

Case Report

Mahaim Fibre Coexisting with Anomalous Origin of the Left Main Coronary Artery From the Right Aortic Sinus and an Interarterial Course

CHARALAMPOS KRIATSELIS¹, KONRAD GOHL², JUERGEN JESSL², MARTIN GOTTWIK²,
ILIAS KARABINOS³

¹German Heart Institute, Berlin, ²Klinikum Nürnberg, Nuremberg, Germany;

³Cardiology Department, Euroclinic of Athens, Greece

Key words:

Mahaim fibre, coronary anomaly, broad QRS tachycardia, right coronary sinus, ablation.

We present the case of a 45-year-old female with a normal heart, who exhibited an episode of a broad QRS tachycardia that was initially suggested to be sustained ventricular tachycardia. Coronary angiography showed the left main stem originating from the right aortic sinus with an interarterial course between aorta and right ventricular outflow tract. Electrophysiological study revealed the presence of a right atriofascicular accessory pathway with decremental antegrade conduction (Mahaim bundle), which was successfully ablated.

Mahaim fibres are accessory fibres that exhibit antegrade conduction with decremental properties. Here we describe a case in which the coexistence of a Mahaim accessory pathway with a rare congenital coronary anomaly impeded the early diagnosis of a tachycardia.

Case presentation

A 45-year-old Caucasian woman presented at the emergency room with palpitations and dizziness. The 12-lead ECG showed a broad QRS complex tachycardia of 188 beats per minute with left axis deviation and left bundle branch block (LBBB) morphology (Figure 1A), which was terminated with administration of 50 mg ajmalin. During sinus rhythm, the 12-lead ECG showed a QRS complex of LBBB morphology and 125 ms duration and a normal PQ interval (150 ms) (Figure 1B). After an acute coronary syndrome was ruled out the patient was referred to the cardiology department for further evaluation. At this time the sus-

pected diagnosis was sustained ventricular tachycardia of unknown aetiology.

The patient had no history of palpitations, angina pectoris, dyspnoea, syncope or family history of sudden cardiac death. The echocardiogram showed normal left and right ventricles with no evidence of systolic or diastolic dysfunction. The heart valves were normal and an Ebstein's anomaly was ruled out. Coronary angiography revealed the left main stem originating from the right aortic sinus and having an interarterial course between the aorta and right ventricular outflow tract (Figure 2), with no evidence of compression. The right coronary artery was normal and also originated from the right aortic sinus. Left ventriculography revealed a left ventricle with normal systolic function. It appeared that the ventricular tachycardia could have been triggered by myocardial ischaemia, secondary to exercise-induced compression of the left main stem. Surprisingly, a ^{99m}Tc sestamibi exercise test showed no evidence of ischaemia or exercise-induced ventricular tachycardia.

Manuscript received:
February 16, 2007;
Accepted:
July 31, 2007.

Address:

Ilias K. Karabinos

17 Solomou St.
111 47 Galatsi,
Athens, Greece
e-mail:
iliaskk@in.gr

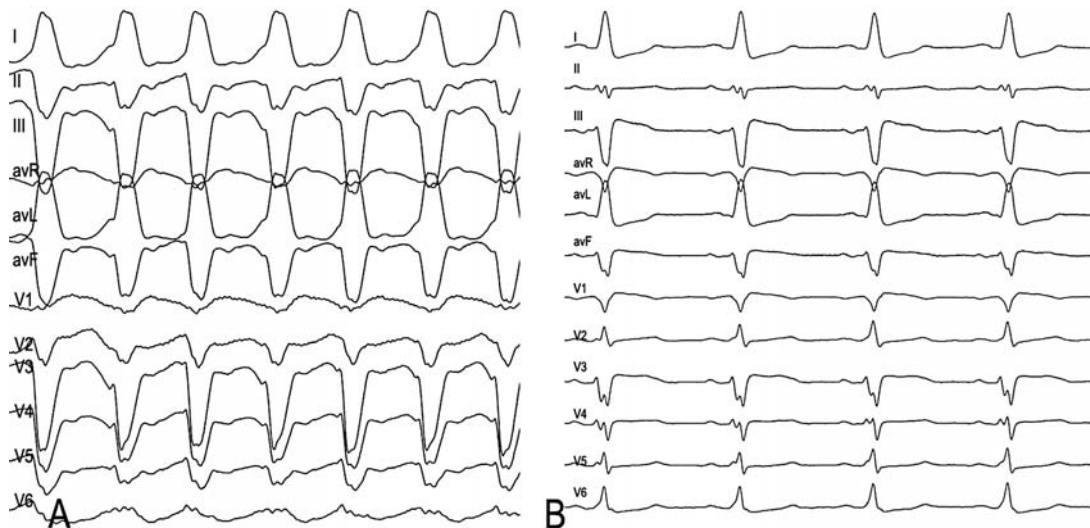


Figure 1. A: Twelve-lead ECG on admission showed a broad-QRS complex tachycardia of 188 beats per minute with left axis deviation and left bundle branch block morphology. B: Twelve-lead ECG after termination of tachycardia with 50 mg of ajmalin revealed a QRS complex of LBBB morphology and a normal PQ interval (150 ms).

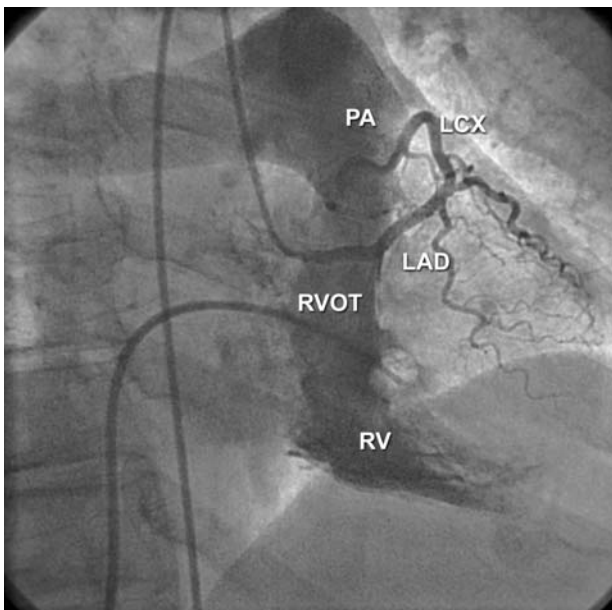


Figure 2. Simultaneous coronary angiogram (right anterior oblique projection) and right ventriculography revealed the left main stem originating from the right aortic sinus and having an interarterial course between aorta and right ventricular outflow tract. LAD – left anterior descending artery; LCX – left circumflex artery; PA – pulmonary artery; RV – right ventricle; RVOT – right ventricular outflow tract.

The patient was referred for electrophysiological study. The A-H interval was normal (105 ms) and the H-V interval slightly short (25 ms) (Figure 3A). Upon atrial stimulation, pre-excitation was generated with a QRS complex morphology identical to that of the clinical

tachycardia. The A-H interval was prolonged to 125 ms, whereas the H-V interval became negative (-8 ms) (Figure 3B), indicating the presence of a right atriofascicular accessory pathway with decremental antegrade conduction (Mahaim bundle). An antidromic atrioventricular re-entrant tachycardia (AVRT) was repeatedly induced by programmed ventricular stimulation with a ventricular extrasystole coupled at 320 ms (Figure 4). The cycle length and morphology of the antidromic AVRT were identical to that of the clinical tachycardia. Conventional mapping during sinus rhythm (Figure 5A) revealed that the accessory pathway was located at the superior-anterior tricuspid annulus. Radiofrequency (RF) energy was delivered at the suggested site of the accessory pathway for a total time of 30 s through an ablation catheter. Two and a half seconds after RF energy application, the pre-excitation disappeared (Figure 5B). The H-V interval returned to normal and tachycardia was no longer inducible. No early or late complications occurred. Thereafter, the patient remained asymptomatic for 18 months.

Discussion

Mahaim fibres have been classified according to their atrial and ventricular insertion as nodofascicular, atriofascicular and nodoventricular. The majority of Mahaim fibres are located along the tricuspid annulus, remote from the atrioventricular node.¹⁻⁴ Because of the decremental antegrade conduction, patients with Mahaim fibres have no or slight pre-excitation on the

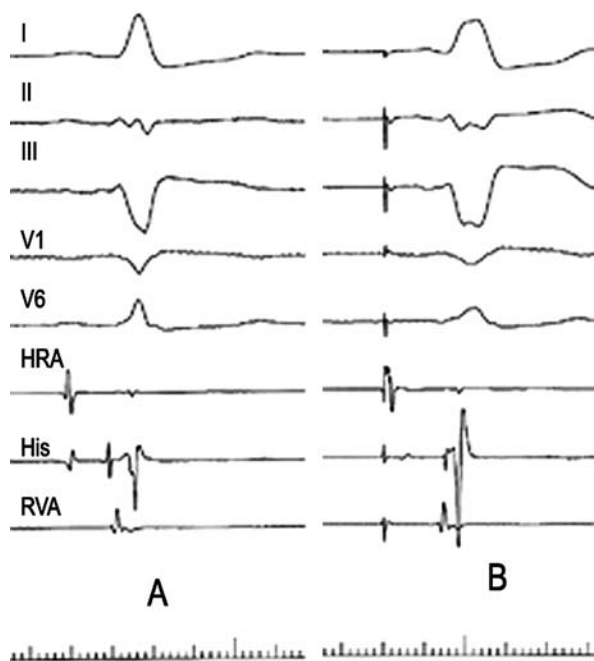


Figure 3. A: Electrophysiological traces. Normal A-H (105 ms) and slightly short H-V interval (25 ms). B: Upon atrial stimulation, pre-excitation was generated with a QRS complex morphology identical to that of the clinical tachycardia, with a prolonged A-H interval (125 ms) and a negative (-8 ms) H-V interval, indicating the presence of a right atriofascicular accessory pathway with decremental antegrade conduction (Mahaim bundle). His – recording from the His bundle; HRA – recording from the high right atrium; RVA – recording from the apex of the right ventricle.

ECG during sinus rhythm. From this point of view, patients with a Mahaim bundle constitute a distinct subgroup of the pre-excitation syndromes. They have

episodes of a pre-excited tachycardia, but during sinus rhythm they exhibit no or only slight ventricular pre-excitation. During electrophysiological study, this accessory pathway exhibits conduction only in the antegrade direction, with a long conduction time and decremental conduction properties. The pre-excited tachycardia (antidromic AVRT) is distinctive, with a left bundle branch block pattern, a long AV interval (due to the long conduction time over the accessory pathway), and a short VA interval. Most of these patients do not have episodes of narrow QRS complex tachycardia, (orthodromic AVRT), because of the absence of retrograde conduction in this accessory pathway.

There is a frequent association of Mahaim pathways with Ebstein's anomaly, as well as additional accessory pathways (Kent) and dual AV nodal pathway conduction.⁵ De Lemos et al reported a case of a male patient with Ebstein's anomaly, an anomalous origin of the right coronary artery arising from the left sinus of Valsalva, and a Mahaim fibre.⁶ In our female patient the Mahaim bundle coexisted with an anomalous origin of the left coronary artery (LCA) arising from the right sinus of Valsalva, in the absence of Ebstein's anomaly. To the best of our knowledge, a Mahaim fibre associated with anomalous origin of the left main coronary artery from the right aortic sinus has never been reported. Anomalous origin of the LCA occurs with an incidence of 0.017%.⁷ The anomalous LCA arising from the right sinus of Valsalva may take either a septal, an anterior, an interarterial or a posterior course. The passage of the LCA between the aorta and the right ventricular outflow tract

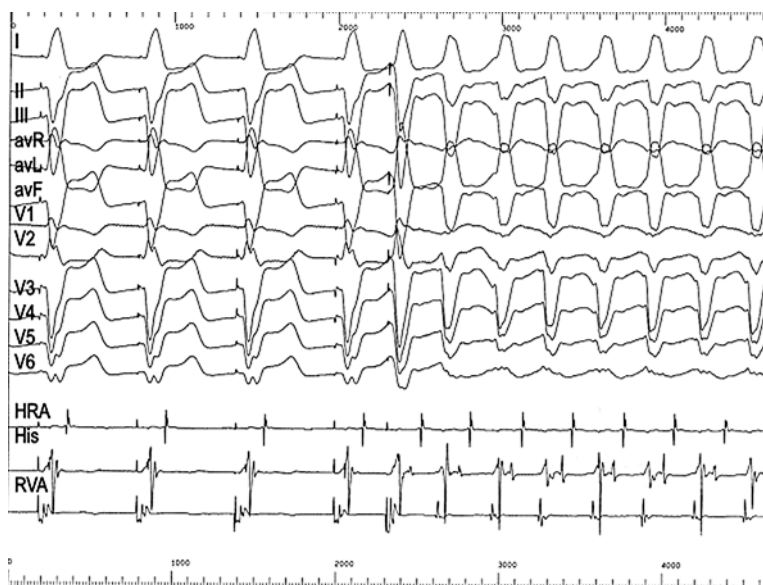


Figure 4. An antidromic atrioventricular (AV) re-entrant tachycardia was repeatedly induced by programmed ventricular stimulation with a ventricular extrasystole coupled at 320 ms. The cycle length and morphology of the tachycardia were identical to that of the clinical tachycardia. After ventricular stimulation retrograde conduction occurred over the His bundle and AV node (first 5 QRS complexes). Tachycardia starts after a premature ventricular stimulation (5th QRS complex) with antegrade conduction over the Mahaim bundle and retrograde conduction over the His bundle and AV node (antidromic atrioventricular re-entrant tachycardia). Abbreviations as in Figure 3.

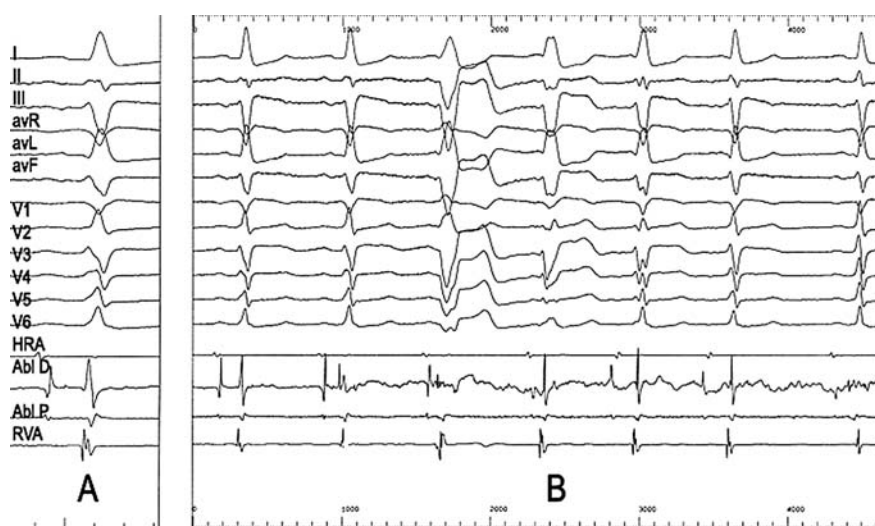


Figure 5. A: Conventional mapping during sinus rhythm revealed that the accessory pathway was located at the superior-anterior tricuspid annulus. The small electrogram between A and V in the Abl D recording is the Mahaim potential. B: Ablation of the Mahaim fibre. Radiofrequency energy delivery started at the end of the second QRS and the seventh QRS is the first without pre-excitation. Abl D – recording from the distal tip of the ablation catheter; Abl P – recording from the ablation catheter proximally; HRA – recording from high right atrium; RV – recording from the apex of the right ventricle.

has been associated with sudden death during or shortly after exercise in young persons.⁷⁻¹⁰ Sudden death is thought to result from transient flow impairment of the anomalous LCA caused by the increase in blood flow through the aorta and pulmonary artery that occurs during exercise.

An association between these two rare congenital anomalies cannot be established. Nevertheless, most of the patients with Mahaim pathways are young and do not usually undergo coronary angiography, so that a possible association of Mahaim pathways with coronary anomalies might have remained unrecognised. In the case we present, antidromic AVRT and ventricular pre-excitation were both misinterpreted as ventricular tachycardia and LBBB, respectively, which led to a thorough cardiac diagnostic workup, including coronary angiography. On electrophysiological study, the LBBB morphology of the QRS complex was attributed to pre-excitation over a Mahaim bundle and not to an organic heart disease. Successful elimination of the accessory pathway returned the 12-lead ECG to normal and tachycardia was no longer inducible.

References

1. Tchou P, Lehmann MH, Jazayeri M, et al: Atriofascicular connection or a nodofascicular Mahaim fiber? Electrophysiologic elucidation of the pathway and associated reentrant circuit. *Circulation* 1998; 77: 837-848.
2. Leitch J, Klein GJ, Yee R, et al: New concepts on nodoven-tricular accessory pathways. *J Cardiovasc Electrophysiol* 1990; 1: 220-230.
3. Benditt DG, Milstein S: Nodoven-tricular accessory connections: a misnomer or a structural functional spectrum. *J Cardiovasc Electrophysiol* 1990; 1: 231-237.
4. Klein GJ, Guiraudon GM, Kerr CR, et al: "Nodoven-tricular" accessory pathway: evidence for a distinct accessory atrioven-tricular pathway with atrioventricular node-like properties. *J Am Coll Cardiol* 1988; 11: 1035-1040.
5. Gatzoulis KA, Katsivas A, Apostolopoulos T, et al: Right posterior atrioventricular ring: a location for different types of atrioventricular accessory connections. *Interv Card Electrophysiol* 1999; 3: 187-191.
6. De Lemos JA, Delacretaz E, Piana RE, et al: Ebstein's anomaly associated with an anomalous coronary artery, 2 myo-cardial bridges, and a Mahaim fiber. *Images in Cardiovascular Medicine*. *Circulation* 1999; 100: 1757-1759.
7. Yamanaka O, Hobbs RE: Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990; 21: 28-40.
8. Cheitlin MD, Decastro DM, McAllister HA: Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva. *Circulation* 1974; 50: 780-787.
9. Kragel AH, Roberts WC: Anomalous origin of either the right or left main coronary artery from the aorta with subse-quent coursing between aorta and pulmonary trunk: Analysis of 32 necropsy cases. *Am J Cardiol* 1988; 62: 771-777.
10. Roberts WC: Major anomalies of coronary artery origin seen in adulthood. *Am Heart J* 1986; 111: 941-963.