Project Name: KDPath

Project Background & Significance:
Kawasaki Disease (KD) has outpaced rheumatic fever to become the leading cause of acquired heart disease in children in developed nations. It is marked by bilateral conjunctival injection (red eyes), cracked lips, strawberry tongue, red and swollen hands and feet, polymorphous rash, and enlargement of cervical lymph nodes. The feared complication of this systemic vasculitis is the development of coronary artery aneurysms, which occur in up to 25% of untreated children (Newburger et al. 2016). Physicians currently use clinical signs coupled with inflammation markers to diagnose KD and treat children with a high dose of aspirin and intravenous immunoglobulin. The unknown etiology makes the disease challenging to diagnose in some cases and hampers the development of better strategies to prevent damage to the coronary arteries. In a study of young adults < 40 years evaluated by cardiac catheterization for suspected cardiac ischemia, 5-7% of patients had lesions consistent with missed KD (Daniels 2012; Rizk et al. 2015). Better understanding of the pathology of the vasculitis will aid in identifying new therapeutic targets and contribute to understanding the natural history of this mysterious condition (Shimizu et al. 2015).

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Inclusion Criteria:
Case Definitions:

1) Definite case: Known history of KD in childhood
2) Probable case: Subject age <40 years with aneurysms of the coronary arteries associated with calcification and without known history of traditional risk factors for atherosclerosis

Cause of death: Any cause of death is of interest, particularly for definite cases with or without coronary artery aneurysms. ***Because there is no system in the U.S. to track KD cases, there is virtually no descriptive pathology of the cardiovascular system in individuals who had KD in childhood and died of unrelated causes***. The history of KD may or may not be available at the time of autopsy, but these cases would be extremely valuable. It is likely that the majority of cases will be young adults with silent aneurysms and myocardial infarction or arrhythmia as the cause of death.

Autopsy: We are seeking paraffin-embedded tissue from the coronary arteries and myocardium for immunohistochemical studies in our laboratory. Key tissues would be the myocardium (all chambers and septum) and coronary artery. Also of interest would be other extra-parenchymal muscular arteries (splanchnic, splenic, renal, etc.) and the ascending and descending aorta.

Consent: The KD Research Center will obtain next-of-kin information from the medical examiner and conduct informed consent for tissue donation. A completed consent form will be provided to the medical examiner.

IRB Approval: This protocol is approved by the UCSD Institutional Review Board (UCSD # 090156).

Resources for Family: Counseling about KD with sharing of lay informational material developed by our Center.

Resources for Forensic Pathologists: Histologic consultation with our team and with Dr. Kei Takahashi of Toho University in Japan, one of the world's leading pathologist for KD and a collaborator with our Center.

Website: https://www.kdpath.org/