G80 Hemoglobin SC Disease Presenting as Sickle Crisis After Outpatient Surgery: A Case Report

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After attending this presentation, attendees will learn the significant signs, symptoms, and sequelae of Hemoglobin SC disease, and implement the use of postmortem hemoglobin electrophoresis in indicated cases.

This presentation will impact the forensic community and/or humanity by providing information on the importance of recognizing an occult hemoglobinopathy and its possible contribution to the cause of death.

Hemoglobin SC (Hb SC) disease is a hemoglobinopathy with approximately the same incidence as that of Hemoglobin SS (Hb SS) disease in American blacks. The typical presentation of Hb SC disease includes fatigue, dyspnea on exertion, frequent upper respiratory infections, sporadic mild jaundice, and arthralgias. The onset of complications due to Hb SC disease is usually in childhood, but many do not present until the teens or later. Rare reports of autosplenectomy in Hb SC disease exist, but most patients with Hb SC disease have splenomegaly. Unlike Hb SS disease, painful crises in Hb SC disease occur more frequently in the muscles and joints than in the abdomen. Although Hb SC can present with the same manifestations as sickle cell anemia, it is generally characterized by a milder anemia and fewer vasoocclusive crises, with a severity that is intermediate to those of sickle cell disease or sickle cell trait.

A 33-year-old black male who underwent outpatient surgery for arthritis that developed following ankle and foot fractures sustained three years prior when he jumped off a roof while fleeing from an offender. His preoperative medical history and physical were within normal limits, and he had a history of sickle cell trait.

The surgery lasted 2½ hours, with two hours of tourniquet time, and consisted of a right subtalar fusion and arthroplasty of the 2nd-4th right toes, with application of a cast. There were no complications during surgery. Ketorolac and local bupivacaine were given, a dressing was applied, and the tourniquet was removed. His airway device was then removed, and on the way to the recovery room he suffered sudden cardiopulmonary arrest and was intubated, but expired despite resuscitative measures.

At autopsy, he had a plaster cast on the right leg, beneath which were intact sutured incisions and orthopedic hardware. The lungs were heavy and congested, and the mucosa of the gastrointestinal tract was hemorrhagic and appeared ischemic. The heart, liver, kidneys, and brain displayed no abnormalities. The spleen weighed 4 grams and had light tan, fibrotic parenchyma. Based on the appearance of the spleen, the family was contacted, and they stated that the patient did have sickle cell trait, and not sickle cell anemia.

Postmortem hemoglobin electrophoresis, however, revealed the presence of Hb SC disease. Microscopy revealed pulmonary congestion with prominent sickling of red blood cells within vessels. Sickling of red blood cells was also prominent in the liver, heart, kidneys, meninges, adrenal glands, thyroid gland, and gastrointestinal tract. Sections of the spleen showed prominent fibrosis and calcification, consistent with autosplenectomy. It is theorized that release of the tourniquet caused a large amount of partially deoxygenated blood to re-enter the circulation and cause a sickle crisis. The cause of death was determined as sickle cell crisis due to hemoglobin SC disease, with ankle surgery following a fall from height as a significant contributing condition. The manner of death was accidental.

Sickle cell crisis in Hb SC disease is rare. Despite the fact that this patient underwent autosplenectomy, he had previously suffered no symptoms of Hb SC disease and believed throughout his life that he had sickle cell trait. The use of postmortem hemoglobin electrophoresis was invaluable in the determination of cause of death in this case.

Forensic Science, Hemoglobin SC Disease, Sickle Cell Crisis

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