Supraventricular Tachycardia in a Patient with Lown-Ganong-Levine Syndrome Associated with Apical Hypertrophic Cardiomyopathy

Motonobu Hayano, M.D., Yoichi Imamura, M.D., Mitsuhiro Tsuruta, M.D., Jun-ichi Inoue, M.D., Hiroshi Nakashima, M.D., Kuniaki Fukuyama, M.D., Yoshiki Eguchi, M.D., Shinsuke Tsuji, M.D., Shuzo Matsuo, M.D., and Katsu Yano, M.D.*

SUMMARY

Electrophysiologic study of a 55-year-old patient with Lown-Ganong-Levine syndrome associated with apical hypertrophic cardiomyopathy is reported. The patient had a history of recurrent attacks of tachyarrhythmia and his electrocardiogram showed a short P-R interval (0.10 sec) with narrow QRS complex and left ventricular hypertrophy with giant negative T waves. His cineangiogram showed severe apical hypertrophy. An electrophysiologic study was performed. The results of programmed atrial pacing show the existence of the dual A-V nodal pathways. The A-H interval at rapid atrial pacing increased maximally by 103 msec. Atrial stimulation could depolarize parts of the atrium without altering the supraventricular tachycardia. These findings suggested that preferential rapidly conducting A-V nodal and intranodal reentry are the responsible mechanisms in this reciprocating tachycardia.

We conclude that the short P-R interval was due to intranodal reentry through the dual A-V nodal pathways. To our knowledge, a case of Lown-Ganong-Levine syndrome with apical hypertrophic cardiomyopathy has not been previously described in the literature.

Additional Indexing Words:
Supraventricular tachycardia  Lown-Ganong-Levine syndrome  Dual A-V nodal pathways  Apical hypertrophic cardiomyopathy

One of the underlying mechanisms for supraventricular tachycardia in patients with Lown-Ganong-Levine syndrome is thought to be reentry utilizing the atrium, the A-V node and the A-V nodal bypass tract.1-3 It
has been also shown that intranodal reentry without utilization of the A-V nodal bypass tract can be responsible for the supraventricular tachycardia in this syndrome.4)-9)

This report describes a patient with Lown-Ganong-Levine syndrome associated with apical hypertrophic cardiomyopathy in whom supraventricular tachycardia is thought to be due to intranodal reentry associated with the dual A-V nodal pathways and rapidly conducting A-V nodal fibers.

CASE REPORT

A 55-year-old woman was admitted to the hospital in July, 1985 because of recurrent palpitations. On cardiac examination, an atrial sound without murmur was heard on auscultation. The chest X-ray showed no cardiomegaly. The electrocardiogram showed sinus bradyarrhythmia with a rate of 50 beats/min, a short P-R interval of 0.10 sec with a narrow QRS complex, and high amplitude and negative T waves in the precordial leads, indicating left ventricular hypertrophy (Fig. 1). Two-dimensional echocardiography showed severe apical hypertrophy of the left ventricle. The interventricular septum was 1.3 cm, and the left ventricular free wall in end-diastole was 1.1 cm. The

Fig. 1. Standard 12-lead electrocardiogram showing marked left ventricular hypertrophy and short P-R interval of 0.10 sec (see text).
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RA = right atrium; RV = right ventricle; PA = pulmonary artery; PCVP = pulmonary capillary venous pressure; LV = left ventricle; Ao = aorta.

Fig. 2. Left ventricular angiogram showing a typical spade-like shaped left ventricle at end-diastole in the right anterior oblique projection. top: end-systole, bottom: end-diastole frames.
most conspicuous finding was a severe thickening of the apical portion of the left ventricle. Systolic anterior movement of the mitral valve was not observed.

A cardiac catheterization study was performed on the 7th day after admission. Pressure studies are presented in Table I. There were no significant peak systolic pressure gradients in the left ventricle. Slightly elevated pressures were seen on the recordings from the pulmonary artery, right ventricle, left ventricle and aorta. The left ventricular and right ventricular end-diastolic pressures were slightly elevated. Left ventriculogram showed an end-diastolic spade-like configuration of the left ventricle indicating severe apical hypertrophy, and a vigorous symmetric contraction in systole was observed (Fig. 2). Coronary arteriogram was normal. On the basis of these findings, apical hypertrophic cardiomyopathy was diagnosed.

An electrophysiologic study was carried out with quadripolar catheters placed in the right atrial appendage, coronary sinus, the A-V junction and right ventricular apex. During sinus rhythm, A-H interval was 60 msec and H-V interval was 35 msec (Fig. 3). Rapid atrial pacing showed a progressive increase in the A-H interval at rates of up to 160 beats/min with 1:1 A-V conduction. The A-H interval at these pacing rates increased maximally by 103 msec, whereas no significant change in H-V interval was observed. The results of programmed atrial pacing shown in Fig. 4 indicate the existence of the dual A-V nodal pathways at a driven cycle length of 600 msec. At longer coupling intervals conduction proceeded over the fast pathway until a criti-
Fig. 4. The A-V conduction curve demonstrating dual A-V nodal conduction time. A1A2 response is abscissa and A2H2 response is ordinate. Open circles indicate response to atrial premature beats introduced at varying prematurity during basic cycle length (600 msec). Closed circles indicate atrial premature beats, resulting in echo beats.

Fig. 5. Supraventricular tachycardia is present at an A1A2 interval of 320 msec. A' shows retrograde atrial activation. Leads I, aVF, V1, high right atrial electrogram (HRA), coronary sinus (CS), His bundle electrogram (HBE) and right ventricular electrogram (RV) are recorded from top to bottom (see text).
Fig. 6. Intracardiac electrocardiograms are recorded at a paced rate of 70 beats/min of right ventricular pacing. $A'$ shows retrograde atrial activation. Leads $V_1$, $V_6$, $V_1$, high right atrial electrogram (HRA), coronary sinus (CS), His bundle electrogram (HBE) and right ventricular electrogram (RV) are recorded from top to bottom.

cal coupling interval was reached, whereupon refractoriness of this pathway resulted, causing conduction to shift over to the slow pathway. At an $A_1A_2$ interval of 380 msec, the corresponding $A_2H_2$ was 222 msec. When the $A_1A_2$ interval was 370 msec, the corresponding $A_2H_2$ suddenly increased to 513 msec. Atrial echoes were noted at $A_1A_2$ intervals of 370 msec or less. Supraventricular tachycardia was initiated at an $A_1A_2$ coupling interval of 320 msec (Fig. 5). Atrial stimulation could depolarize parts of the atrium without altering this supraventricular tachycardia. This finding indicates that the atrium is not necessary to sustain this tachycardia. As the coupling interval ($A_1A_2$) shortened, the corresponding $A_2H_2$ interval was prolonged progressively by 62 msec during the fast pathway conduction. Intracardiac electrocardiograms were also recorded during various rates of right ventricular pacing. Fig. 6 shows the recording of right ventricular pacing at a paced rate of 70 beats/min. With incremental stimulation from the right ventricular apex, 1:1 ventriculoatrial conduction was maintained, and the ventriculoatrial conduction time increased gradually up to a paced rate of 170 beats/min.

**Discussion**

A short P-R interval may reflect the abbreviation of A-V conduction time manifested by a short A-H interval in patients with Lown-Ganong-Levine syndrome. In patients with this syndrome a blunted response of
A-V nodal conduction to rapid atrial pacing has been demonstrated. Supraventricular tachycardia in this syndrome has been thought to be due to an atrionodal reentry involving the A-V nodal bypass tract (James fiber), the atrium and the A-V node. On the other hand, it has also been reported that an intranodal reentry through dual A-V nodal pathways can be responsible for the supraventricular tachycardia in this syndrome.81-10

The following electrophysiologic findings were observed in the present case: (1) a short A-H interval (60 msec) during sinus rhythm showing a maximal prolongation of 103 msec in response to rapid atrial pacing; (2) dual A-V nodal pathways with an increase of 62 msec in A-H interval during the fast pathway conduction; (3) supraventricular tachycardia initiated by atrial premature stimulation; (4) the atrium is not necessary for maintenance of supraventricular tachycardia.

These findings are similar to those previously described by Josephson et al8 who have suggested that preferential rapidly conducting A-V nodal fibers and intranodal reentry are the mechanisms responsible in some patients with Lown-Ganong-Levine syndrome and reciprocating tachycardia.

Thus, we concluded that in the present case the short P-R interval was not due to the anterograde A-V nodal bypass tract but due to intranodal reentry through the dual A-V nodal pathways. The present case showed the typical spade-like left ventricle on left ventriculography which was compatible with the diagnosis of the apical type of hypertrophic cardiomyopathy.11

The association of WPW syndrome in patients with hypertrophic cardiomyopathy has been reported.12-14 In a review of published cases, Marriott(12) suggested that this association was seen with some frequency in familial cardiomegaly (12%), and to a lesser extent in idiopathic cardiomegaly (1.5%). Perosio et al(14) reported 105 patients with preexcitation syndrome. In their cases, 8 (7.6%) had an associated hypertrophic cardiomyopathy, and 1 of these 8 patients had Lown-Ganong-Levine syndrome. However, the ECG of this case showed flat T waves in leads I, II, aVL, V5 and V6.

Although cardiac catheterization and cineangiography data from this case were not shown in their paper, the apical type of hypertrophic cardiomyopathy was not likely because of the absence of typical giant negative T waves on the ECG. To the best of our knowledge an association of Lown-Ganong-Levine syndrome and "apical" hypertrophic cardiomyopathy as shown in the present case has not been described in the literature.

References

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