Sickle cell disease and common clinical manifestations and treatment: a case study

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ABSTRACT

Introduction: Sickle cell disease (SCD) is a blood disorder that affects the framework of red blood cells and everyday function, leading to many concerns including anemia, pain, and increased risk of infection. Oral characteristics can be overlooked but can majorly affect the patient's day-to-day activities and quality of life. Case description: This short communication reports the clinical and radiographic oral manifestations of SCD and the critical nature of early intervention and collaborative maintenance. Discussion: This case study describes a 13-year-old African American male with sickle cell trait and classic oral findings such as enamel hypomineralization, pulp stones, hypercementosis, and the "stepladder" trabecular bone pattern. Other important characteristics include poor oral hygiene and decreased access to care due to socioeconomic factors contributing to increased disease migration. Conclusion: Preventive strategies including routine assessments, effective patient education, and consistent collaboration with the medical team are critical in improving oral and systemic health outcomes.

Keywords: anemia, sickle cell; anti-inflammatory agents, non- steroidal; caries, dental; hemoglobin, sickle; hypomineralization, dental enamel; malocclusion; socioeconomic status, low

CDHA Research Agenda category: oral health frameworks

PRACTICAL IMPLICATIONS OF THIS RESEARCH

- This report increases awareness of oral health professionals regarding radiographic and clinical characteristics for patients with sickle cell disease
- Common conditions for a patient with sickle cell disease may be overlooked as another condition by dentists and dental hygienists
- Collaborating with many medical and oral health specialists may help manage the ill effects of sickle cell disease

INTRODUCTION

Sickle cell disease is an autosomal recessive disease causing "abnormal Hemoglobin S which distorts the shape of red blood cells into a sickle or crescent moon shape."[1] The red blood cells become rigid, and blood turns very viscous. This condition may cause clots to form in smaller vessels and organs causing blockages in blood flow and decreased oxygen levels to bones and other tissues leading to anemia, severe pain, infections, and other complications. Most critically, organs such as the brain, heart, kidneys, and lungs are affected by the lack of oxygen levels. Because hemoglobin in red blood cells plays a vital role in transporting oxygen, sickle cell anemia (SCA) causes widespread systems of oxygen deficiency, such as fatigue, irritability, dizziness, lightheadedness, tachycardia, and shortness of breath.[1]

Sickle cell disease is a rare blood disorder that is more evident in the United States (approximately 100,000 cases) than in Canada (approximately 6,000 cases).[2,3] When evaluating statistics from developing countries such as Angola, Nigeria, and the Democratic Republic of Congo, sickle cell disease (SCD) is more prevalent. In comparison, world prevalence is very low at approximately 7% but is rising due to global immigration.[4,5] SCD has a very high mortality rate with many patients only living into their thirties and forties.[1] Dental infections have been linked to ophthalmic or pulmonary disorders that cause premature crises and death.[6] Survival age is normally in the 40's for both males and females; however, newer treatment therapeutics are resulting in longer life spans.[6]

Genetically predisposed patients who have lower socioeconomic status (SES), lower access to care, and limited or no insurance coverage are more prone to have SCD with concomitant dental pathologies.[7] When patients have SCD, low SES, as well as dietary insecurity, lack of access to care may affect the severity of the disease.[8] Several co-morbidities

are found in patients with SCD and may cause severe complications with treatment and coping mechanisms in everyday life situations.[7]Faith-based treatments help some cope with all the dynamics of this complicated disease.[8]

Patients with SCD often have caries, mostly due to poor oral hygiene, which elevates the risk for caries, gingivitis and periodontal disease.[5,9] This is most likely linked to lack of resources and knowledge.[9] In certain instances, patients with SCD have increased caries rates due to enamel hypomineralization; and plaque accumulation exacerbates the condition affecting much of the dentition.[5] A decrease in the prevalence or severity of caries may be seen, if patients are taking daily antibiotic therapy which is commonly given in the first five years of life for those with SCD.[5] Systemic amoxicillin and metronidazole in unison are a gold standard for SCD and do show promise for treatment.[4] Non-steroidal anti-inflammatory drugs (NSAIDs) such as Ibuprofen and Naproxen have been shown to help patients control pain and inflammation and may decrease the incidence of periodontal disease and destruction of periodontal structures.[6] However, their use should be in consultation with the patients' physician if renal disease is suspected. In addition, the daily use of folic acid has shown some promise as an adjunctive treatment to lower the chance of developing pulpal necrosis which may be associated with hypoxia from vasoconstrictor use.[4,10] Along with folic acid, hydroxyurea may be prescribed for patients with SCA.[11] In severe cases of SCD, blood transfusions can be used to help the patient's condition, however, this will make them more prone to different forms of infection.[11]

Sickle cell anemia is a specific type of SCD and tends to be very common in patients who are of African descent[10] are from Brazil (1 in 200 births).[5,12] Prevalence also is found in people of Mediterranean descent. Being an autosomal recessive gene, both parents must have the affected gene to transfer it to future generations. If only one parent has the gene for SCA, the child is

recognized as a 'carrier' of the sickle cell trait.[10] This condition not only affects the patient, but the entire family unit. Not only do mental health struggles plague the patient with SCA due to chronic pain, but families have additional stress and concern for their loved one due to the potentially high mortality rates associated with this disease.[13] Delays in growth and puberty along with small gonad development and endocrine dysfunction are common in children and adolescents with SCA and cause complications later in life.[4]

The importance of discovering clinical manifestations of SCD assists the dental professional with assessing, diagnosing, and treating this condition in the dental setting. In children, delayed tooth development, hypodontia, gingival enlargement, and, in some cases, abnormal tooth eruption sequence, and pallor are seen in gingival tissues.[6,14] Class II occlusion and overjet caused by prominent maxillary jaw growth and dysplasia, are commonly seen in these patients .[15,16] The prominent maxilla is due to a high erythropoietic drive in SCD, such as in thalassemia which creates the tendency for maxillary hyperplasia and malocclusion.[17] Along with malocclusion, patients with SCD can have more crowding and protrusion with the anterior teeth.[10] Periodontal conditions are evaluated early via images, periodontal screening that includes periodontal index, gingival index, and bleeding on probing (BOP); and higher measurements are common for patients with SCD.[6]

"Several dental effects are commonly found in children with sickle cell disease, such as delayed tooth eruption, hypoplasia, and enamel hypomineralization on labial surfaces of maxillary incisors, hypercementosis, glossitis, pulp stones or pulp denticles, and asymptomatic pulpal necrosis due to thrombosis in the blood vessels." [5,6,13,18]

Patients with SCD can have many concerning crises caused by dehydration, infections, extreme temperature fluctuations, hypoxia, and pulmonary disease.[6] The long bones, spinal column, and joints in the body may develop severe pain, which in turn can cause weakness, chronic fatigue, and bone deformity.[6,19] A promising development is the use of gene therapy to decrease levels of pain or to even cure SCD.[20] Across the world, iron chelation therapy has been used as a variable treatment to blood transfusions for SCD.[21] The primary purpose of iron chelation is to protect tissue from iron toxicity resulting from Hb breakdown and to eliminate the excessive amount of stored iron in the body.[21] Many times, this type of treatment will lower the risk of organ failure in SCD patients.

Inflammation may be seen in the temporomandibular joint (TMJ) area in SCD patients, but TMJ pain is not prevalent.[12] However, about 50% of SCD patients may experience orofacial pain due to facial swelling in other locations than the TMJ, such as the mandible, frontal bone, and maxilla and this contributes to headaches in 77% of the cases.[12,14]

"Mental nerve neuropathy or "numb chin syndrome" can occur in patients with some form of mandibular bone injury or disease in adults."[22] This condition has been discovered more readily in adult patients with SCA.[23] The numbness usually affects the chin and lower lip due to decreased oxygen levels causing osteomyelitis following surgery; or spontaneously by relative compression of the mental nerve as it exits the mental foramen, or when the SCD is in an active state.[8,22] This specific type of anesthesia or paresthesia with the mental nerve can also involve the inferior alveolar nerve as well causing more profound numbness.[18] In certain instances of mental nerve numbness, it is possible that patients may develop ulcerations on the lip region due to excessive and repetitive trauma.[24] The numbness may resolve in days, months, or up to a year depending on the severity of the case.

Different types of imaging (ie. cephalometric imaging) are utilized to assess the patient regarding size and development of bones within the head and neck region. In certain types of computed tomography (CT) scans, an increased prominence of the zygomatic, frontal, and parietal bones can be seen—this can contribute to the craniofacial pain of some patients with SCD.[8]

On standard periapical or panoramic images there are several characteristics that can be assessed. Very common radiographic discoveries may include enlarged medullary space, "stepladder" appearance, pulpal sclerosis or pulpal obliteration, and decreased thickness of the inferior border of the mandible.[6] The lamina dura can be disrupted or completely missing on some teeth and evidence of hypercementosis with thickness or change in the apex of the root structure can be observed.[25] When assessing the trabeculation in the jaw, a decrease in trabecular bone or "honeycomb effect" is evident with excessive radiolucency on the dental images.[26] Panoramic images are used to investigate the radiomorphometric indices to evaluate for decreased bone density.[27]

The problem is that sickle cell disease and trait have well-defined systemic effects; however, some oral manifestations, especially in children, are often misinterpreted or not properly diagnosed by dental professionals. Primary and specialized care and maintenance in both the dental and medical professions are imperative, and if not accomplished, this can delay effective treatment modalities and impact dental treatment throughout the life span for patients with SCD or sickle cell trait.

This short communication reports clinical and radiographic characteristics of a 13-year-old African American male with sickle cell trait. This report proposes to increase awareness among oral health professionals about the diverse clinical manifestations that may mimic other

diseases or conditions.

CASE DESCRIPTION

A 13-year-old black male was seen for a routine examination and radiographic survey including four bitewings and one periapical around teeth #46 and #47. At a subsequent appointment, select periapical radiographs and a panoramic radiograph were exposed for evaluation. The patient reported a medical history including asthma, attention deficit hyperactivity disorder (ADHD)—both of which the patient is in treatment with a physician, and an allergy to Penicillin, animal dander, and pollens. No other health history or family history is noted. In addition, the patient does not have any restrictions or issues with playing sports; has had no emergency room visits, and his physician does not have any concerns with the current health of the patient. He also reported bleeding gums when brushing or flossing and sensitivity to hot, cold, and sweets. He indicated that toothaches "come and go" with pain being more localized, but he reported feeling sensitivity in certain teeth while chewing food. His updated periapical and panoramic radiographs and clinical examination revealed multiple areas of gross primary and secondary caries, a progression since his last appointment, as well as the necessity for a referral for root canal therapy (RCT). Periodontal charting and assessment found localized 4- and 5-millimeter pockets on the posterior areas along with the presence of heavy plaque. Radiographs also revealed several enlarged pulp-chambers with large pulp stones and possible cemento-osseous dysplasia on the panoramic radiograph for #41and #45 (Figure 1), which is a common characteristic bone involvement with African Americans and patients with SCD. Stepladder trabeculation was also interpreted on the panoramic radiograph between #36 and #35 (Figure 1).

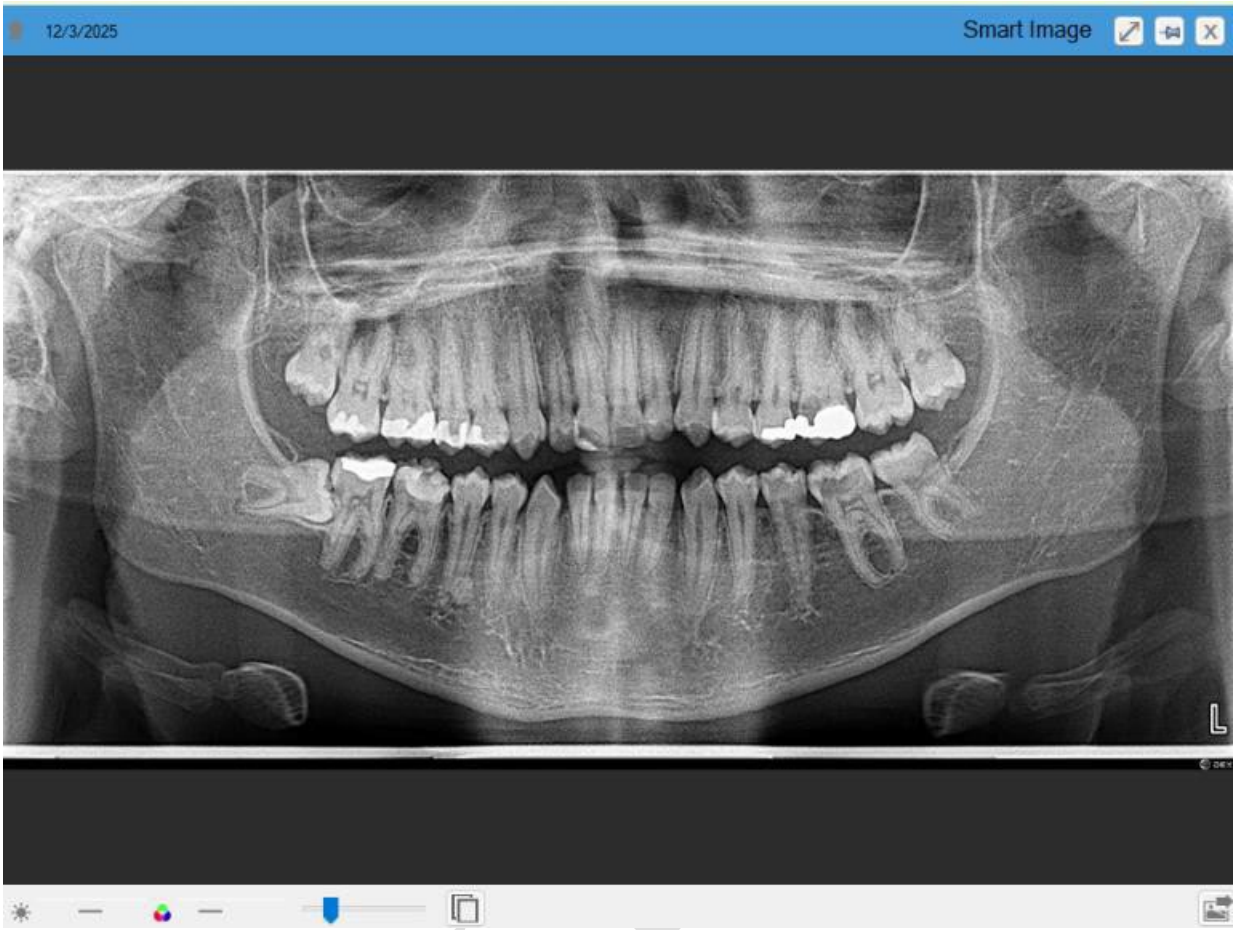

Figure 1. Image showing pulp stones in many canals, condensed round radiopacities on #41 and #45, and stepladder trabeculation pattern on the mandibular right and left quadrants

Multiple restorations were failing. A periapical radiopacity was associated with the apex of #45 and his mandibular bone was uncharacteristically radiolucent with interproximal and interradicular decrease in trabeculation with a stepladder appearance (Figure 2). The child and his mother were asked if there was any history of SCD or SC trait in the family, and they both reported that the patient has only the SC trait and not SCA. There were frequent lapses in his dental care, and when he did present, he was very active in the dental chair and salivated profusely. Oral hygiene regimens consisted of brushing twice a day with a manual toothbrush (poor brushing technique), no flossing, and the use of occasional mouthwash products (no

specific brand or type). Dietary habits consisted of no candy, regular meals with very low amounts of fruits and vegetables, and eating chicken, pork, and beef (grilled or fried), and a moderate consumption of fast foods.

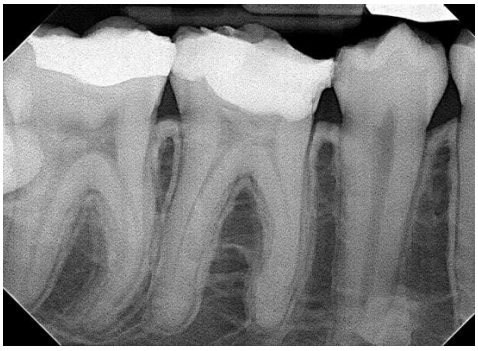

Figure 2. Image showing dense radiopacity on apex of #45 and interradicular "stepladder" appearance of alveolar bone #46

DISCUSSION

Many of the radiographic and oral characteristics displayed with this 13-year-old align with SCD. This patient exhibited many moderate carious lesions as well as hypercementosis, pulp stones, pulpal sclerosis, and the classic "stepladder" characteristic of the bone on the periapical shown, and these conditions are demonstrated in patients with higher levels of inflammation. Much of the patient's social and familial history leans toward limited access to oral care, and this can create a long-term negative effect on the health of the patient's oral cavity. This case stresses the importance of methodical oral assessments and medical history as well as investigating past oral health constraints and lack of care throughout the family unit. Early assessment, collaboration with all medical providers, and the primary focus on prevention are critical to minimizing compromises in the oral health status and overall health outcomes.

Some limitations within the study's design were that there was only one case evaluated, and results may not be generalizable to other patients known to have the SC trait. In addition, since this is one patient's clinical and dental history data, comparisons can only be made to other published case reports.

CONCLUSION

Early treatment and intervention with a pediatric dental patient are imperative to establish goals for care and to prevent oral infection and possible exacerbation of sickle cell diseases. Restorative management can be beneficial to prevent caries spreading to other areas of the mouth. Effective treatment modalities that include proper oral hygiene, early intervention and detection, and providing preventive products such as fluoride and calcium phosphate may help patients with SCD maintain their oral health as effectively as possible.

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