Case Report: Sickle Cell Anemia Part 1

Name

Course

University

Date

Introduction

Sickle Cell Disease (SCD) is a hereditary disorder hindering the synthesis of hemoglobin - a fundamental component of the red blood cells (RBC) – and is a worldwide disease affecting millions of people, but the most sensitive part of the disease is that it has no cure; therefore, this case report has been authored to investigate the cause, symptoms, diagnosis, control and treatment of SCD with the express purpose of contributing to the extensive medical, pharmaceutical and scholarly efforts committed to making the world SCD-free. SCD is associated with aplastic, sequestration and vaso-occlusive crises exposing children to further medical complications and possible deaths. SCD normally shows in early childhood. For the first six months of life, babies are protected mostly by raised levels of HbF, but shortly after, the condition turns conspicuous (Peterson, 2008).

Symptoms of SCD

The commonest symptom of SCD is vaso-occlusive crisis. This crisis takes event when the microcirculation is blocked by sickled red blood cells (RBCs), prompting what hematologists call ischemic injury to the organ supplied and resultant pain. Pain complications are the most distinguishable clinical manifestations of SCD and are the largest cause of emergency room appointments and admissions for impacted patients. Pain crises start abruptly and can last for a couple of hours to a couple of days and end as sudden as it started. The pain has the ability to affect any body part. Some of the most affected parts are bones, soft tissue, the abdomen, and joints. In addition, the pain involves acute necrosis or avascular necrosis, or acute abdomen and dactylitis (paining and swollen feet and/or feet in children). With frequent incidents in the spleen, auto-splenectomy and infarctions leading to life-threatening infection are not uncommon.

In some extreme episodes, liver infarction and resultant liver failure take center stage. Sickle cell anemia is also a source of mental illness as observed with patient QC035 (Bloom, 2009).

Case Presentation

Reasons for Evaluation

QC035 is a 32-year old African American male with Nigerian ancestry. He was born to Nigerian father and a white American mother. They moved to Boston when he was 3 years old. Three weeks ago, he was in Nigeria to visit his paternal relatives. He was brought to the psychiatric outpatient department of *Paramount Medical Center (PMC),* Milton, in Boston MA with a third incident of mental sickness within a year. QC035 is an SCD patient. His hemoglobin genotype is Hb.SS. He was an unmarried Christian. Having spent most of his high school in hospital, QC035 went to college late and completed his degree in Environmental Science at 31 and is now an intern at a local NGO in (Boston). He was the only child to a Christian family. . His biological father succumbed to bone and spleen cancer which spread to vital organs such as the liver and gall bladder. He was only 9 at the time. His mother remarried four years later. He was presented by his mother and step-father to the psychiatric outpatient center on the ground of being too conversational, verbal and physically aggressive. His parents (mother and step-father) also reported abnormal sleep patterns and his refusal to eat his food, citing deliberate food poisoning. In addition to accusing his neighbor of being the reason his girlfriend left him for another man nine months ago, QC035 was expressing his desire to have a professional boxing match with Floyd Mayweather and promised to knock him down within two rounds. In general, the patient was wearing a mood of physical aggression, retribution and assigning blame (Al-Salem, 2015).

History of Psychiatric and General Medical Sickness

QC035 was well until 2 weeks before presentation when he was observed to be over-talking and more than often talked out of the subject being discussed. Small and simple things would provoke and agitate him and in most cases he would opt for a fight. For instance, he went to his neighbor's apartment and almost threw him out through the window from the third floor for being the reason his girlfriend abandoned him. In the real sense, the neighbor just moved in and has no idea who his girlfriend was. His thought of a ring fight against Floyd Mayweather Jr further confirmed the worries of his family. The manner in which he wanted to start fighting suggested that he perceived everyone around him very small and weak, including his step-father and mother. He has not been sleeping well for most parts of the night. When not in sleep, QC035 would be heard talking to himself, praying for long hours and reading the Bible. He turned down his meals prepared at home claiming that they have been poisoned somehow by his enemies, especially his neighbors.

QC035 did not manifest any depressive symptoms. However, in the past he has had suicidal thoughts in particular during the times of bone pain crises. He did not show any definitive plan to take his own life. The first incident of mental sickness happened three years ago, when he was 29. The second and third episodes took event within the past 13 months and that is why his family is showing concerns that his illness could only be worsening. The first episode involved inadequate sleep, aggressive behavior, visual hallucination, and blame of others for things happening to him. The second was characterized by all the aforementioned plus selective preference of meals, but the latest one is even worse because he is now refusing food. The last two cases lasted one week only thanks to good medical care from an anonymous private hospital. His medical records were shared with ***Paramount Medical Center.***

His first diagnosis of SCD was at the age of 6 during a bone pain crisis. He was blood-transfused two times; at ages 8 and 14 on accounts of hemolytic crises with PCV lower than 16% on each incident. PCV stands for packed cell volume. Except for recurrent incidents of bone pain crises happening thrice per annum, QC035 had been very stable after the last transfusion of blood.

Family, Development and Social History

QC035 was born as the only child and his biological father died from complications of bone and spleen cancer when he was 9 years old. The one who brought him to *Paramount Medical Center* is his step-father who raised him like his own. Interestingly, the thought of good memories about his biological father brings him happiness and calm especially during episodes. His relationship with his step-brother (his father step-father moved in with a child – 6 years older than QC035 - from his first marriage which ended in divorce) is amazing, except for when he is having episodes. He is working as a paralegal in a certain Boston law firm. In the house lives QC035, his mother, step-father and a maid.

QC035's Nigerian grandfather had SCD with Hemoglobin genotype (Hb.SS). His mother had no SCD, but his biological father had sickle cell trait with Hemoglobin genotype (Hb.AS). However, none of the family members showed signs of mental disease aside from QC035.

He spent most of his high school in hospital and finished his college late. Some of the hospital visits were not associated with SCD. He was described as a shy individual with a handful of friends. He never talked much, avoided group activities and most of his time was dedicated to reading.

Mental States Evaluation

Laboratory examination revealed the following. Physical evaluation showed a pale person with mild jaundice and moderate dehydration. Neurological check-up showed no gross abnormality. Other systems were examined and showed no medical normalcy. His PCV was 22%. Hematological examination revealed that total and differential WBC counts were within optimal range. His blood also showed reticulocytosis and one plus of malaria parasite – *Plasmodium falciparum* - Urea and electrolytes were normal. Lastly, Liver Function Test (LFT) did not show any abnormality (Bloom, 2009 and Peterson, 2008).

Diagnosis and Treatment

Bloom (2009) and Peterson (2008) studies are consistent with the fact that QC035 was diagnosed with paranoid schizophrenia (F-20) and co-morbid SCD (D-57) and malaria (B-50) in accordance with International Classification of Diseases and World Health Organization (W.H.O). He was treated with malarial drugs. He was also given a dose of Risperidone 2 mg daily to treat the symptoms of psychosis. In-patient medication of 15 days completely eliminated symptoms of psychosis. The patient was referred to the hematology department and was counselled to engage in regular malarial medication. After 3 months of hematological medication, QC035 became stable with no symptoms of psychosis (Al-Salem, 2015 & Ashley-Koch, 2000). In addition, he was advised to seek immediate medical examination the next time he comes back to America from Nigeria, should he feel the need to visit his relatives in the future. Nigeria is a tropic country prone to malaria.

References

Al-Salem, A. (2015). Medical and Surgical Complications of Sickle Cell Anemia. Springer.

Ashley-Koch, A, Yang, Q and Olney, R.S. (2000). Sickle Hemoglobin (*HbS*) Allele and Sickle Cell Disease: A HuGE Review. American Journal of Epidemiology. Retrieved http://aje.oxfordjournals.org/content/151/9/839

Bloom, M. (2009). Understanding Sickle Cell Disease. University Press of Mississippi.

Peterson, JM. (2008). Sickle Cell Anemia. The Rosen Publishing Group.