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SCLERODERMA RENAL CRISIS IN PROGRESSIVE SYSTEMIC SCLEROSIS: A CASE REPORT

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We report a case of progressive systemic sclerosis with scleroderma renal crisis 10 years after onset. The patient (female) had progressive renal dysfunction, hypertension which was difficult to control, and massive gastrointestinal bleeding. An angiotensin converting enzyme inhibitor (enalapril) could not control her hypertension. Only intravenous nicardipine had a slight effect on her hypertension. Hemodialysis and plasma exchange, did not reverse the renal crisis, and the patient died. Microscopic examination of her kidney showed thickening of the capillary walls and mild nephrosclerosis.

Key words: Progressive systemic sclerosis, Scleroderma renal crisis

INTRODUCTION

Scleroderma renal crisis is a common cause of death in patients with progressive systemic sclerosis. It is usually accompanied by malignant hypertension. Angiotensin converting enzyme inhibitors are thought to be effective. However, we report a case of progressive systemic sclerosis with scleroderma renal crisis on whom an angiotensin converting enzyme inhibitor (enalapril) was not effective.

CASE REPORT

A 37-year-old female noticed edema, chills in the fingers and Raynaud's phenomenon in 1979. She was diagnosed as having progressive systemic sclerosis in 1982 and was treated primarily with urokinase and prostaglandin E₁ for five years. However, since 1986, she had not received any medication. In February 1989, she showed general fatigue and dyspnea, and was found to have renal dysfunction and hypertension. On March 11, 1989, she required institutionalization in another hospital for hemodialysis. She was admitted to our hospital on March 14, 1989, due to massive bleeding from the stomach and intestine.

On physical examination, her face was mask-like, her lips were dry and her mouth

opened with difficulty. Her skin was dry and tight, but she had no ulcers or scars on her fingers.

The laboratory data included white blood cell count of 30,900/mm³, hemoglobin value of 9.5 g/dl, platelet count of 9.5×10^4 /mm³, bleeding time of 7 minutes 30 seconds, and erythrocyte sedimentation rate of 7 mm/hr. Serological examination included serum creatinine level of 9.45 mg/dl, blood urea nitrogen of 174.9 mg/dl, bicarbonate of 10.7 mEq/l, creatinine clearance of 1.7 l/day, and plasma renin activity of 31.78 ng/ml/hr. Urinalysis revealed proteinuria of 0.88 g/day. Immunological examination revealed that the antinuclear factor (ANF) was weakly positive in speckled pattern at a 1:80 titer, and none of the other specific antibody levels were significantly high.

Fresh blood, platelet transfusion and gabexate mesilate were administered for treatment of disseminated intravascular coagulation (DIC). However, bleeding from the intestine still continued (Fig. 1).

She was given nicardipine, nifedipine and enalapril to control her blood pressure. Because oral administration of antihypertensive drugs was not possible due to gastrointestinal bleeding she was administered nicardipine intravenously. During the

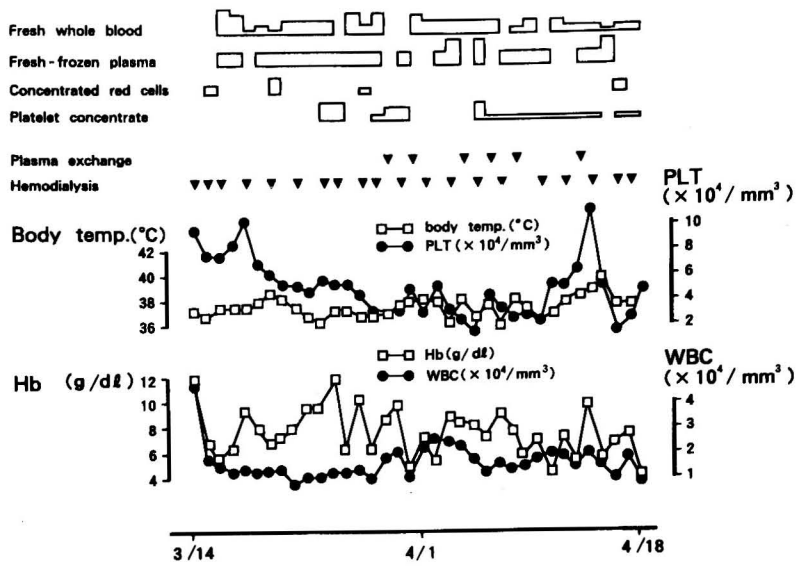


Fig. 1. Clinical course

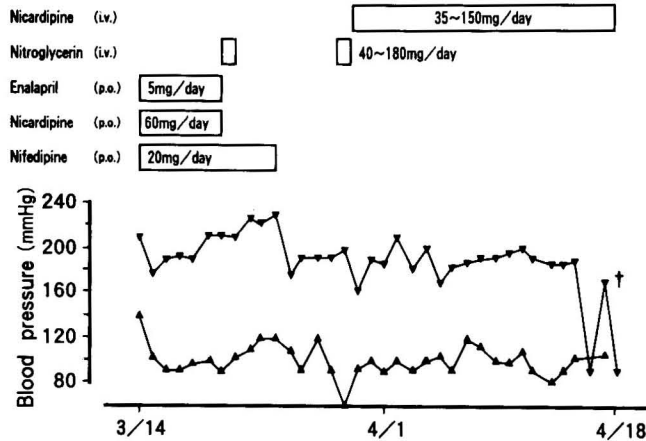


Fig. 2. Blood pressure during the clinical course.

course of therapy, her blood pressure remained high despite a variety of antihypertensive medications (Fig. 2).

Despite almost daily hemodialysis, as a treatment for renal failure, as well as plasma exchange for the progressive systemic sclerosis itself, she eventually died of DIC.

Microscopic examination of her kidney showed thickening of the capillary walls, which indicated mild nephrosclerosis (Fig. 3).

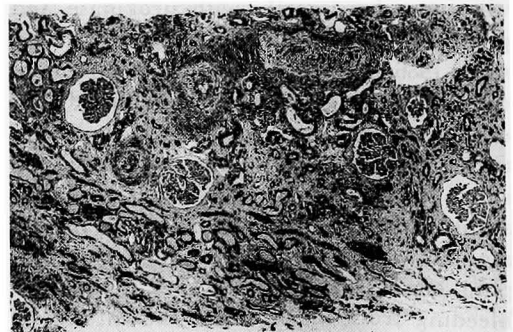


Fig. 3. Renal autopsy showing thickening of the capillary walls. (HE $\times 10$)

DISCUSSION

Scleroderma renal crisis is the rapid deterioration of the renal function with malignant hypertension found in patients with progressive systemic sclerosis. It is still an important and often fatal complication for these patients. Brown et al. reported that 7% of progressive systemic sclerosis patients developed this complication³⁾, and according to Traub et al., it was present in 8.3% of all cases⁴⁾.

The factors predicting scleroderma renal crisis are thought to be: a relatively short duration from onset of progressive systemic sclerosis, urinalysis abnormalities—such as hematuria and proteinuria, malignant hypertension, high renin value and decreasing renal plasma flow. However, neither clinical nor laboratory findings are significant¹⁾. Steen and her co-workers reported that rapid progression of skin thickening and early onset of anemia were likely to be warning signs⁵⁾.

Several treatments for scleroderma renal crisis have been tried, such as hemodialysis after nephrectomy, kidney transplantation and antihypertensive drugs⁴⁾. However, recent studies showed that the most effective therapy for scleroderma renal crisis was the administration of angiotensin converting enzyme inhibitors^{2,6-8)}. The administration of other antihypertensive drugs, such as nifedipine and prazosin, not used in combination with angiotensin converting enzyme inhibitors are thought to be ineffective^{8,9)}.

Ejima et al. listed the patients with scleroderma renal crisis who had been treated by angiotensin converting enzyme inhibitors reported before 1987. Most of the patients who took angiotensin converting enzyme inhibitors before severe renal dysfunction appeared, survived⁹⁾.

In our patient, the renal function had already been severely deteriorated and DIC had occurred when she was admitted

to the hospital. In addition, intestinal bleeding rendered the oral administration of enalapril ineffective, which ultimately caused the malignant hypertension to continue. The causes of death were thought to be uncontrolled DIC and malignant hypertension.

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和文抄録

腎クリーゼを起こした強皮症の1例

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発症より10年後腎クリーゼにて急性腎不全を起こした進行性全身性硬化症の1例を経験した。急激な腎機能低下，コントロール不良な高血圧，大量の消化管出血を認め，治療開始約1ヵ月後死亡した。本症例では入院時すでにDICと腎不全が高度であったこと，消

化管出血によりアンギオテンシン変換酵素阻害剤であるenalaprilが経口投与できなかったことが予後不良であった原因と考えられた。剖検時の腎の光顕所見で動脈内膜の肥厚と軽度の腎硬化像が認められた。

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