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Oral Pathology

“Sickle Cell Anemia”

Sickle cell anemia is a genetic blood disorder. It is defined as a condition of not having an

adequate amount of healthy red blood cells capable of carrying necessary oxygen to the body

tissues. Normal RBC’s are disc-shaped and look like doughnuts without holes in the center. The

'Sickle' reference is in relation to the disordered shape red blood cells take, having a crescent

shape instead of a disc shape. The three most common form of the disease in the United States

are: hemoglobin SS or sickle cell anemia, hemoglobin SC disease, and hemoglobin sickle beta-

thalassemia. Sickle cell anemia is caused by a mutation of a gene that is responsible for the

production of hemoglobin. The disease is acquired through hereditary genes. A child is affected

only if both parents pass on the defective gene. For example, one parent has normal hemoglobin

gene ( type AA) and the second parent has defective form gene ( Type AS, or the sickle cell

“trait”), there is a 50% chance that each child will have the sickle cell trait, but they will not have

sickle cell disease (type SS).

Clinical manifestations of the disease include increased blood viscosity and vascular

blockage caused by clumped sickled cells. The abnormal crescent shape of SCD makes them

sticky, rigid, and less able to freely travel in the blood stream. This shortage of  RBCs results in

less oxygen transport to the body tissues, fatigue, anemia, severe pain, tissue damage and in

extreme situations to stroke and even death. RBC’s live about 120 days in the blood stream and

then die. In sickle cell anemia the abnormal sickle cells usually die after only about 10 to 20

days. The bone marrow cannot make new blood cells fast enough to replace the dying defective

ones. There is a severe pain that the patient will experience due to sickle cell anemia, which is

the episodic pain in chest, abdomen, joints, and bone. Severe pain occurs when blockage of the

blood flow happens. In general, tissue damage, and infection occurs when there is a slower blood

flow occurring. Sickle cell anemia is not a contagious disease; it is a genetic disorder inherited

form parents. Factors that can precipitate episodes of sickle cell anemia are, changes in

temperature, hypoxia, dehydration, infection, physical exertion, and psychological stress.

The sickle cell gene is more common in people of Indian, Middle Eastern, Hispanic and

Mediterranean heritage as well as in African Americans.

Different methods are used to diagnose this disease. One simple common one is called

hemoglobin electrophoresis. Sickle cell anemia has no widely available cure, people are born

with it. However, treatments to improve the anemia and lower complications can help both

children and adults. Blood and marrow stem cell transplants, and bone marrow transplant is a

treatment that has been used, however finding an adequate donor is very difficult.  Treatment

includes regular medical check-ups, blood transfusion, and administration of supplemental

oxygen. Pain relieving medication is used during acute pain crisis. Sickle cell anemia varies from

person to person, however, with proper care and treatment, many people who have the disease

can have improved quality and reasonable health most of the time. One of the treatments for this

disease is called Hydroxyurea and Sulphasalazine which is actually a drug that decreases the

number of nucleotides inside cells, and reduces the concentration of defective hemoglobin and

also works by, reducing the number of "sticky" molecules on red blood cells in SCD. Because of

improved treatment and care, people who have sickle cell anemia are now living into their forties

or fifties, or longer.

As clinician, we see the manifestations that can vary from patient to patient. The oral

 mucosa may be pale or yellowish due to jaundice and most of the clinically detectable signs of

anemia can be observed. The increased production of red blood cells in the bone marrow

promotes a physical expansion in  the size of the marrow that ultimately results in bone loss.

Radiographically the ensuing striped and circumscribed bone pattern result in areas of decreased

bone density, as well as in areas of increased density and bone thickening. Radiographs can often

be diagnostic; bone marrow spaces are enlarged, trabeculate bone is present and prominent.

Abnormal bone patterns result from increased hematopoiesis occurring in the bone marrow

spaces. Radiographic analysis of mostly African Americans with SCD revealed increased

trabecular spacing,  prognathic maxillary profile, and class II malocclusion. Other conditions can

include the tendency towards osteomyelitis of the mandible, prolonged paresthesia of the

mandibular nerve, and asymptomatic pulpal necrosis, however the teeth and lamina dura are

unaffected. Clinical features of SCD show the disruption of bone development in the jaws in

response to the increased amount of red blood cells, expansion of bone marrow, and loss of bone

visible on dental radiographs. Chronic anemia associated with SCD can result in painful crises in

the jaws particular, the mandible, and it can mimic osteomyelitis both clinically and

radiographically. Oral treatment and clinical management for a SCD patient is root debridement

of 4 quadrants selective polishing for stain removal, and toothbrush removal of bacterial biofilm.

Refer the patient for a medical consultation and electrophoresis blood tests to determine the

presence of Hb S and a medical diagnosis and treatment protocol.

 It is essential that dentist and dental hygiene professionals be aware of the potential for clinical

complications that may affect the therapy of patients with SCD-trait, particularly if a patient is

scheduled for surgery, local anesthesia, nitrous oxide sedation, or general anesthesia, because

patient could go into a hypoxic shock which is where patient does not receive an adequate

oxygen supply during stress, which could cause serious complications. SCD is relevant to dental

practitioners, and hygienist because dental infections can trigger a crisis, and poor wound healing

is expected with this type of patients. Educating our patients is extremely important for the

individual, and should be the anchor for a consistent oral care provider relationship with the

patient.

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