Japanese Circulation Journal (1998.05) 62巻6号:458～461.

A Case of Cardiac Sarcoidosis —Significance of Ventricular Tachycardia Originating From the Septum— (心サルコイド症の1例—中隔起源心室頻拍の意義—)

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Jpn Circ J 1998; **62:** 458–461

A Case of Cardiac Sarcoidosis
—— Significance of Ventricular Tachycardia Originating From the Septum ——

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A 65-year-old woman was admitted for assessment of recurrent tachycardia. Cross-sectional echocardiography showed that the anterobasal portion of the ventricular septum was thin and dyskinetic. An electrophysiologic study revealed ventricular tachycardia, during which marked fragmented potentials could be obtained from the anterior septal aspect of the right ventricle. The site of earliest activation was in the vicinity of the His bundle. A diagnosis of cardiac sarcoidosis was made by based on endomyocardial biopsy combined with the clinical manifestations. Ventricular tachycardia originating from the anterior septum may be an indicator of underlying cardiac sarcoidosis. (*Jpn Circ J* 1998; **62:** 458–461)

Key Words: Cardiac sarcoidosis; Ventricular tachycardia; Electrophysiologic study; Echocardiography; Fragmented potentials

Sarcoidosis is a granulomatous disease of unknown cause that involves multiple organ systems, including the heart. Associated ventricular tachycardia (VT) is the leading cause of death in cardiac sarcoidosis[1,2] Although several previous reports have described the occurrence of cardiac sarcoidosis, its inducibility by programmed electrical stimulation, and its treatment[3,4] there has been little study of the site of the VT substrate, which can be determined by electrophysiologic mapping[5] We describe here a patient with VT associated with cardiac sarcoidosis in whom detailed electrophysiologic study, including right endocardial catheter mapping, was performed. In this case, the morphology of the electrocardiogram (ECG) and the site of origin of the VT reflected the location of focal dyskinesis.

Case Report

A 65-year-old woman was admitted to the Takikawa Municipal Hospital with severe palpitations and lightheadedness. Seven years previously a diagnosis of sarcoidosis had been made on account of bilateral hilar lymphadenopathy and characteristic chorioretinitis with secondary glaucoma. On presentation at the emergency room, the patient was found to have a wide QRS complex tachycardia with a heart rate of 174 beats/min. In leads II and V_4 of the tachycardic ECG (Fig 1), the final portion of the QRS complex exhibited negative notches (▲), giving the appearance of retrograde P waves. Intravenous procainamide and disopyramide were ineffective in terminating the tachycardia. The cardiac rhythm was converted to sinus rhythm by electrical cardioversion. The 12-lead surface ECG during the sinus rhythm (Fig 1) showed a frontal axis of +10°, right bundle branch block morphology, and a QRS duration of 150 msec, mimicking the documented tachycardia. These findings misled the attending physician into making a diagnosis of paroxysmal supraventricular tachycardia. The patient was transferred to our section in order to perform a detailed diagnosis, assess total cardiac function, and control the recurrent tachycardia.

Laboratory data on admission, including the serum angiotensin-converting enzyme level, were completely normal. Chest radiography exhibited characteristic bilateral hilar lymphadenopathy. Gallium scintigraphy revealed abnormal uptakes corresponding to the hila. Cross-sectional echocardiographic images (Fig 2) showed that the anterior basal portion of the ventricular septum was thin and dyskinetic, whereas the other parts were of normal thickness and only slightly hypokinetic. Left ventriculography in the left anterior oblique projection revealed poor contraction of the basal lateral portion, which was not clearly detected by echocardiography, in addition to the focal dyskinesis of the basal septal portion.

An electrophysiologic study was performed in the absence of antiarrhythmic drug therapy. Sinus and atrioventricular nodal function were normal. The interval between the His-bundle electrogram and the ventricular electrogram was prolonged by up to 100 msec and the ventricular electrogram was widely split. Tachycardia was reproducibly induced with double extrastimuli from the right ventricular apex. The morphology of the 12-lead surface ECG during the induced tachycardia completely coincided with the clinical tachycardia (Fig 1) and the intracardiac electrocardiogram revealed atrioventricular (AV) dissociation (Fig 3), thereby proving that this tachycardia was VT. A decapolar catheter to be used for endocardial mapping was then inserted via the inferior vena cava and advanced toward the right basal anterior

(Received September 19, 1997; revised manuscript received December 15, 1997; accepted December 16, 1997)
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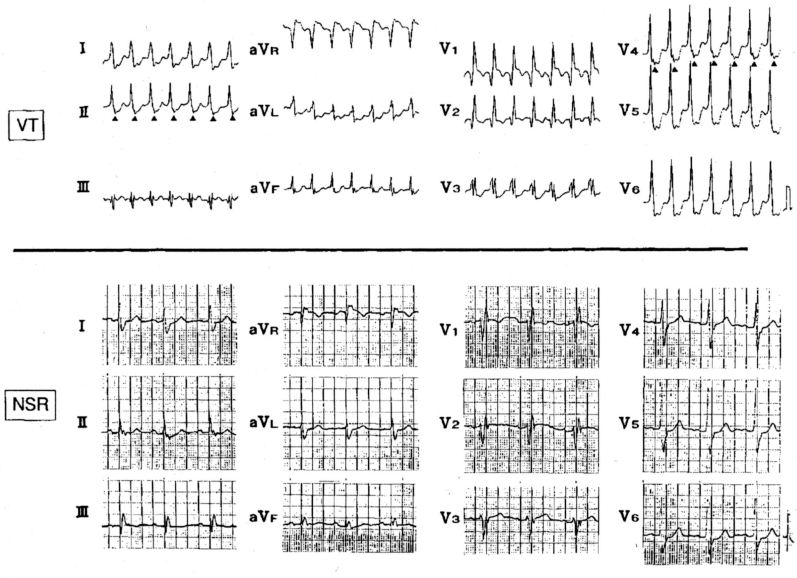

Fig 1. Twelve-lead ECG recordings. Top: ECG recorded in the emergency room showing a wide QRS complex tachycardia. Note the negative notches in the final portion of the QRS complex in leads II and V_4 (▲). Bottom: ECG during sinus rhythm in which the QRS morphology mimicked that of the tachycardia. VT, ventricular tachycardia; NSR, normal sinus rhythm.

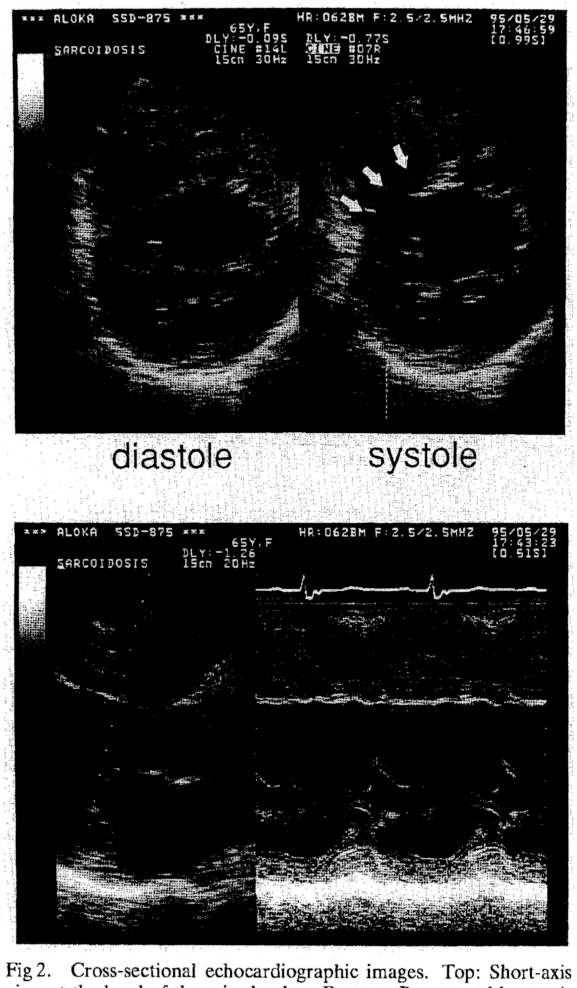

Fig 2. Cross-sectional echocardiographic images. Top: Short-axis view at the level of the mitral valve. Bottom: Parasternal long-axis view and M mode view at the level of the mitral valve. Note that the anterior basal portion of the interventricular septum was thin and dyskinetic (white arrows).

septum, 2–3 cm distal to the electrode for recording the His-bundle electrogram (Fig 4). During the VT marked diastolic fragmented potentials, occupying 62% of the total cycle length, were obtained from the multiple bipolar recordings from this catheter (Fig 5). The site of earliest activation in this mapping was at electrodes 5–6, at which the onset of the potential (thin arrow) preceded the onset of the surface ECG (thick arrow) by 75 msec. The timing of the potentials recorded from the right ventricular inflow tract, apex, and outflow tract were later than the onset of surface ECG by 50, 50, and 40 msec, respectively, and no abnormal diastolic potentials were seen in these areas. In the right ventricular endomyocardial biopsy, findings of interstitial fibrosis and myocardial cellular hypertrophy were demonstrated, although characteristic non-caseating epithelial cell granulomata were not identified. Fatty degeneration was not observed. From these histopathologic findings combined with the above-mentioned clinical manifestations, a diagnosis of cardiac sarcoidosis was made.

Discussion

Left ventricular wall motion abnormalities of various degrees are a common manifestation of cardiac sarcoidosis[6,7] Focal asynergy in the basal portion of the interventricular septum in particular is postulated to be a typical finding of this disease[6] The site of origin of the VT must be closely related to the localization of the wall motion abnormalities in cardiac sarcoidosis, as well as in myocardial infarction or other myocardial disorders, because both the substrates of the VT and asynergy may be associated with myocardial granulomas or other pathologic changes[3,6] Jain et al[5] reported on a surgical case of VT related to sarcoidosis showing the area of earliest activation to be at the apical margin of the inferior left ventricular aneurysm.

Other evidence, such as the late potential[8] or inducibility by the programmed extrastimulus[3,8] supports the hypothesis that the mechanism of the VT related to sarcoidosis is re-entry. In our case, clinical VT was induced and detailed endocardial mapping of the VT was per-

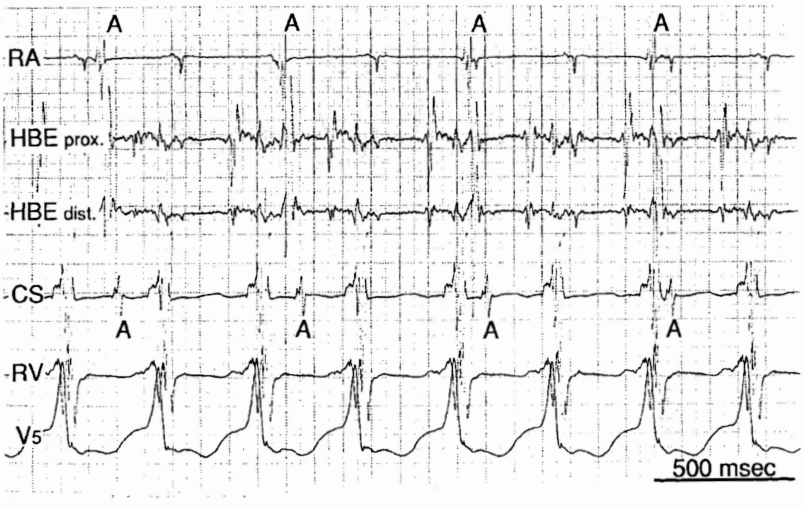

Fig 3. Electrograms of the tachycardia showing AV dissociation. RA, right atrial potential; HBE, His-bundle electrogram; prox., proximal; dist., distal; CS, coronary sinus; RV, right ventricle; A, atrial depolarization. In the HBE prox. and HBE dist. recordings, ventricular potentials were fragmented and therefore His potential was unclear.

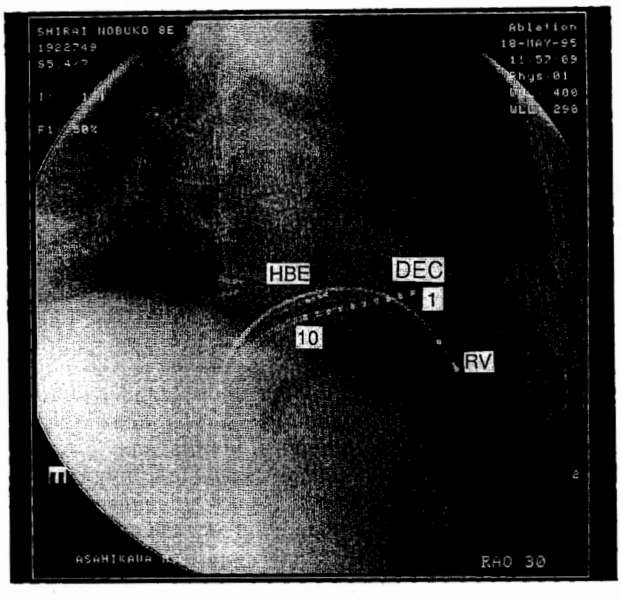

Fig 4. Catheter positioning in the 30° right anterior oblique view for recording the intracardiac potentials shown in Fig 5. A decapolar catheter (DEC), whose electrodes were numbered in order from the tip, was placed at the right basal anterior septum. RV and HBE, catheters for recording the right ventricular and His-bundle electrograms, respectively.

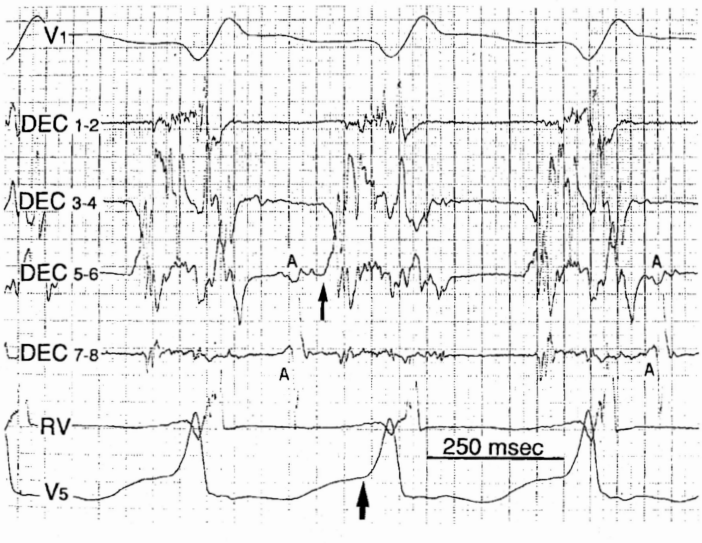

Fig 5. Electrograms of the tachycardia. Note a diastolic fragmented bipolar potential (thin arrow) recorded from the decapolar catheter (DEC). The earliest diastolic potential was seen at the pair of the 5th and the 6th electrodes (5–6). RV, right ventricle; A, atrial potential.

formed during an electrophysiologic study. Abnormal diastolic fragmented activation, indicating a slow conduction zone of a re-entrant tachycardia, was recorded in the anterior basal septum, where focal dyskinesis was detected by echocardiography and left ventriculography. The site of earliest activation in our mapping was also located in this area, near the electrode for His-bundle electrogram recording. Kaltenbrunner et al[9] analyzed the intraoperative maps for postinfarctional VT and reported that VTs showing an epicardial breakthrough on the right ventricle originated from the interventricular septum. This activation pattern is similar to that for normal sinus depolarization[10] thereby indicating participation of the His-Purkinje system in the VT process. In our case, however, the depth of the site of origin of the VT is unclear because left ventricular endocardial mapping was not performed. There is a possibility that there is an earlier activation site in the left septal site.

In summary, the septal localization of the VT substrate in our case may account for the similarity in the ECG morphology between the VT and the sinus rhythm. When considering that a focal lesion in the basal septum is specific for cardiac sarcoidosis[6] it is proposed that a VT originating from the septum and therefore showing a similar ECG morphology to the sinus rhythm should be a manifestation suggesting sarcoidosis as an underlying disorder.

References

1. Roberts W, McAllister H, Ferrans V: Sarcoidosis of the heart: a clinicopathologic study of 35 necropsy patients (group I) and review of 78 previously necropsy patients (group II). *Am J Med* 1977; **63:** 86–108
2. Silverman K, Hutchins G, Bulkley B: Cardiac sarcoid: a clinicopathological study of 84 unselected patients with systemic sarcoidosis. *Circulation* 1978; **58:** 1204–1211
3. Huang PL, Brooks R, Carpenter C, Garan H: Antiarrhythmic therapy guided by programmed electrical stimulation in cardiac sarcoidosis with ventricular tachycardia. *Am Heart J* 1991; **121:** 599–601
4. Winters SL, Cohen M, Greenberg S, Stein B, Curwin J, Pe E, et al: Sustained ventricular tachycardia associated with sarcoidosis: Assessment of the underlying cardiac anatomy and the prospective utility of programmed ventricular stimulation, drug therapy and an implantable antitachycardia device. *J Am Coll Cardiol* 1991; **18:** 937–943
5. Jain A, Starek PJK, Delany DL: Ventricular tachycardia and ventricular aneurysm due to unrecognized sarcoidosis. *Clin Cardiol* 1990; **13:** 738–740
6. Valantine H, McKenna WJ, Nihoyannopoulos P, Mitchell A, Foale RA, Davies MJ, et al: Sarcoidosis: a pattern of clinical and morphological presentation. *Br Heart J* 1987; **57:** 256–263
7. Burstow DJ, Tajik AJ, Bailey KR, DeRemee RA, Taliercio CP: Two-dimensional echocardiographic findings in systemic sarcoidosis. *Am J Cardiol* 1989; **63:** 478–482
8. Kasanuki H, Isogai K, Hosoda S: Detection and significance of late potential in patients with cardiac sarcoidosis. *Nippon Rinsyo-Jpn J Clin Med* 1995; **53:** 463–468 (in Japanese)
9. Kaltenbrunner W, Cardinal R, Dubuc M, Shenasa M, Nadeau R, Tremblay G, et al: Epicardial and endocardial mapping of ventricular tachycardia in patients with myocardial infarction. Is the origin of the tachycardia always subendocardially localized? *Circulation* 1991; **84:** 1058–1071
10. Dürrer D, van Dam RT, Freud GE, Janse MJ, Meijler FL, Arzbaecher RC: Total excitation of the isolated human heart. *Circulation* 1970; **41:** 899–912