Histiocytoid Cardiomyopathy associated with Wolff-Parkinson-White syndrome: A rare fatal combination in infancy

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Background: Histiocytoid cardiomyopathy (HC), initially described by Voth, is a rare genetic myocardial disease characterized by the presence of foamy histiocyte-like cells. It is a disease of infancy and has female predominance.

Case history: An asymptomatic 3 month old female, presented for a murmur evaluation. Examination was consistent with branch peripheral pulmonic stenosis. Family history was negative for congenital heart defects, cardiomyopathies, arrhythmias or sudden cardiac death. Echocardiogram was normal. EKG revealed pre-excitation consistent with Wolff-Parkinson-White (WPW) syndrome. The patient was followed up after 6 months at which time the EKG showed persistent pre-excitation. A 24 hour Holter monitor showed no arrhythmias but persistent pre-excitation at all heart rates. The patient was not on any medical treatment. Unfortunately, the patient died during sleep a few weeks later and autopsy was compatible with HC.

Discussion: Although sudden cardiac death has been reported to be associated with HC, the etiology is often unknown. Some theories suggest that HC is associated with abnormal conduction system development. There is a known association of HC with WPW. WPW, alone, is associated with sudden death. The ancillary finding of WPW in this patient raises the issue of a primary arrhythmia due to WPW causing the patient's demise and not the cardiomyopathy per se. Currently, there is no standardized treatment for HC and the management mainly focuses on using anti-arrhythmic drugs to prevent fatal arrhythmias. Although the patient's Holter was negative for arrhythmias, the persistent pre-excitation did place the patient at risk for fatal arrhythmias. Whether empiric beta-blocker therapy would have prevented this, is unknown.

Conclusion: Our patient presented with asymptomatic WPW and suffered sudden cardiac death with the diagnosis of HC made only on autopsy. This raises the question of the use of anti-arrhythmic therapy in infants at risk for sudden death associated with WPW.