G44 Myocarditis With Giant Cells in an Infant: A Case Report and Review of the Literature

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After attending this presentation, attendees will be able to recognize the various entities associated with myocarditis with giant cells, most notably idiopathic giant cell myocarditis, and its clinico-pathologic features.

This presentation will impact the forensic science community by highlighting a case of an uncommon disease entity which is commonly fatal, and generally affects young, healthy adults, but can also affect the pediatric population.

The subject was a 26-day-old, Asian female infant born at 31 weeks gestation with no complications at birth. While under the care of her parents, she vomited once and then was reported to be feeding poorly. She was taken to her pediatrician's office where she was "sick appearing." In the clinician's office she became unresponsive, was subsequently admitted to the nearest hospital, and died within four hours. Family history included a "head cold" in an older sibling and her mother was believed to suffer from an autoimmune-type disease which was undiagnosed.

At autopsy, the subject's growth parameters were between the 10th to 90th percentiles when corrected for prematurity, her skin was free of rashes, and her abdomen was distended. Within the abdominal cavity, there was 60 cc of ascites. The lungs were congested and heavy with a combined weight of 53 grams. The heart weighed 16 grams; it was normally formed, and had a probe patent ductus arteriosus. Externally, the epicardium of the heart was mottled pale tan to erythematous. Cut sections of the myocardium were equally mottled. The other major organs were appropriate weights and unremarkable for an infant of her age. No lymphadenopathy was identified. Blood cultures obtained from the hospital and at autopsy were negative. Toxicology and vitreous electrolytes were unremarkable.

Histological sections of the heart revealed patchy myocyte necrosis with mononuclear cells, a prominent collection of eosinophils, and ancient ischemia. scattered multinucleate giant cells. No granulomas were identified. The intramyocardial vessels and epicardial fat were free of inflammation. Histological sections of the other organs were free of granulomatous inflammation, viral cytopathic effect, or vasculitis.

Myocarditis with giant cells is seen in association with many recognized entities including tuberculosis, fungal infections, rheumatic myocarditis, measles, syphilis, foreign body reaction, Wegener's granulomatosis, hypersensitivity reaction, and sarcoidosis. Idiopathic giant cell myocarditis is as the name implies a myocarditis with giant cells, but of unknown etiology. It is a rare, but commonly fatal form of myocarditis which has been recognized since the beginning of the 20th century. This disease generally affects previously healthy, young adults (mean age 42 years); however, approximately 16 cases have been reported in the pediatric population. The youngest to date was 6-weeks- of-age; however, the majority of reported pediatric patients are teenagers. Symptoms generally are due to congestive heart failure, although numerous other symptoms have been reported including sudden death and palpitations. Diagnosis has classically been made at autopsy, although, the disease is being diagnosed by endomyocardial biopsy and following cardiac transplant. Gross identification of the disease ranges from "normal" to serpiginous areas of myocardial necrosis. Histology demonstrates myocyte necrosis, with lymphoplasmacytic inflammation with eosinophils and multinucleate giant cells. While the disease generally affects previously healthy people, approximately 20% of patients have immunologic disorders including inflammatory bowel disease, optic myocytis, thyroid disorders, systemic lupus erythematosus, Takayasu's arteritis, myasthenia gravis as well as others. The most successful treatment consists of cardiac transplantation with immunosuppression. Giant cell myocarditis has, however, been known to recur post-cardiac transplant at a rate of 20-25%. Without treatment, the average survival time from diagnosis to either death or cardiac transplantation is 5.5 months.

Based on the history including no known exposure to any drugs, maternal history of an autoimmune disease, and following review of the histology and other studies, the cause of death of this infant is due to idiopathic giant cell myocarditis. Based on the literature review, this is the youngest reported patient with the disease.

Myocarditis, Heart Failure, Sudden Death in Infants

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