better to the endothelium, thereby impeding the blood flow and causing tissue anoxia, necrosis and pain [10].

The main oral manifestations and complica-

tions of sickle cell disease are mucosal pallor, yellow tissue coloration, radiographic abnormalities, delayed tooth eruption, disorders of enamel and dentine mineralisation, changes to the superficial cells of the tongue, malocclusion, hypercementosis, and a degree of periodontitis that is unusual in children [9,11-14].

For dental treatment to be carried out, it is recommended that dental surgeons have an understanding of the pathophysiology of this disease, enabling them to determine treatment plans so that they can also take systemic conditions into consideration [15-17].

Case Report

A 17-year-old female patient with sickle cell anaemia, accompanied by her guardian, attended the outpatient dental clinic of a university hospital to undergo restorative treatment on her anterior upper teeth, which had been fractured during a convulsive crisis. As a result of a bilateral ischaemic stroke at the age of six years, she was paraplegic. She gave a history of rheumatic fever at the age of eight years and had arterial hypertension. She was undergoing blood transfusion every 30 days due to

a past history of strokes and was taking captopril, digoxin, furosemide, and folic acid.

Knowing that the patient was paraplegic and had sickle cell anaemia, a dental treatment plan was developed to address these two problems before any oral therapy was implemented. A dental office accessible to wheelchairs was selected, antibiotic prophylaxis (2g of penicillin one hour prior to the dental procedure) was prescribed, and consultation with her assistant physician was undertaken to make sure that there were no other impediments to the dental treatment. With all these precautions implemented, the patient was called in for consultation.

Palpation and auscultation of the temporomandibular joint did not detect any irregularities. There were no cervical lymph nodes palpable in the cervical or submaxillary region. Evaluation of the oral soft tissues showed signs suggestive of sickle cell anaemia, including mucosal pallor and abnormalities of tongue morphology known as “smooth tongue” (Figures 1, 2 and 3). An examination of the periodontal tissues was then performed and consisted of an assessment of gingival recession, pocket-probing depth, tooth mobility, plaque and bleeding indices. The results of the bleeding index (53%)

and the plaque index (43%) suggested the diagnosis of gingivitis, indicating that the patient’s gingival response to plaque was enhanced by her underlying systemic problem.

After a dental prophylaxis, an oral examination of the hard tissues was performed with a plane mirror and a World Health Organization probe. It revealed the absence of tooth 12, fractures in teeth 11, 21 and 22 (as a result of past trauma), and active caries lesions in the enamel of teeth 36, 37 and 46.

Cervical muscle incompetence made it impossible to produce panoramic radiographs. Thus, the radiographic examination was limited to periapical and interproximal radiographs, which revealed bone rarefaction and trabecular bone coarsening (Figure 4), some regions with subgingival calculus, and loss of height of alveolar bone with bone crests rounded.

After carrying out the physical and complementary examinations, composite resin restorations were placed at 11, 21 and 22 (mesial, distal incisal faces, and lingual). Upper and lower impressions were taken so that a mouthguard could be made. The patient’s guardian was shown how to brush the patient’s teeth and gingival crevices, and fluoride toothpaste was recommended. Local anaesthetic

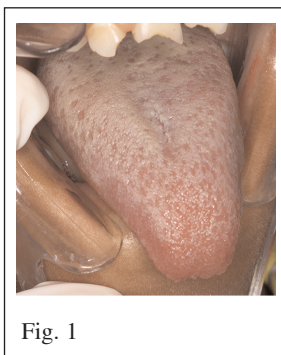


Fig. 1



Fig. 2



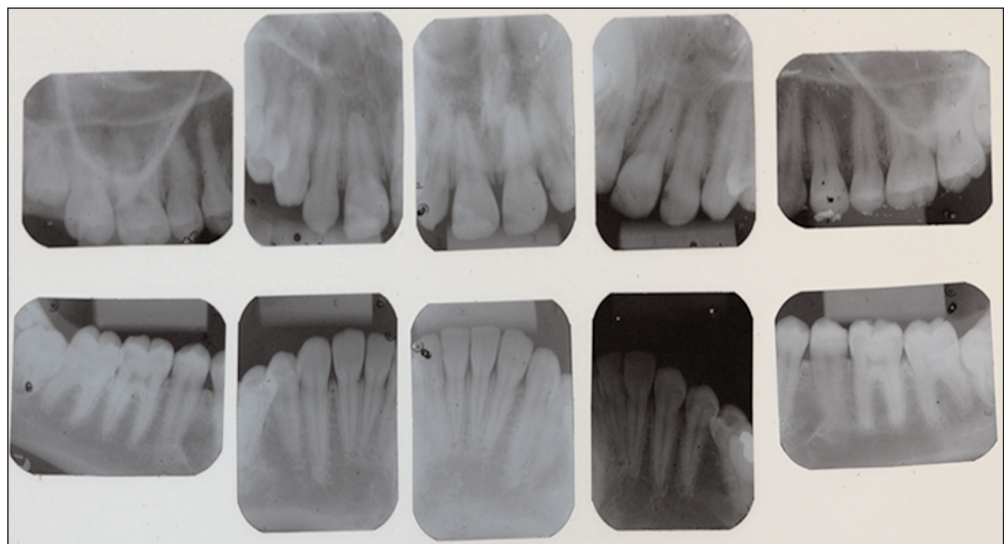
Fig. 3

Figure 1. Abnormalities of the superficial cells of the tongue.

Figure 2. Fractured incisors and the missing tooth 22.

Figure 3. Mucosal pallor.

Figure 4. Periapical radiographs showing the formation of a coarse trabecular pattern bone (teeth 37, 36, 35 and 47, 46, 45) and medullary spaces formed by loss of thin bone rarefaction and loss of height of the alveolar bone (units: 15, 16, 17, 23, 24, 25, 26, 27, and 41, 42, 43).



was not required because these procedures were non-invasive and did not cause pain.

After this initial therapy, the patient and her family were advised about future oral health care, were given dietary advice to promote a non-cariogenic meal plan, and received further instruction in tooth-brushing, the use of dental floss and tongue cleaning. Return visits at 3-4 weekly intervals for fluoride applications and oral hygiene checks were arranged.

Unfortunately, the patient suffered a new cerebrovascular accident that resulted in her death and this treatment plan could not be carried out. The patient's guardian gave written informed consent to use the images reported in this article and to report the case.

Discussion

Sickle cell anaemia can lead to many systemic complications especially in areas that are most compromised by hypoxia and infarction [4,17]. The most frequent complications are joint pain, haemolytic anaemia with jaundice, skin and mucosal pallor, leg ulcers, spleen and liver sequestration and infarction, haematuria, pulmonary dysfunction, osteomyelitis and stroke with headaches, convulsions and hemiplegia [4,18].

Although children are more vulnerable to cerebral ischaemic events, haemorrhagic events prevail in adults [4,7]. The patient presented in this report had suffered an ischaemic stroke resulting in the impairment of her muscular system that made her dependent on a wheelchair.

Repeated splenic infarctions result in spleen hypofunction with abnormalities in opsonisation, alternative complementary paths, antibody production, leukocyte function, and cell immunity [18,19]. The patient's diagnosis of rheumatic fever was closely related to malfunctioning of the spleen and had occurred at a time when she was suffering from a number of infections.

Although the oral manifestations are not exclusive to this disease, they may suggest a diagnosis of sickle cell disease [13]. Mucosal pallor and yellowed discoloration of the gingiva result from the deposition of blood pigments secondary to hyperbilirubinemia caused by large-scale destruction of erythrocytes [6,11]. In this report, the mucosal pallor was very evident and observed primarily in the oral and labial mucosa. The smooth appearance of the tongue was not compounded by poor hygiene of this organ because of the absence of a tongue coat-

ing. Hypocalcification of the enamel and tooth fractures were also observed in several teeth (*Figures 1, 2 and 3*).

The radiographic findings of distinct radiopaque areas caused by repairs to bone infarction [6] were present in both the maxilla and the mandible. The radiographs also revealed loss of the normal trabecular pattern with increased radiolucency due to decreased number of trabeculae and increased medullary spaces secondary to compensatory hyperplasia; thinning of the lower edge of the mandible. There was a coarse trabecular pattern of "staircase" shape (present mainly in the interproximal bone because of trabeculae that formed horizontal rows), presence of projections similar to "hair strands" due to secondary formation of bone tissue as compensation for resorption that occurred during bone marrow expansion. There was also thickening of the lamina dura, loss of height of the alveolar bone, and distinct radiopaque areas caused by repairs to bone infarctions [10]. The periapical radiographs showed a coarse trabecular pattern, increased medullary space, bone rarefaction and loss of height of the alveolar bone, thereby minimising the bone manifestations of the disease in a significant manner. These signs should alert clinicians to the possibility that a patient has sickle cell anaemia. The condition is more prevalent in Afro-Caribbean populations and is therefore common in Brazil where the authors of this case report work. However, it is rarely seen in a number of other countries, including those of Eastern Europe.

Conclusions

Sickle cell anaemia presents with variable clinical manifestations, and different degrees of severity that depend on the stage at which this disease is found, the patient's age, number of hospitalisations, need for blood transfusions and need for continuous drug use, among others. It is important that all clinicians are aware of the physiopathology and oral manifestations of sickle cell anaemia and that dental surgeons should carefully obtain the patient's clinical history and information about particular features so that they can plan any dental treatment such that it is appropriate to the patient's limitations and needs. As with all treatment plans, its goals should be to achieve and maintain oral health and to reduce the risks of dental infections.

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Contributions of each author

- SM was the dental surgeon responsible for the clinical care of the patient's oral problems.
- SM, RPCA and CA participated in the design of the study, literature search, analysis and interpretation of data, drafting and critical review of the final version of the manuscript.

- All authors acknowledge that they have participated sufficiently in the work to take responsibility for its content.

Statement of conflict of interest

The authors state that there is no financial or other relationship that might lead to conflict of interest.

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