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### **Rare Case of Recurrent Hyperhemolysis Syndrome (HHS) and Methemoglobinemia in a Patient with Sickle Cell Anemia**

Ashbina Pokharel

Pradeep Khanal

Dilip Khanal

Samikshya Pandey

Bipin Ghimire

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Program: Beaumont Hospital – Royal Oak

Director: Sandor Shoichet, MD, FACP

Presenter: Ashbina Pokharel

Additional Authors: Pradeep Khanal, Dilip Khanal, Samikshya Pandey, Bipin Ghimire.

### **Rare Case of Recurrent Hyperhemolysis Syndrome (HHS) and Methemoglobinemia in a Patient with Sickle Cell Anemia**

Hyperhemolysis syndrome (HHS) is a rare but serious and potentially life-threatening complication of red blood cell (RBC) transfusion and has been described in both sickle cell disease and non-sickle cell disease patients. It is characterized by destruction of both donor and host RBC. Methemoglobinemia is another rare condition leading to cyanosis, tissue hypoxia and potentially death. We present a middle-aged female with sickle cell anemia who developed methemoglobinemia and hyperhemolysis after a surgical procedure.

41 yo female with sickle cell anemia (SC type) was hospitalized for elective right total hip resection arthroplasty. Patient received 3 units blood transfusion during the surgery. Post operatively patient developed cyanosis and was found to have methemoglobinemia. One week after surgery hemoglobin dropped precipitously from 8.4 gm/dl to 2.5 gm/dl within 24 hours. Lab investigation showed LDH >4500 U/L, haptoglobin <8 mg/dl, bilirubin 12.1 mg/dl, AST 2672 U/L. Coombs test was negative. Reticulocyte count was elevated 202 bil/L. Peripheral smear showed marked normocytic normochromic anemia with an absolute reticulocytosis and moderate anisopoikilocytosis including occasional spherocytosis. G6PD testing showed normal enzyme level. Methemoglobinemia resolved with use of methylene blue. Hyperhemolysis was treated initially with IVIG and Tocilizumab without improvement. Patient then underwent splenic embolization and plasmapheresis. She was also treated with Rituximab. Despite aggressive treatment patient continued to have hemolysis and developed multi organ failure. Ultimately family decided to pursue comfort care and patient subsequently died.

Hyperhemolysis syndrome is a rare transfusion related complication that can occur in patient with sickle cell disease. It can pose a significant therapeutic challenge and can unfortunately have devastating consequence including death.