Familial Mahaim Syndrome

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We describe the occurrence of Mahaim syndrome in a mother and her son. The occurrence of such a rare disorder in two members of a family is noteworthy, has not been reported before, and suggests the possibility of genetic transmission. A genetic transmission of supraventricular tachycardia has been described only in rare cases for the Wolff-Parkinson-White syndrome. No such data is available for the Mahaim syndrome.

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CASE 1

A 23-year-old man presented with recurrent palpitations present since childhood age. There were no other medical problems. His physical exam was normal and one resting 12-lead ECG was within normal limits except for ST changes consistent with an early repolarization pattern at one time. Another ECG taken on a different day showed a short (and variable) PR interval and a left bundle branch block QRS pattern with R wave transition between lead V⁴ and V⁵ (Fig. 1). The palpitations were documented to be due to a regular tachycardia with a heart rate of 150–180 beats/min. The QRS complex during tachycardia showed left bundle branch block morphology, a normal frontal plane axis, and precordial R wave transition between lead V⁵ and V⁶ (Fig. 2). In an electrophysiological study, the clinical tachycardia was induced and was documented to be due to an antidromic AV re-entry tachycardia using a "Mahaim-type" atrioventricular accessory pathway. Intracardiac mapping localized this pathway to the right lateral tricuspid annular position. Mapping in this area revealed probable pathway potentials (Fig. 3). Radiofrequency energy catheter ablation at this site eliminated pathway conduction and the ability to induce tachycardia.

CASE 2

The patient's mother had a history of recurrent paroxysmal supraventricular tachycardia for many years. The tachycardia was reported as having left bundle branch block morphology. At the age of 43, she had an electrophysiological study performed, which confirmed the diagnosis of a "Mahaim-type" accessory pathway. Successful radiofrequency energy catheter ablation was performed at the mid right lateral right ventricular cavity, at sites where catheter manipulation resulted in transient mechanical pathway block (personal communication: Dr Pedro Brugada).

DISCUSSION

To the best of our knowledge, this is the first report of familial occurrence of the Mahaim syndrome. Due to the extremely low prevalence of this disease in the general population, we suspect a possible genetic transmission in this family.

Mahaim syndrome describes an antidromic atrioventricular re-entry tachycardia due to a slow, antegrade-only, conducting accessory pathway. During tachycardia the QRS complex has a left bundle branch morphology. The prevalence of this disease in the general population is unknown, but is believed to be extremely low. For comparison, the prevalence in the general population of the

Figure 1. Standard 12-lead ECG (25 mm/s, 10 mm/mV) showing sinus rhythm and left bundle branch block QRS pattern with R wave transition between lead V⁴ and V⁵, consistent with "Mahaim pathway" conduction. The short (and somewhat variable) PR interval is suspected to be due to automaticity originating from the Mahaim tract.

Figure 2. Standard 12-lead ECG (25 mm/s, 10 mm/mV) showing the clinical tachycardia with a regular rate (155 beats/min), left bundle branch block QRS pattern with precordial R wave transition between lead V⁵ and lead V⁶.

Figure 3. Four surface ECG leads (I, aVF, V¹, and V⁶) as well as five intracardiac electrogram leads recorded during sinus rhythm (100 mm/s). The ablation catheter (ABL D) is located at the lateral tricuspid annulus (subvalvular position). Mahaim pathway potentials are recorded in this region. Radiofrequency catheter ablation at this site eliminated Mahaim pathway conduction. (HRA = high right atrium, HBEP = HIS bundle electrogram proximal, HBE = HIS bundle electrogram, HBED = HIS bundle electrogram distal).

more common WPW syndrome is approximately 0.15%.¹ While initially understood as a "nodoven-tricular" accessory pathway, recent data from pa-tients undergoing curative radiofrequency catheter ablation has shown that the pathway usually con-nects the lateral right atrial wall to the apical right ventricular region.²

Most cases of SVT do not have a genetic cause or familial association. A familial occurrence of the WPW syndrome has been described. In the largest published series on this subject,³ the prevalence of pre-excitation in 2343 first-degree relatives of 383 patients with WPW syndrome was 0.55%. This was significantly higher than the 0.15% prevalence found in the general population.¹ The genetic trans-mission pattern was autosomal dominant with variable penetrance. The genetic locus of this iso-lated familial WPW syndrome is not known. WPW may also be associated with a familial form of

hypertrophic cardiomyopathy.⁴ Genetic linkage between these two conditions has been established in two families with this disorder. The gene locus was mapped to the chromosome 7q3.⁵ In rare cases WPW syndrome was associated with a mitochon-drial disorder (Leber hereditary optic neuropathy) suggesting an extranuclear maternal transmission pattern.⁶

Despite linkage to specific chromosomal regions in some cases of familial WPW syndrome, the un-derlying genetic defect and molecular mechanism that permits the accessory pathways to form is unknown. No other case reports or genetic data is available with regard to the Mahaim syndrome.

Analysis of the pattern of inheritance of cardiac arrhythmias and gene identification is important since it may allow genetic counseling in affected families. In addition, knowledge of the genetic transmission pattern may aid in understanding the

pathophysiological mechanism of arrhythmias as has been demonstrated in the long QT syndrome.⁷

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