H132 Sudden Death Due to Undiagnosed Rheumatic Heart Disease in a Child

Pauline Saint-Martin, MD, PhD, Service de Medecine Legale, Hopital Trousseau, CHRU Tours, Tours 37000, FRANCE; Camille Rerolle, MD, Service de Medecine Legale, Hopital Trousseau, CHRU Tours, Tours 37000, FRANCE; Maxime Faisant, MD, Service de Médecine Légale, Hôpital Trousseau, Tours, Centre 37044, FRANCE; and Thierry Lefrancq, MD, Le Vauban, BP 549, 16 rue Clerget, Nevers 58009, FRANCE*

After attending this presentation, attendees will better understand an unusual presentation of rheumatic heart disease which may lead to sudden death in young people and, in particular, the histology of heart lesions typically caused by this disease.

This presentation will impact the forensic science community by presenting an uncommon cause of sudden death in childhood and describing how forensic scientists should recognize the signs which can lead to this diagnosis.

Introduction: Sudden death in childhood is rare. The case of a young boy who died from complications of rheumatic heart disease with atypical presentation is presented.

Case report: A five-year-old Caucasian boy was pronounced dead at his home by paramedics. He had a recent medical history that had started one month prior to death with symptoms of gastroenteritis. Twelve days before death he suffered a painless, discrete cutaneous rash on the legs and one hand; scabies was suspected and he was treated accordingly. The following day there was fever and bilateral, diffuse thigh pain. Eight days before death he had trouble walking, handling a pen, and exhibited general psychomotor retardation. He was hospitalized for seven days for evaluation of the neurological symptoms. No explanation was found and all exams were normal, although there was an inflammatory syndrome and the brain Magnetic Resonance Imaging (MRI) showed what seemed to be signal abnormalities in the left centrum semiovale. All symptoms had disappeared during hospitalization and the boy was feeling well, so viral encephalitis was suspected and the boy was authorized to leave the hospital. He was scheduled to have an MRI six months later.

The boy died the day after he was discharged. An autopsy was requested. External examination of the body showed no injury. There were numerous small, crusted skin lesions on the legs. At autopsy, the organs were congested and there was pulmonary edema with enlargement of multiple lymph nodes. Toxicology tests were negative. Histological analysis revealed pancarditis associated with Aschoff bodies, aseptic mitral valve endocarditis, and myocarditis of the septum. The main neuropathological finding was a sub-acute cerebral infarction of the left centrum semiovale. It was concluded that the cause of death was an acute cardiac arrhythmia secondary to heart failure due to rheumatic heart disease. The boy had suffered undiagnosed rheumatic fever. The brain lesion was attributed to ischemic stroke resulting from an embolism secondary to the heart condition.

Discussion: Rheumatic heart disease is the most serious complication of rheumatic fever, a systemic disease that affects children with a previous Group A beta-hemolytic Streptococcus infection. Around 40% of patients with acute rheumatic fever develop some degree of pancarditis with associated heart failure. Acute rheumatic fever and rheumatic heart disease are thought to be due to an autoimmune response, but the exact pathogenesis remains unclear. The prevalence of rheumatic fever has decreased and is less than 0.05 per 1,000 population in industrialized countries, making it an uncommon cause of sudden death in childhood.

In this case, the issue of medical negligence could be raised, because the boy died the day after he had left the hospital with an inappropriately reassuring diagnosis; however, the presentation was atypical, with prominent neurological signs that were not specific to rheumatic fever and that were subsequently attributed to ischemic stroke. Following hospitalization and investigation, viral encephalitis was suspected which was consistent with the medical history. The diagnosis of rheumatic heart disease was made postmortem on the basis of histology. Although rheumatic fever is infrequent, clinicians should consider this diagnosis when confronted with neurological symptoms in a child with a previous history of fever and rash.

Forensic Pathology, Sudden Death, Rheumatic Heart Disease