J Cardiol 1998; 31: 381-384

Cardiovascular Imaging In-a-Month

● Technetium-99m (V) Dimercaptosuccinic Acid Myocardial Scintigraphy in a 74-Year-Old Woman With Congestive Heart Failure

Takashi HATORI, MD
Ryotaro SEKI, MD
Takuji TOYAMA, MD
Akira HASEGAWA, MD
Tsutomu IWASAKI, MD
Takahiro YAMAGISHI, MD
Yoshiaki KANEKO, MD
Ryozo NAGAI, MD, FJCC

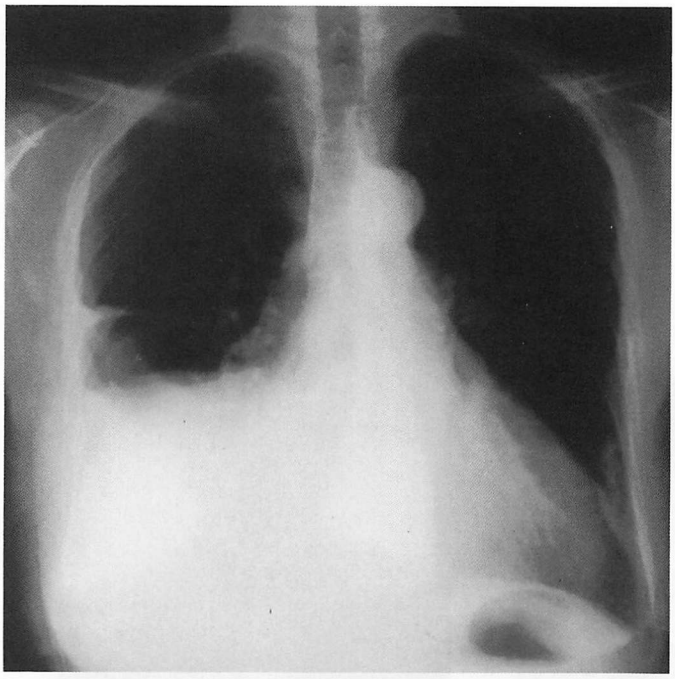

Fig. 1

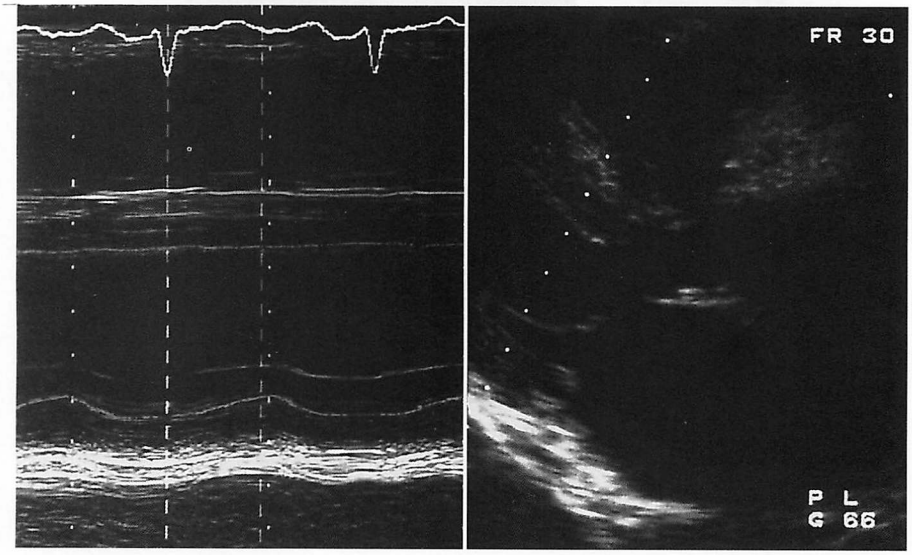

Fig. 2

群馬大学医学部 第二内科：〒371-8511 群馬県前橋市昭和町 3-39-15
The Second Department of Internal Medicine, Gunma University School of Medicine, Gunma
Address for reprints: HATORI T, MD, The Second Department of Internal Medicine, Gunma University School of Medicine, Showa-machi 3-39-15, Maebashi, Gunma 371-8511
Manuscript received May 7, 1998

CASE

A 74-year-old woman with a one-year history of atrial fibrillation was admitted to our hospital because of the gradual onset of dyspnea, malaise and loss of appetite over the past 3 months. Physical examination revealed macroglossia, hepatomegaly and ankle edema. Radiography showed cardiomegaly and right-sided pleural effusion with lower lobe atelectasis (**Fig. 1**). Echocardiography revealed increased left ventricular wall thickness with markedly reduced contraction (**Fig. 2**). Technetium-99m (V) dimercaptosuccinic acid [^{99m}Tc (V) DMSA] scintigraphy showed prominent accumulation of the tracer in the myocardium, right lower lobe of the lung, thyroid gland and tongue with nonspecific accumulation in the urinary tract (**Fig. 3**).

Fig. 3

Discussion of Diagnosis

Laboratory examination revealed the presence of serum M-protein and urinary Bence-Jones protein. Bone marrow aspiration disclosed plasmacytoma and radiography of the bone showed osteolytic lesions in the skull. Biopsy of the right pleura revealed deposition of amyloid (**Fig. 4**). Based on these findings, the diagnosis was amyloidosis associated with multiple myeloma. We could not perform cardiac catheterization because of rapid progression of congestive heart failure refractory to medical therapy. No chemotherapy was given and the patient died three months after admission to the hospital. Necropsy of the heart revealed massive amyloid deposition in the myocardium (**Fig. 5**).

Amyloidosis consists of a group of diseases that are characterized by the deposition of one of several proteinaceous materials known as amyloids in one or more organs. Such lesions are rare and occur mainly in the respiratory, gastrointestinal and urinary tracts and the heart in patients with plasmacytoma. The presence of amyloidosis can be suspected from various signs and symptoms, but the definitive diagnosis can only be made by histochemical analysis of biopsy specimens.

^{99m}Tc(V)DMSA is a tumor imaging agent that has been used to evaluate amyloidosis with plasmacytoma[1] and primary amyloidosis[2]. Patients with cardiac amyloidosis are frequently hemodynamically unstable for biopsy procedures, as in the present case. In such patients with cardiac amyloidosis, ^{99m}Tc(V)DMSA scintigraphy is a useful alternative for detecting the distribution of amyloid deposits localized in the heart, liver and other organs.

Diagnosis : Cardiac amyloidosis

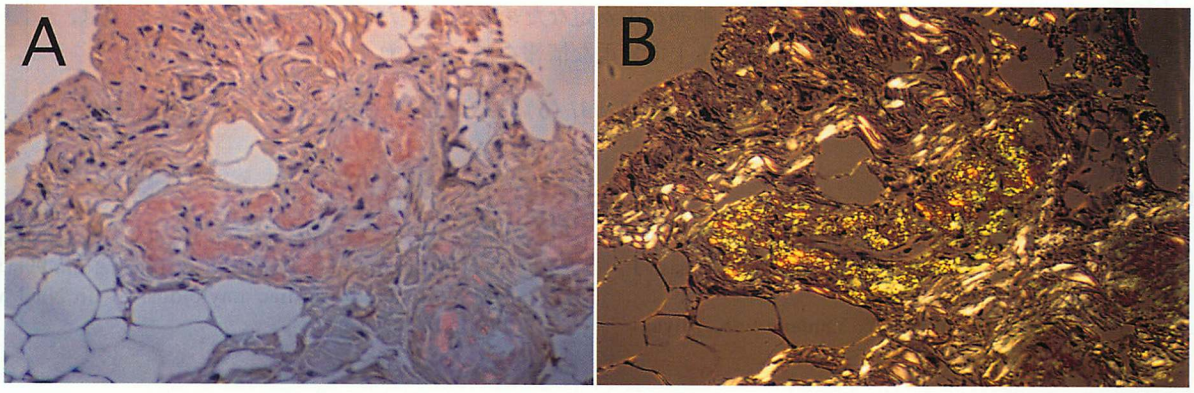

Fig. 4

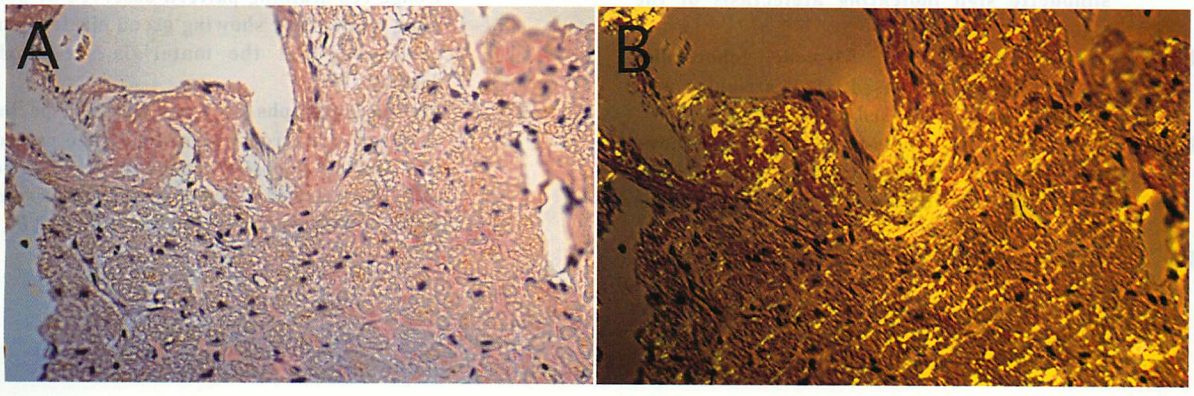

Fig. 5

要　約

Technetium-99m(V) Dimercaptosuccinic Acid心筋シンチグラフィーが診断に有用であった心アミロイドーシスの1例

羽鳥　貴　岩崎　勉　関　亮太郎　山岸高広
外山卓二　金古善明　長谷川　昭　永井良三

症　例　74歳，女性

現病歴：1年前より心房細動で近医に通院中，3ヵ月前より労作時呼吸困難，全身倦怠感，食欲不振が出現し，精査加療のため当院入院．入院時，巨舌，肝腫大，下腿浮腫を認めた．胸部X線写真では心陰影の拡大，右側胸水貯留と右下葉の無気肺を認め，心エコー図上，左室壁は肥厚し，壁運動は著明に低下していた．検査では尿中Bence-Jones蛋白と血中M蛋白を認め，骨髄穿刺，胸水穿刺にて異型形質細胞が増加しており，頭蓋骨X線写真にて骨融解像を認めた．

診断のポイント：以上より多発性骨髄腫に合併したアミロイドーシスを疑い，technetium-99m(V) dimercaptosuccinic acid [^{99m}Tc(V)DMSA] シンチグラフィーを施行したところ，心筋，右肺下葉，甲状腺，舌にアミロイドの沈着を示唆するトレーサーの異常集積を認めた．右側胸膜生検にて胸膜にアミロイドの沈着を認め，アミロイドーシスと診断したが，その後，急激に進行する治療抵抗性の心不全のため心臓カテーテル検査，化学療法は施行出来ず，入院3ヵ月後に死亡．剖検にて心筋にアミロイドの沈着が証明され，^{99m}Tc(V)DMSAシンチグラフィー像の所見は臓器へのアミロイドの沈着を示すものであることが確認された．

心アミロイドーシスはアミロイドーシス患者の予後を規定する重要な臓器障害であり，組織生検でのみ診断されうる．^{99m}Tc(V)DMSAシンチグラフィーは軟部腫瘍やアミロイドーシスの画像診断に用いられており，本例のように血行動態が不安定なため心筋生検が困難であるような症例では，アミロイドの浸潤部位，予後を推定する上で貴重な情報を与えてくれる．

J Cardiol 1998; 31 (6): 381–384

References

1) Ohta H, Endo K, Katoh T, Konishi J, Kotoura H: Technetium-99m(V)DMSA uptake in amyloidosis. J Nucl Med 1989; **30**: 2049–2052
2) Ohta H, Okada T, Fukukawa Y, Yamamoto K, Miyaki Y, Shizuki K, Kotoura H, Endo K, Konishi J, Ohshima C, Nakamura Y, Kawai C: Technetium-99m(V) DMSA uptake in cardiac amyloidosis. Clin Nucl Med 1991; **16**: 673–675

Fig. 1 **Chest radiograph on admission demonstrating cardiomegaly and right pleural effusion with silhouette sign indicating atelectasis of the right lower lobe**

Fig. 2 **Two-dimensional and M-mode echocardiograms showing increased left ventricular wall thickness and decreased contraction**

Fig. 3 **^{99m}Tc(V)DMSA whole-body scintigram showing abnormal high accumulation in the myocardium, lower lobe of right lung, tongue and thyroid gland**

Fig. 4 **Photomicrographs of the pleural biopsy specimen showing a large amount of Congo red positive proteinaceous materials (*A*), and the Congo red staining pattern observed by polarizing microscopy showing green birefringence, confirming that the materials are amyloid deposits (*B*)**

Fig. 5 **Photomicrographs of a myocardial specimen obtained at necropsy showing Congo red positive amyloid deposits around myocardial cells (*A*), and amyloid deposits with green birefringence under the polarizing microscope (*B*)**