Identical Anatomical Location of Accessory Pathway in a First-Degree Relative with Atrioventricular Reentrant Tachycardia

Khairul Mawaris

Introduction: Introduction Accessory conduction pathway has been thought to result from a cardiac developmental defect in electrical insulation between atria and the ventricles. Wolf-Parkinson-White (WPW) syndrome can cause paroxysmal supraventricular tachycardia. The prevalence is 0.1-0.3% in the general population and 3.4% in first-degree relatives. The location of accessory pathway (AP) could be anywhere along AV rings. During past 2 decades a significant heritable factor is increasingly recognized as well as familial WPW syndrome has also been described. This report presents a case of first degree relative with history of tachyarrhythmia and identical AP location as identified by electrophysiologic study (EPS).

Methods: Case 1 A 16-years-old female student (Patient A) was referred to our hospital for history of several palpitation episodes. The resting ECG showed pre-excitation. There were no history of other disease. Laboratory examination and echocardiography was normal. EPS identified eccentric activation at left side. Mapping via coronary sinus revealed AP at left lateral area of mitral ring. Application of multiple RFA during sinus eliminated pre-excitation. There were no inducible tachyarrhythmias nor retrograde conduction after procedure. Case 2 A 51-years-old female (Patient B, the mother of patient A) was referred to our hospital also for history of palpitation. There was no pre-excitation from resting ECG. Echocardiography showed mild LA dilatation with moderate mitral regurgitation. Mapping via coronary sinus identified the AP was at left lateral area of mitral ring. Several application of RF energy at this level during tachycardia eliminated retrograde conduction without any inducible arrhythmias.

Results: Discussion Accessory pathways are thought to result from developmental failure to eradicate the remnants of AV connections, other than specialized conduction system, during cardiogenesis. The PRKAG2 gene has been described to be associated with WPW syndrome in autosomal dominant pattern associated with heart disease. However, the genetic basis of isolated sporadic WPW syndrome and AP formation is still unknown. In this case, the daughter had a manifest left lateral Kent pathway and her mother had a unidirectional retrograde AP at the same position. Both patient have no evidence of associated cardiomyopathy. The exact mechanism resulting in the formation of AP is still unclear, it may be determined by mosaic somatic mutation or by environmental exposure.

Conclusion: Conclusion This report presents a case of first-degree relative presenting with AVRT. Electrophysiology study revealed a similar location of accessory pathway on left lateral mitral annulus. This finding supports a suggestion that a genetic factors may have a role in the patogenesis of formation of accessory pathway.