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Two Cases of Peripartum Cardiomyopathy

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Abstract

Two cases of peripartum cardiomyopathy, a rare type of dilated cardiomyopathy, are reported.

A 36-year-old woman developed congestive heart failure 1 month after delivering her third child. Cardiac catheterization revealed diffuse hypokinesis of the left ventricle and an ejection fraction of 28%. The second study, 20 months later, demonstrated an ejection fraction of 46%. Endomyocardial biopsy showed mild interstitial edema.

A 42-year-old woman developed toxemia during pregnancy. She delivered her second child at 38 weeks of gestation. Two weeks later, she developed congestive heart failure. Cardiac catheterization demonstrated diffuse hypokinesis of left ventricle with an ejection fraction of 40%. Endomyocardial biopsy revealed dense fibrosis. Follow-up angiography performed 8 months later showed similar findings with an ejection fraction of 34%.

These two cases suggest the importance of evaluation of endomyocardial biopsy to determine the degree of interstitial fibrosis that may reflect the prognosis for patients with peripartum cardiomyopathy confirmed by measurements of ejection fraction.

Key Words

cardiomyopathy (peripartum), angiocardiography, myocarditis, pathology, endomyocardial biopsy

INTRODUCTION

Peripartum cardiomyopathy (PPCM) is a rare type of dilated cardiomyopathy of unknown origin[1–17]. We present two patients with PPCM confirmed by the clinical courses and results of cardiac catheterization and endomyocardial biopsy.

CASE PRESENTATIONS

Case 1

A 36-year-old, gravida 3, para 2, woman delivered her third child after an uneventful pregnancy on December 26, 1989. Approximately 1 month later, she experienced coughing and dyspnea. A local physician diagnosed congestive heart failure and she was transferred to our hospital. Her past history was uneventful. A chest radiograph and electrocardiogram taken prior to this pregnancy showed no abnormalities. Her family history revealed that her mother had died of myocardial infarction aged 61 years.

On admission the patient showed signs of pulmo-

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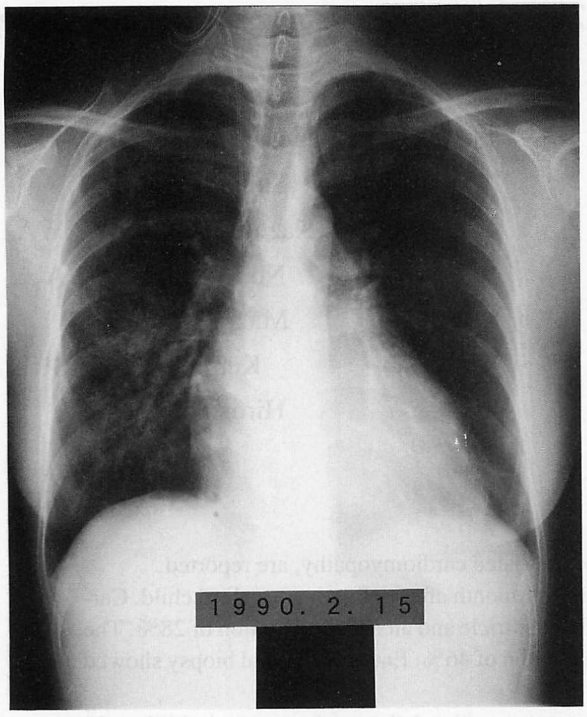

Fig. 1 Case 1. Chest radiograph on admission showing cardiomegaly, cardiothoracic ratio of 70%, and pulmonary congestion

nary congestion and bilateral, lower extremity pitting edema. Laboratory data revealed a normal complete blood count and urinalysis, but slightly increased serum C-reactive protein level, hypoproteinemia, and hypoalbuminemia were noted. Transaminase and renal function tests were initially abnormal, but gradually improved after treatment with diuretics and vasodilator. Antinuclear antibody and CH_{50} titers were within normal limits. Paired titers of Coxsackie virus and other viruses were also normal.

A plain chest radiograph showed cardiomegaly, a cardiothoracic ratio (CTR) of 70%, and evidence of lung field congestion (**Fig. 1**). An electrocardiogram (ECG) displayed a normal sinus rhythm, with frequent premature ventricular contractions, and inverted T wave in leads V_2–V_6 (**Fig. 2**). An echocardiogram demonstrated diffuse hypokinesis and dilatation of the left ventricular wall. Cardiac catheterization was performed twice, in July 1990 and March 1992. The initial study revealed normal coronary arteries, diffuse hypokinesis of the left ventricular wall, and a left ventricular ejection fraction (EF) of 28%. The cardiac index (CI) by the thermodilution method was 2.47 *l*/min/m^2. However, the second study demonstrated hypokinesis in the apical segment with an EF of 46% and a CI of

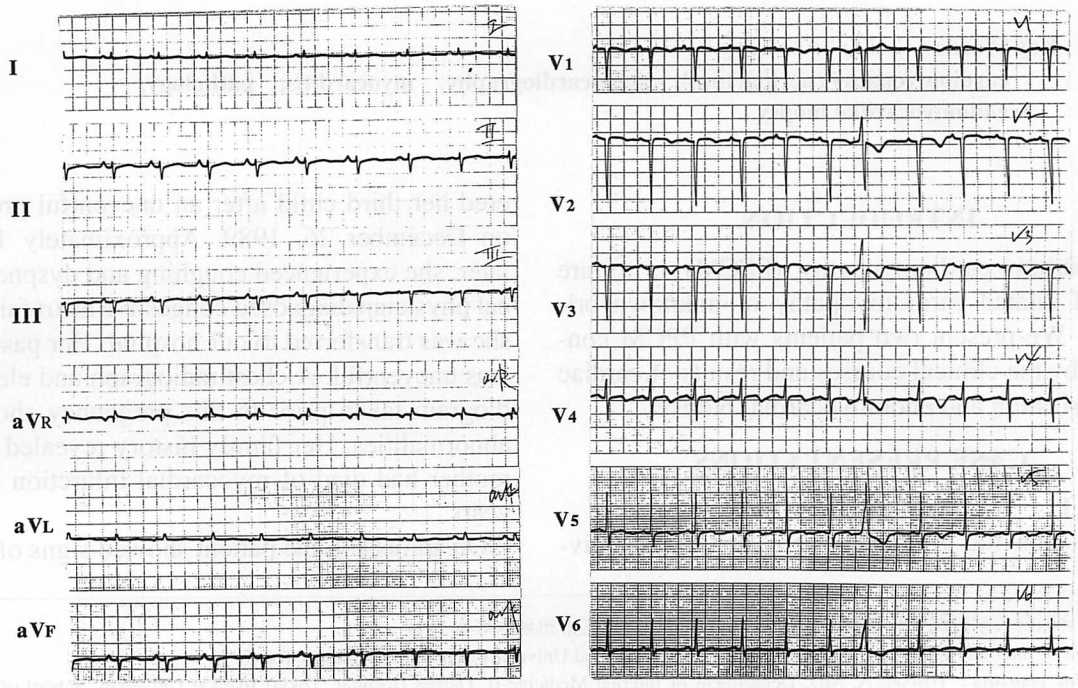

Fig. 2 Case 1. Electrocardiogram on admission revealing low voltage in the frontal leads, poor R wave progression in the chest leads V_1–V_3, inverted T waves in leads V_2–V_6, and extrasystolic beat

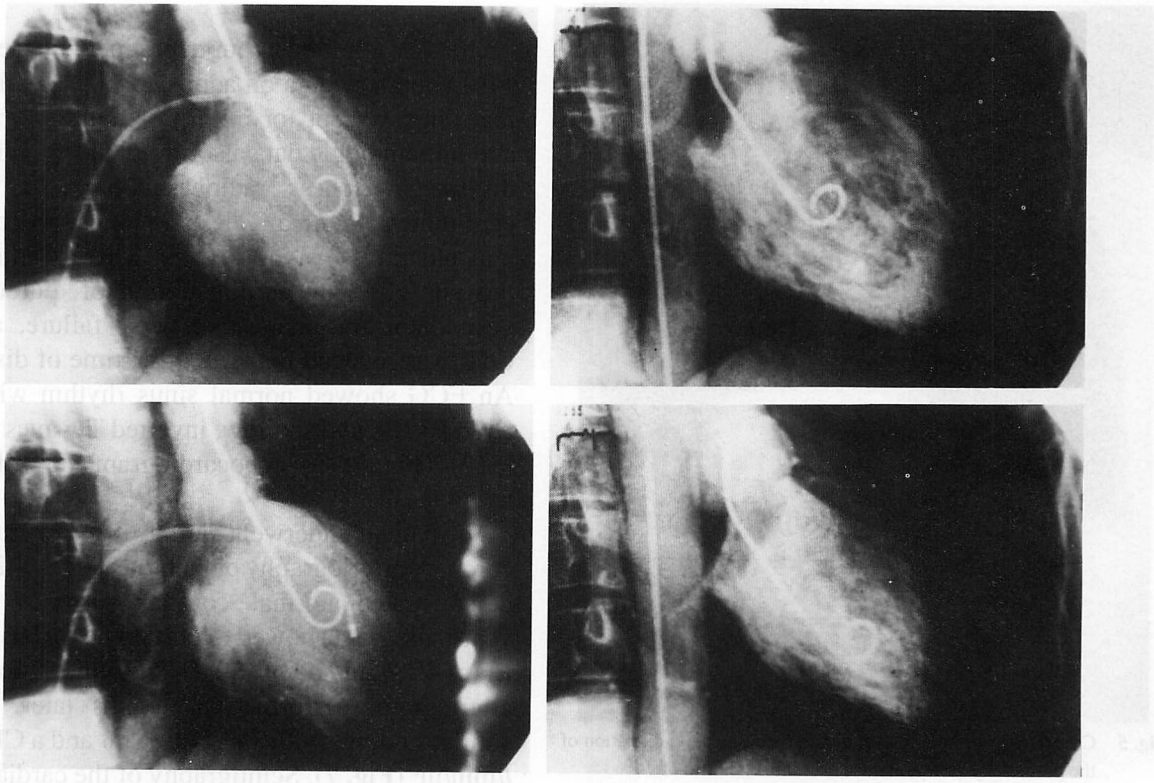

Fig. 3 Case 1. Serial left ventriculograms showing improvement in left ventricular contraction (*left*: July 1990; *right*: March 1992; *above*: diastolic phase; *below*: systolic phase)
The ejection fraction on the left was 28%, on the right 46%.

4.18 *l*/min/m^2 (**Fig. 3**).

An endomyocardial biopsy was performed simultaneously. Light microscopy of the biopsy specimens demonstrated mild, focal, interstitial edema without inflammatory cell infiltration. Myocytes showed anisonucleosis. Mild interstitial fibrosis was present but confined mainly to the subendocardial layer (**Fig. 4**). A second biopsy specimen revealed similar features. For the past 3 years, the patient has been followed up at the outpatient clinic. She is NYHA class 1 and receives medication only for ventricular arrhythmia.

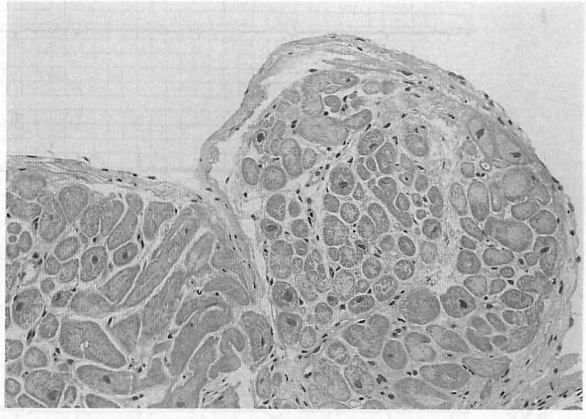

Fig. 4 Case 1. Light microscopy of the endomyocardial biopsy specimen from the left ventricle performed during initial cardiac catheterization, showing mild endocardial fibrosis, interstitial fibrosis, myocyte swelling and a few inflammatory cells (hematoxylin & eosin stain, original magnification ×50)

Case 2

A 42-year-old, gravida 1, para 1, woman had a past history of gastric ulcers 5 years earlier and a 1 year history of chest pain. Coronary angiography in January 1987 demonstrated no lesions. A left ventriculogram showed good contraction with an EF of 61%. An ergonovine test was negative. Her family history was negative for cardiac diseases. She developed toxemia during pregnancy diagnosed at 37 weeks of gestation. Hypertension and edema were treated by fluid restriction and diuretics. She delivered an infant at 38 weeks of gestation. She was discharged from the hospital on November 11, 1991. Two weeks later, she complained of dyspnea on exertion and facial and lower extremity

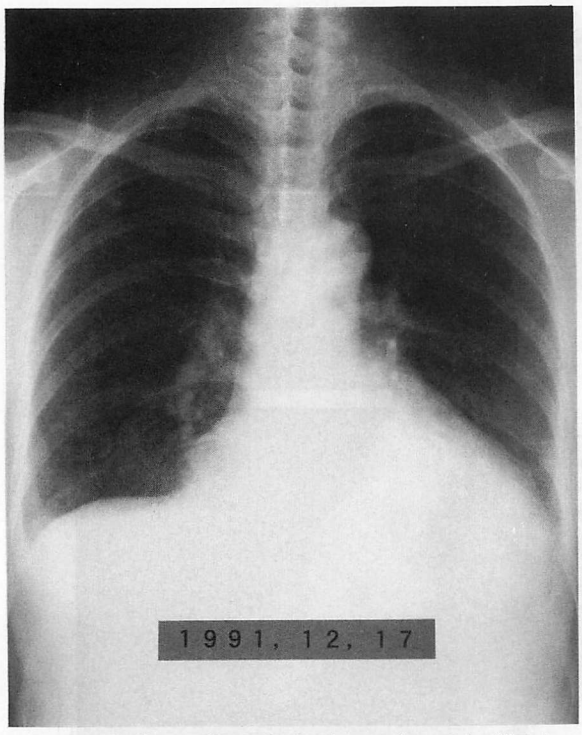

Fig. 5 Case 2. Chest radiograph showing cardiomegaly, dilatation of the pulmonary artery, and a cardiothoracic ratio of 74%

edema.

She was admitted to our department on December 17. Laboratory data on admission were within normal limits except for mild proteinuria. Viral titers and immunologic data also revealed no evidence of disease. A chest radiograph showed marked cardiomegaly with a CTR of 74% and a small bilateral pleural effusion (**Fig. 5**). These findings improved following administration of diuretics and vasodilator for congestive heart failure, and the CTR was reduced to 51% at the time of discharge. An ECG showed normal sinus rhythm with scattered PVC's and shallow, inverted T waves in leads V_1–V_6 (**Fig. 6**). An echocardiogram showed diffuse hypokinesis.

The patient underwent cardiac catheterization and coronary arteriography on February 12, 1992, which demonstrated normal coronary arteries. A left ventriculogram showed diffuse hypokinesis with an EF of 40% and a CI of 2.21 *l*/min/m^2. A follow-up arteriogram, performed 8 months later, showed similar findings with an EF of 34% and a CI of 2.66 *l*/min/m^2 (**Fig. 7**). Scintigraphy of the cardiac circulation revealed an EF of only 47% by first pass images.

An endomyocardial biopsy was performed. Light

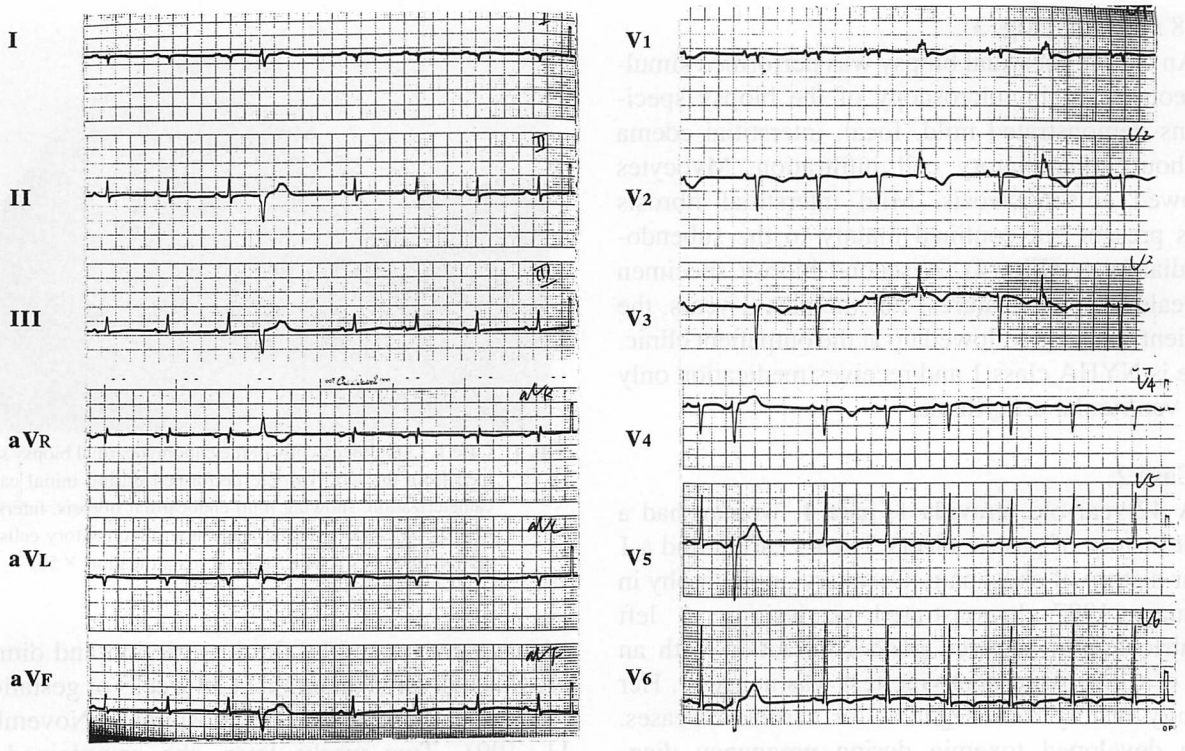

Fig. 6 Case 2. Electrocardiograms on admission exhibiting right axis deviation, frequent premature ventricular contractions, poor R wave progression in leads V_1–V_4, and inverted T waves in leads V_2–V_6

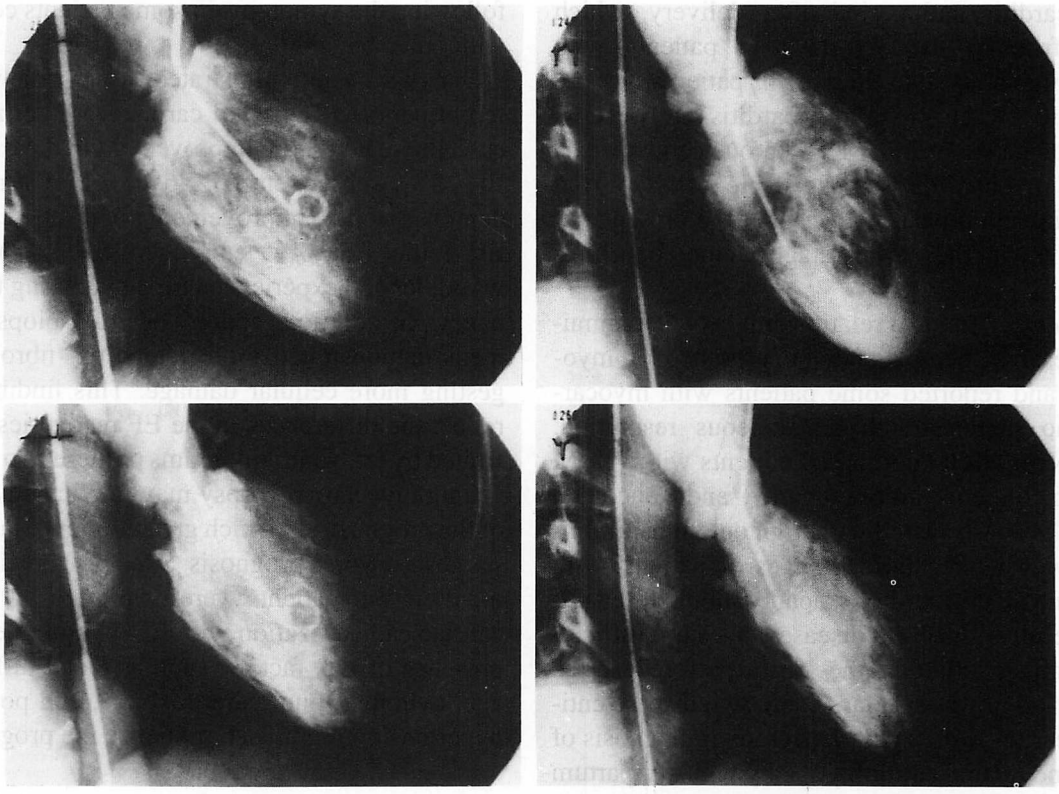

Fig. 7 Case 2. Serial left ventriculograms showing no improvement in left ventricular contraction (*left*: February 1992; *right*: October 1992; *above*: diastolic phase; *below*: systolic phase)
The ejection fraction on the left was 40%, on the right 39%.

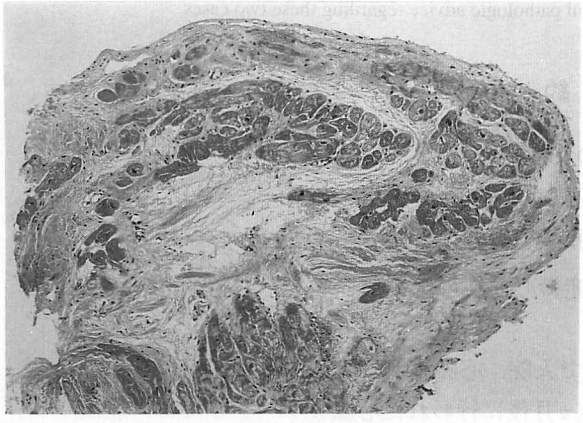

Fig. 8 Case 2. Light microscopy of an endomyocardial biopsy specimen from the left ventricle demonstrating extensive interstitial fibrosis, but few inflammatory cells (hematoxylin & eosin stain, original magnification ×25)

microscopy of the biopsy specimen revealed dense fibrosis located in the subendocardial layer with extension into the myocardial layer. No interstitial inflammatory cellular infiltrates were present (**Fig. 8**). A follow-up biopsy sample demonstrated essentially the same features. The patient has been followed up periodically at our outpatient department for 2 years as NYHA class 2. A recent echocardiogram was essentially unchanged.

DISCUSSION

The etiology of PPCM is unknown, but pregnancy and/or related conditions are potential causes[15]. Criteria for diagnosis include demonstration of no definite etiology prior to the last month of pregnancy and development of peripartum heart failure[15]. The former is difficult to fulfill because not every pregnant woman receives detailed medical examination. Our case 2 received cardiac catheterization and coronary arteriography to evaluate chest pain which showed no lesion. Previous reports have suggested that risk factors for PPCM include: black race, older mothers, twinning, toxemia during pregnancy, multipara, and the presence of postpartum hypertension[5,10]. Our case 1 was multipara, while case 2 demonstrated toxemia during pregnancy and an older mother. Recent endomyocardial biopsy studies have shown no relationship between presence

of myocarditis and period after delivery, which ranged from 6 to 195 days in patients with myocarditis, and 33 to 96 days in patients without myocarditis[8,16]. However, myocarditis rarely occurs in the late stages of PPCM[8,16]. In our patients, evidence of myocarditis was negligible in tissue from both initial and repeat biopsies, but the presence of myocarditis in the earlier stage cannot be eliminated.

Midei *et al*[16] found a relationship between immunosuppressive therapy and the presence of myocarditis, and reported some patients with myocarditis who demonstrated spontaneous resolution. Martin-Neto *et al*[17] compared patients with PPCM and with dilated cardiomyopathy, and concluded that PPCM is associated with high output heart failure. However, the clinical stages of patients with PPCM and dilated cardiomyopathy should be evaluated carefully, since the disease stages are difficult to determine, although the period before cardiac catheterization was similar. These two disease entities appear to be different. Differential diagnosis of PPCM and dilated cardiomyopathy with peripartum congestive heart failure is possible based on the criteria of two entities, since the cardiac condition before pregnancy is different.

The prognoses for patients with PPCM vary. The clinical and laboratory parameters recover rapidly following the symptoms in some patients compared with others[6,7,10,14].

Our two patients illustrated these somewhat different courses. Repeated cardiac catheterization in case 1 revealed some improvement in the EF (initial EF 28%, subsequent EF 46%). However, case 2 demonstrated no improvement in the left ventricular EF (initial EF 40%, subsequent EF 39%). Case 2 was older and experienced toxemia during the pregnancy. In addition, endomyocardial biopsy specimens demonstrated more interstitial fibrosis, suggesting more cellular damage. This finding might reflect the differences in the EF recoveries demonstrated by left ventriculograms of these two patients.

Endomyocardial biopsy may facilitate evaluation of the prognosis, in which greater interstitial fibrosis suggests a worse prognosis. Such prognostic assessments in our patients were confirmed by repeated cardiac catheterization with EF measurements. The presence of risk factors, such as racial differences, age, twinning, multipara, toxemia, and postpartum hypertension might also influence the prognosis[10].

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要　約

産褥心筋症の2症例

外丸　晃久　後藤　義久　三浦　茂　瀧川　和俊
香川　昇　工藤　眞　我妻　賢司　岡野　弘

われわれは最近，拡大型心筋症としてはまれな産褥心筋症の2例を経験し，心臓カテーテルと心筋生検により確診しえたので報告した．

症例1は36歳，女．第3児分娩1ヵ月後心不全で入院．心臓カテーテル所見は左室のび漫性壁運動の低下で，左室駆出率は28%であった．20ヵ月後に行われた心臓カテーテルでは，駆出率は46%に回復していた．心筋生検では軽度の浮腫を示すのみであった．

症例2は42歳，女．妊娠中に中毒症を経験している．38週で第2児を分娩し，2週間後に心不全で入院した．心臓カテーテルでは左室のび漫性壁運動の低下が認められ，左室駆出率は40%であった．心筋生検は高度の心内膜下の線維化を示していた．8ヵ月後の心臓カテーテル所見は前回同様で，左室駆出率は34%であった．

これら2症例より心筋生検の評価で産褥心筋症の予後の判定が可能であることが示唆された．これは左室駆出率の経時的測定により確認した．

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