G14 Sudden Death of a Fourteen-Year-Old Female With Hb S-C Disease

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The goal of this presentation is to review the sudden death of a child with sickle cell anemia – Hb S-C type. The attendees will learn the different genetic forms of sickle cell disease, their complications, and the potential mechanisms of death with sickle cell anemia.

This will impact the forensic community and/or humanity by demonstrating an unusual sudden death in an adolescent with an element of past physical abuse.

According to her caretakers (mother and a grandmother), this 14-yearold black female child had been complaining of vague headache and back pains for several days. At the morning of her death she had complained of her "eyes turning color," increased headache "behind her eyes," and difficulty in "straightening out the fingers of her left hand." Her mother interpreted her daughter's complaints as predominantly an attempt to avoid school that day, and dismissed them from having any serious medical implications. At 1:00 p.m. that day, she was found lethargic and unresponsive. The mother and grandmother attempted to help her stand up, but failed. Of note, despite the grandmother's wish to immediately call for help, the mother rejected such initially, and the 911 call was not placed until some time later when the mother "could not feel a pulse." Paramedics arrived at 3:30 p.m., and she was pronounced dead after resuscitative efforts.

On initial external examination at the Medical Examiner's Office healed, patterned loop-type scars were noted on the deceased's torso, buttocks, and extremities. No acute injuries were present. Autopsy examination revealed a well-developed, well-nourished young adolescent female with scleral icterus and an overall slight jaundice appearance. Internal examination was remarkable for bilateral pulmonary edema, massive splenomegaly (spleen weight of 1,190 grams), and evidence of extreme anemia. No internal injuries were present. Microscopic examination was remarkable for extensive sequestering of sickled red blood cells within the spleen and a hypercellular bone marrow with areas of scarring. Postmortem toxicology was negative for alcohol or drug/medication substances other than a small quantity of acetaminophen. Vitreous electrolytes were unremarkable. Postmortem viral and bacterial cultures were negative, although a blood sample was positive on immunoassay for parvovirus B 19 antibodies IgM and IgG.

Sickle cell anemia is an autosomal recessive disease caused by a point mutation in beta hemoglobin gene chromosome 11p 15.4 (Hb S; 6 Glu leads to Val and Hb C; 6 Glu leads to Lys.). Approximately 8% of black Americans are heterozygous HbS. The carrier rate for HbC in is about 2% to 3%. HbC has a greater tendency to aggregate with HbS that does HbA, and hence those with HbS and HbC (designated HbSC) have a more severe disease than do whose with HbS and HbA. On deoxygenation, abnormal hemoglobins undergo aggregation and polymerization. This converts the hemoglobin from freely flowing liquid to viscous gel and results in distortion of the red cells, which acquire a sickle shape. Patients have to deal with problems ranging from severe anemia, vaso-occlusive complication, and chronic hyperbilirubinemia to severe infection. In children painful vaso-occlusive crises are extremely common, as well as hand-foot syndrome. An aplastic crisis represents a temporary cessation of bone marrow activity usually induced by parvovirus infection of erythroid cells. Sequestration crisis may occur in children with splenomegaly. With modern treatment approximately 90% of patients survive to the age of 20 years, and close to 50% survive beyond the fifth decade. No reported case of rapid death from Hg S-C type sickle cell anemia was found in the literature.

In this case the child had an acute infection with parvovirus B19, which was confirmed by blood serology. It is believed the cause of death was acute sequestration of blood with an aplastic crisis induced by the parvovirus, and thus ruled the death as natural. The mother of the deceased did not promptly call for medical help. She acted such in fearing the discovery of previous child abuse – which does suggest possible medical neglect and, thereby, a potential for other interpretations as to manner of death.

Sickle Cell Anemia, Hb SC Type, Parvovirus B19

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