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Cardiovascular Imaging In-a-Month

•A 68-Year-Old Man With Complete Atrioventricular Block and Congestive Heart Failure

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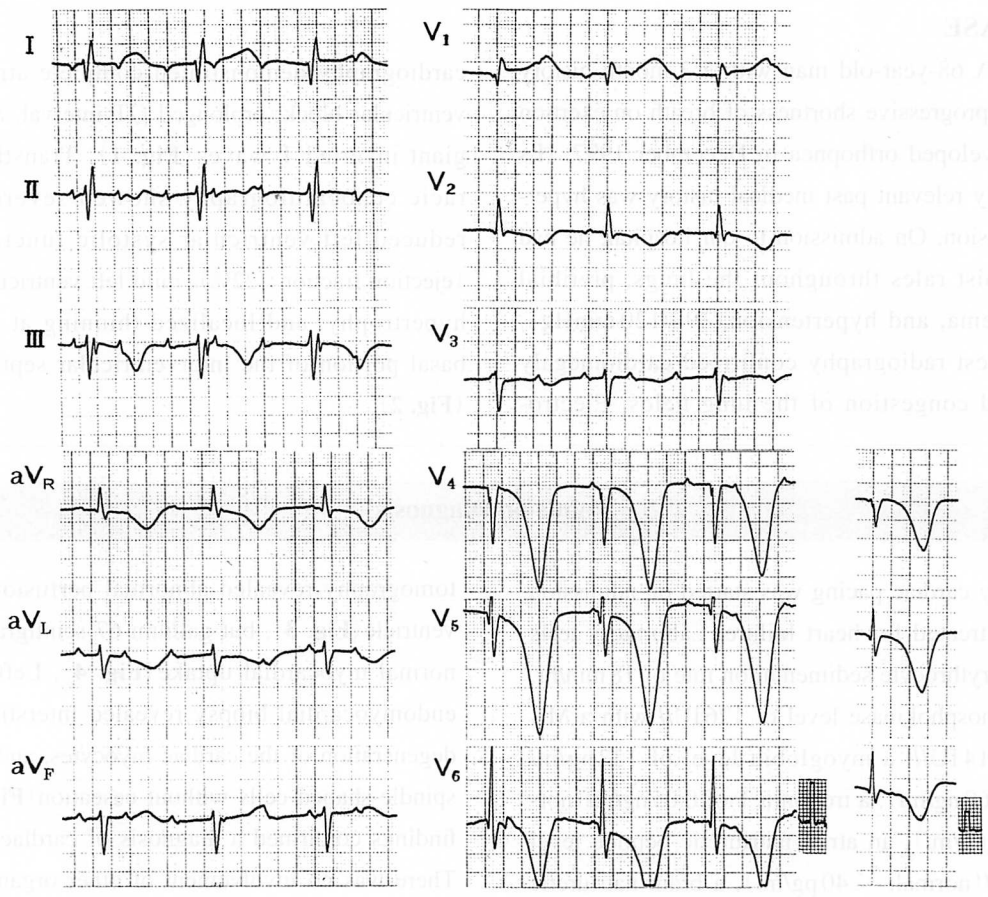

Fig. 1

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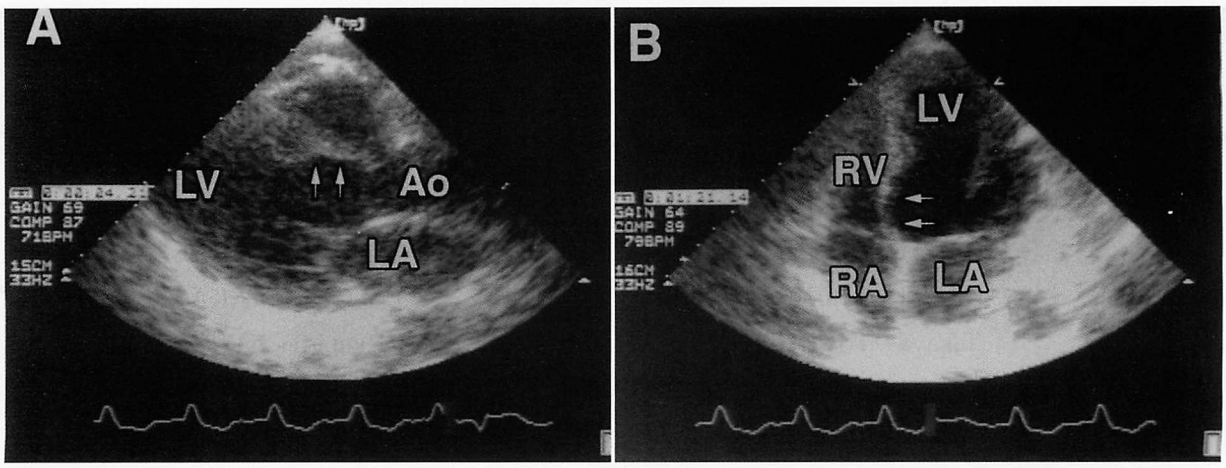

Fig. 2

CASE

A 68-year-old man with a 3-month history of progressive shortness of breath on exertion developed orthopnea in December 1997. His only relevant past medical history was hypertension. On admission to our hospital, he had moist rales throughout the lungs, pretibial edema, and hypertension (198/120 mmHg). Chest radiography confirmed cardiomegaly and congestion of the lung fields. Electrocardiography demonstrated complete atrioventricular block, prolonged QT interval, and giant inverted T waves (**Fig. 1**). Transthoracic echocardiography showed severely reduced left ventricular systolic function (ejection fraction: 22%), mild left ventricular hypertrophy, and localized thinning at the basal portion of the interventricular septum (**Fig. 2**).

Points on Diagnosis

Temporary cardiac pacing was started on admission and he was treated for heart failure. Laboratory tests showed an erythrocyte sedimentation rate of 18 mm/hr, a creatine phosphokinase level of 126 IU/*l* with a MB fraction of 14 IU/*l*, a myoglobin level of 132 ng/m*l* (normal, $<$ 60 ng/m*l*), a troponin T of 0.05 ng/m*l* (normal, $<$ 0.25 ng/m*l*), an atrial natriuretic peptide level of 150 pg/m*l* (normal, $<$ 40 pg/m*l*), a brain natriuretic peptide level of 1,870 pg/m*l* (normal, $<$ 20 pg/m*l*), an angiotensin converting enzyme level of 24.2 IU/*l* (normal, 8.3−21.4 IU/*l*), and a lysozyme level of 18.4 μg/ml (normal, 5.0−10.2 μg/ml). Coronary angiography was normal. Technetium-99 m methoxy-isobutyl isonitrile and iodine-123 betamethyl-*p*-iodophenyl-pentadecanoic acid dual single photon emission computed tomography revealed abnormal perfusion of the left ventricle (**Fig. 3**), but gallium-67 scintigraphy showed normal myocardial uptake (**Fig. 4**). Left ventricular endomyocardial biopsy revealed interstitial fibrosis, degeneration of the cardiac myocytes, and a cluster of spindle-shaped cells without caseation (**Fig. 5**). These findings confirmed a diagnosis of cardiac sarcoidosis. There was no involvement of other organs. A permanent pacemaker was implanted because of recurrent ventricular tachycardia and the prolonged interval between the His bundle and ventricle on electrograms. The patient also received corticosteroid therapy after pacemaker implantation. The left ventricular ejection fraction improved from 22% to 40% and the cyclic variation of myocardial integrated backscatter, which

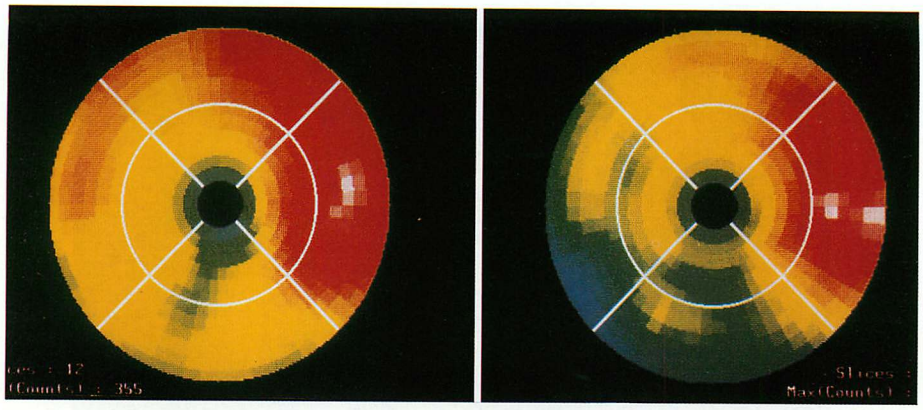

Fig. 3

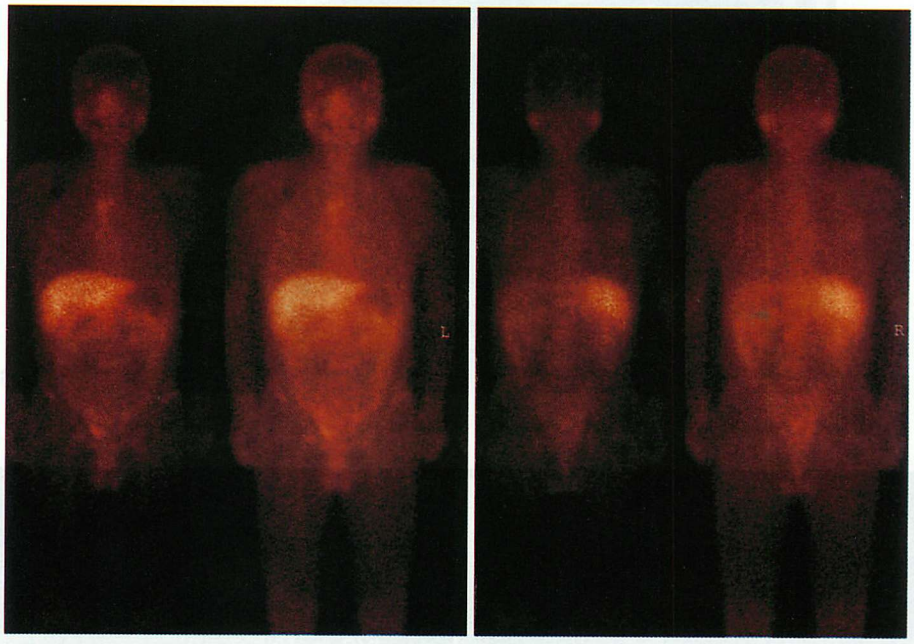

Fig. 4

reflects intramyocardial contractile function[1], reappeared 1 month after the start of steroid therapy (**Fig. 6**).

In our experience, it is difficult to detect cardiac involvement at an early stage in patients with sarcoidosis. Valantine *et al.*[2] studied various morphological abnormalities and found that focal thinning of the anterior basal portion of the interventricular septum was specific to cardiac sarcoidosis. Echocardiographic demonstration of this abnormality in a patient with or without complete atrioventricular block is indicative of cardiac sarcoidosis.

Diagnosis: Cardiac sarcoidosis

Acknowledgments

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References

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2) Valantine H, McKenna WJ, Nihoyannopoulos P, Mitchell A, Foale RA, Davies MJ, Oakley CM: Sarcoidosis: A pattern of clinical and morphological presentation. Br Heart J 1987; **57**: 256–263

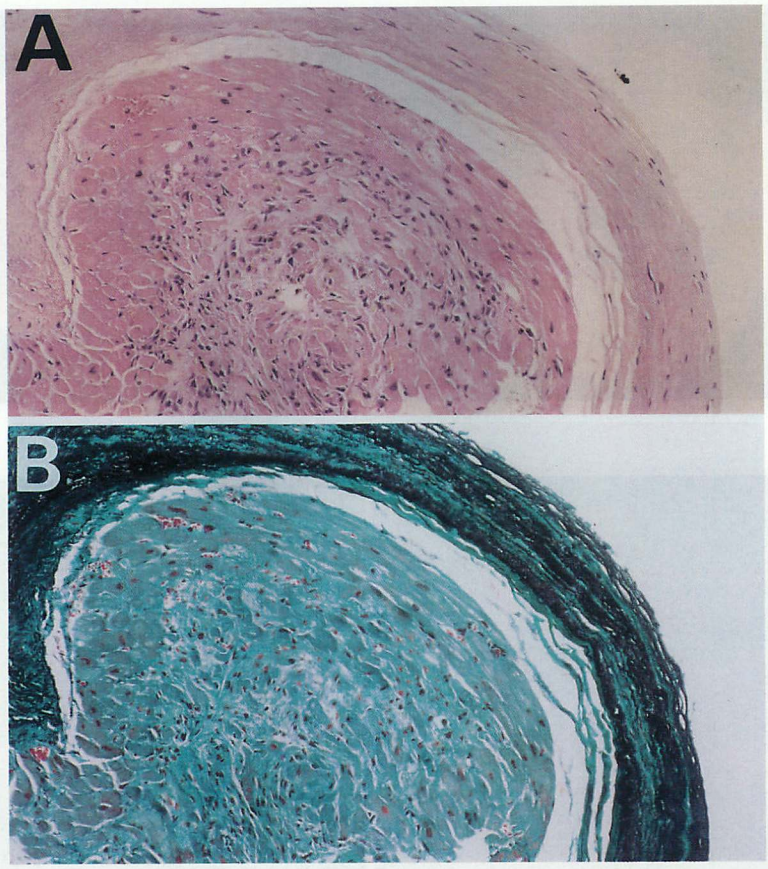

Fig. 5

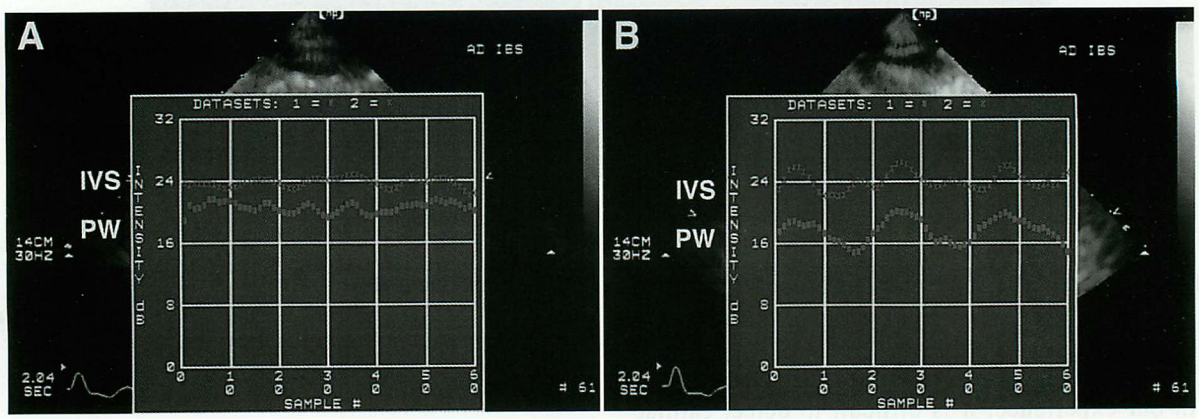

Fig. 6

Fig. 1 Twelve-lead electrocardiogram on admission showing complete atrioventricular block, giant negative T waves in leads V_4–V_6, and a prolonged QT interval (QTc: 0.657 sec)

Fig. 2 Two-dimensional echocardiograms showing focal thinning localized to the anterior basal portion of the interventricular septum (*white arrows*)

A: Parasternal long-axis view, *B*: Apical four-chamber view.

Ao = ascending aorta; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

Fig. 3 Technetium-99 m methoxy-isobutyl isonitrile (^{99m}Tc-MIBI) and iodine-123 betamethyl-*p*-iodophenyl-pentadecanoic acid (^{123}I-BMIPP) dual single photon emission computed tomography images obtained at rest

The normalized polar map displays regional perfusion and metabolic abnormalities. Uptake of ^{123}I-BMIPP (*right*) is less than that of ^{99m}Tc-MIBI (*left*) in the anteroseptal, inferoposterior, and apical regions.

Fig. 4 Gallium-67 scintigrams showing normal uptake in the myocardium

Fig. 5 Photomicrographs of left ventricular endomyocardial biopsy specimens showing interstitial fibrosis, degeneration of cardiac myocytes, and a cluster of spindle-shaped cells

A: Hematoxylin-eosin stain, *B*: Elastica-Masson-Goldner stain.

Fig. 6 Cyclic variation of integrated backscatter of the left ventricular myocardium before (*A*) and 1 month after the beginning of the corticosteroid therapy (*B*)

The magnitude of cyclic variation was significantly reduced before steroid therapy and returned to the baseline after 1 month of therapy.
IVS = interventricular septum; PW = left ventricular posterior wall.

要　約

完全房室ブロックと心不全を呈し心エコー図の特徴的な所見が診断に有用であった心サルコイドーシスの1例

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症　例　67歳，男性
主　訴：呼吸困難

現病歴：27年間高血圧の治療を受けてきたが，次第に労作時の息切れが強くなり，起坐呼吸となったため当院集中治療室へ入室となった．入室時，心不全を呈していたが，心電図は完全房室ブロック，QT延長，巨大陰性T波を示し，経胸壁心エコー図では軽度左室肥大，瀰慢性左室収縮能低下（駆出率22%）および心室中隔基部に限局する異常な菲薄化を認めた．^{99m}Tc-MIBI，^{123}I-BMIPP dual SPECTでは両トレーサーの取り込みに乖離が顕著であったが，^{67}Gaシンチグラフィーでは異常な取り込みは認められなかった．

集中治療室入室後，心不全の治療とペースメーカー植え込み術が施行された．原因疾患として主に高血圧性心疾患，虚血性心疾患，拡張型心筋症などの鑑別が必要とされたが，心エコー図の所見で心室中隔基部に限局した異常な菲薄化を認めたことから，心サルコイドーシスも鑑別疾患の一つに加えられた．生化学的検査，冠動脈造影，左室心内膜生検の結果から心サルコイドーシスと診断され，ステロイド療法を行った．断層・Mモード心エコー図により心機能の改善をみたが，それに先立ってintegrated backscatterのcyclic variationの改善が認められた．

心サルコイドーシスの診断は時に困難であるが，本例のように典型的な心エコー図所見を呈する場合は必ず，鑑別診断の一つに加える必要がある．

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