Degos' Disease With Constrictive Pericarditis

— A Case Report —

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A 47-year-old man with Degos' disease was examined by echocardiography, which showed hypokinesis of the apical left ventricular wall with pericardial effusion. To evaluate the myocardial perfusion and coronary flow reserve, ^{201}Tl scintigraphy and intracoronary Doppler flowmetry were performed. The coronary flow reserve was not decreased nor was there angiographical coronary stenosis, although a pressure study revealed constrictive dysfunction of both ventricles. The constrictive pericarditis might have been induced by pericardial vasculitis, thereby causing the left ventricular wall motion abnormality. (*Jpn Circ J* 2000; 64: 464–467)

Key Words: Constrictive pericarditis; Coronary flow; Degos' disease

Degos' disease (malignant atrophic papulosis) has been rarely reported. Histology reveals systemic lymphocytic vasculitis with thrombi in small or medium-sized vessels of the skin, gastrointestinal tract and central nerve system.^{1–6} Although pericarditis was detected in a series of autopsies,⁴ the incidence of clinically apparent myocardial dysfunction was rare.^{1–4}

We present a case of Degos' disease with heart failure and demonstrable constrictive pericarditis that developed into the cardiac dysfunction with minor impairment of the coronary circulation.

Case Report

A 47-year-old man was admitted to the Dermatology Department with multiple papular lesions distributed around the trunk and neck in a typical central white zone with an erythematous border. Histopathology of a cutaneous biopsy showed markedly thin smooth muscle of vessels with invasion of lymphoid cells (Fig 1). In addition, the patient had a past history of surgery for gastrointestinal perforation and had subsequently been diagnosed with Degos' disease in 1996.

He had had dyspnea for 2 months before he presented to hospital in June 1999. On admission, body temperature was 38.0°C, blood pressure was 90/60 mmHg, and heart rate was 100 beats/min with a regular rhythm. Multiple papular lesions were observed. The liver was palpable (2.0 cm at epigastric region), there was distention of the jugular vein, and marked edema of the extremities. Wet rales were audible in the lower lung field, and heart auscultation revealed a diastolic monophasic friction rub. The electrocardiogram showed ST-T depression in V₃ and V₄ and

negative T in II, III, and aV_F. Blood tests revealed a white blood cell count of 3,200 cells/mm³, red blood cell count of 357×10⁴/mm³ (hemoglobin of 9.9 g/dl), platelet count of 43.3×10⁴/mm³, creatine kinase of 62 U/L, and C-reactive protein of 3.0 mg/dl. Chest X-ray showed pleural effusion of the right lung, and a cardiothoracic ratio of 47%. Two-dimensional echocardiography showed hypokinesis of the left ventricular apical wall with pericardial effusion and a high-echo-intensity partial lesion of the pericardium (Fig 2). M-mode echocardiogram revealed narrowing of the left ventricle and two-peaked diastolic septal motion (Fig 2). A chest computed tomographic (CT) scan revealed pleural effusion of the right lung, pericardial effusion, and pericardial calcification (Fig 3). Exercise stress ^{201}Tl -betamethyl-p-iodophenyl-pentadecanoic acid (BMIPP) myocardial scintigraphy showed normal distribution. Cardiac catheterization was performed and showed a central venous pressure of 15 mmHg, right ventricular pressure of 25/16 mmHg (end-diastolic pressure of 16 mmHg), and left ventricular pressure of 81/6 mmHg (end-diastolic pressure of 16 mmHg). Left ventriculography revealed hypokinesis of the left ventricular apical wall, which was also seen on the echocardiography, and an ejection fraction of 65%, despite no angiographical stenosis of the coronary artery at baseline or during hyperemia (intracoronary infusion of 0.14 mg·kg body weight⁻¹·min⁻¹ of adenosine). To evaluate the impairment of myocardial microcirculation, coronary blood flow was measured using a 15-MHz Doppler wire (FloWire, 0.014 inch, Cardiometrics, Inc, CA, USA) and coronary flow reserve was obtained from the hyperemia/baseline flow ratio using a previously reported formula.⁷ Baseline coronary flow showed rapid acceleration and more rapid deceleration in diastole and retrograde flow in systole.⁸ Coronary flow reserve was 4.8 of the left anterior descending artery and 5.4 of the left circumflex artery (Fig 4), and impairment of the myocardial circulation was disregarded.

The patient was treated with aspirin 100 mg and dipyridamole 150 mg/day for the Degos' disease and diuretics (Flosemide 10 mg) for the general edema after the cardiac catheterization. After 6 months, the patient continued this

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Fig 1. Cutaneous biopsy showing perivascular infiltration of lymphocytes (H&E, $\times 400$).

Fig 2. Two-dimensional (Upper) and M-mode (Lower) echocardiographic images. The left ventricular apical wall (arrows) was hypokinetic and the M-mode echocardiogram showed abnormal diastolic septal motion. LV, left ventricle; LA, left atrium.

Fig 3. Chest CT scan showing pleural effusion, pericardial effusion and pericardial calcification (arrows).

Fig 4. Coronary flow of the left anterior descending artery (Left: baseline, Right: hyperemia because of intra-coronary infusion of $0.14 \text{ mg} \cdot \text{kg body weight}^{-1} \cdot \text{min}^{-1}$ of adenosine).

therapy, and his condition has continued to be stable.

Discussion

In the present case, we demonstrated that constrictive pericarditis associated with Degos' disease developed into cardiac dysfunction with left ventricular wall motion abnormality, but the coronary microcirculation was relatively unimpaired.

Cardiac involvement of Degos' disease is seldom reported. Cabré et al presented an infant case who died with disseminated myocardial infarcts² and 2 cases of constrictive pericarditis have been described^{3,4}. One of the 2 cases underwent a pericardiectomy but died from intestinal hemorrhage about 4 months after the operation³. The other case reported by Pierce and Smith⁴ also died after developing both heart failure and respiratory disorder about 3 months after a decortication of the left lung. Histology of the heart of both cases showed myocardial scarring or myocardial fibrosis as well as pericardial impairment, and Pierce and Smith suggested that myocardial impairment

was present without evidence of associated vascular lesions⁴.

In the present case, cardiac catheterization revealed neither epicardial or microvascular lesions of the coronary artery nor a decrease in coronary flow reserve, although the wall motion abnormality was observed by echocardiography and left ventriculography^{9,10}. These findings indicate that there was no evidence of myocardial vasculitis inducing the wall motion abnormality, despite histopathological systemic lymphocytic vasculitis with thrombi in small or medium-sized vessels of the skin, gastrointestinal tract and central nerve system.

Pleural adhesion or fibrous thickening as the result of organization of effusion or scarring secondary to vasculitis has been reported^{3,4}. In the pericardium, Pierce and Smith⁴ also demonstrated that the pathologic process might lead to constrictive pericarditis, the possible major source of morbidity, but they did not describe an effective treatment for the disease. On the other hand, Drucker⁵ and Stahl et al⁶ reported platelet dysfunction with the disease and successful treatment with antiplatelet therapy, aspirin and dipyri-

damole. In the present case, the patient has been given antiplatelet therapy, following the cardiac catheterization, and his condition has remained stable.

We agree that vasculitis in pericardium with microthrombus may play an important role in the development of pericardial organization and consider that antiplatelet therapy is effective for patients with Degos' disease.

References

1. Strole WE, Clark WH, Isselbacher KJ: Progressive arterial occlusive disease (Kohlmeier-Degos): A frequently fatal cutaneous systemic disorder. *N Engl J Med* 1967; **276**: 195–201
2. Cabré J, Noguer-Debray S, Bosch-Castané J, Huguet P, Gonzalez JA, Moraga F, et al: Papulose atrophique maligne de Degos chez un nourrisson. *Bull Soc Fr Dermatol Syphiligr* 1974; **81**: 652
3. Roenigk H, Farmer R: Degos' disease (malignant papulosis). *JAMA* 1968; **206**: 1508–1514
4. Pierce RN, Smith GJW: Intrathoracic manifestations of Degos' disease (malignant atrophic papulosis). *Chest* 1978; **73**: 79–84
5. Drucker CR: Malignant atrophic papulosis: Response to antiplatelet therapy. *Dermatologica* 1990; **180**: 90–92
6. Stahl D, Thomsen K, Hou-Jensen K: Malignant atrophic papulosis. *Arch Dermatol* 1978; **114**: 1687–1689
7. Akasaka T, Yoshida K, Hozumi T, Takagi T, Kaji S, Kawamoto T, et al: Retinopathy identifies marked restriction of coronary flow reserve in patients with diabetes mellitus. *J Am Coll Cardiol* 1997; **30**: 935–941
8. Akasaka T, Yoshida K, Yamamuro A, Hozumi T, Takagi T, Morioka S, et al: Phasic coronary flow characteristics in patients with constrictive pericarditis: Comparison with restrictive cardiomyopathy. *Circulation* 1997; **96**: 1874–1881
9. Voelkel AG, Pietro DA, Folland ED, Fisher SL, Parist AF: Echocardiographic features of constrictive pericarditis. *Circulation* 1978; **58**: 871–875
10. Engel PJ, Fowler NO, Tei C, Shah PM, Driedger HJ, Shabetai R, et al: M-mode echocardiography in constrictive pericarditis. *J Am Coll Cardiol* 1985; **6**: 471–474