Case Report

Acute Type A Aortic Dissection in a Patient with Systemic Lupus Erythematosus

Shigeaki AOYAGI,¹ MD, Hidetoshi AKASHI,¹ MD, Hiroyuki OTSUKA,¹ MD, Hideki SAKASHITA,¹ MD, Teiji OKAZAKI,¹ MD, and Kei-ichiro TAYAMA,¹ MD

SUMMARY

The case of a 27-year-old Japanese woman with type A acute aortic dissection who had been diagnosed with systemic lupus erythematosus (SLE) is presented. The patient also had aortic regurgitation due to non-infective endocarditis and systemic hypertension, and had been maintained on steroid therapy for 15 years. Her twin sister was also diagnosed with SLE. The patient was admitted to emergency due to severe back pain. A chest x-ray showed enlargement of the upper mediastinum. Echocardiography revealed a thickened and deformed aortic valve with aortic regurgitation and dissection of the ascending aorta, but pericardial effusion was not found. Computed tomography demonstrated aortic dissection extending from the ascending aorta to the abdominal aorta. Graft replacement of the ascending aorta and proximal aortic arch was performed under hypothermic circulatory arrest with retrograde cerebral perfusion. The patient recovered uneventfully. Aortic dissection complicated with SLE is extremely rare, and this is only the 15th case reported in the English or Japanese literature. (*Jpn Heart J* 2002; 43: 567-571)

Key words: Systemic lupus erythematosus, Aortic dissection, Steroid therapy

SYSTEMIC lupus erythematosus (SLE) is a chronic systemic inflammatory disease, characterized by malar and discoid rashes, and involves multiple organs. In the cardiovascular system, it frequently causes pericarditis, nonbacterial verrucous endocarditis, coronary artery disease, and cardiomyopathy.¹⁾ However, aortic dissection is a rare cardiovascular manifestation of SLE.²⁻⁵⁾ To our knowledge, only 14 cases of aortic dissection in SLE have been previously reported.^{2,6)}

In this paper, we describe a case of type A acute aortic dissection associated with SLE.

From the ¹ Department of Surgery, Kurume University School of Medicine, Kurume, Fukuoka Japan.

Address for correspondence: S. Aoyagi, M.D., Department of Surgery (2), Kurume University School of Medicine, 67 Asahi-machi, Kurume Fukuoka 830-0011, Japan.

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CASE REPORT

A 27-year-old Japanese woman was admitted to emergency due to severe back pain. She had been diagnosed with SLE at the age of 11 years, and was maintained on steroid therapy with prednisolone of 25 mg/day for 15 years. The patient was also diagnosed with psychosis, and mild aortic regurgitation (AR) resulting from non-infective verrucous endocarditis, as described by Libman and Sacks.⁷⁾ Her twin sister also suffered from SLE. On admission, the patient had no fever or complaints of arthralgia. Her height was 140 cm and body weight was 30.4 kg. Blood pressure was 180/120 mmHg and heart rate was 90/min with a regular rhythm. A Grade 2 to and fro murmur was heard at the 3 - 4th intercostal space on the left sternal border. Hemoglobin was 14.9 g/dL and the white blood count was 11900/mm³. Laboratory studies revealed blood urea nitrogen of 12.7 mg/dL, creatinine of 0.5 mg/dL, and serum albumin of 3.4 g/dL. C-reactive protein was 0.1 mg/dL, and the C₃, C₄, and CH₅₀ concentrations were 78 mg/dL, 11 mg/dL, and 29 units/dL, respectively. Antinuclear antibody was 1:1280. A chest x-ray showed enlargement of the upper mediastinum. Echocardiography revealed a thickened and deformed aortic valve with mild AR, but no pericardial effusion was found. Computed tomography revealed a dilated ascending aorta (4.5 cm in diameter) and a dissection extending from the ascending aorta to the abdominal aorta, as shown in Figure. A diagnosis of Stanford type A acute aortic dissection was made and emergency surgery was performed.

No bloody effusion was found in the pericardial cavity during the operation.

Figure. Computed tomography showing aortic dissection extending from the ascending aorta to the descending aorta.

Cardiopulmonary bypass was instituted through cannulae inserted into the right axillary artery and both caval veins. After aortic cross-clamping, the ascending aorta was transected at 2 cm distal to the coronary ostia, and cold blood cardioplegia was achieved through both coronary ostia. The aortic valve was bicuspid, and small granular, gray nodules with calcification were found on the thickened anterior cusp, however, commissural prolapse resulting from extension of the dissection was not observed. The proximal aortic wall was reconstructed with Teflon felt strips. When the rectal temperature reached 20°C, the aortic cross-clamp was released under retrograde cerebral perfusion with hypothermic circulatory arrest. An intimal tear was located in the descending aorta distal to the left subclavian artery. Graft replacement of the ascending aorta and the proximal aortic arch was performed using a 24-mm Dacron graft. Administration of 40 mg/day prednisolone early after the operation was stepwise reduced to 25 mg/day over a period of 20 days. The concentrations of C₃ and C₄ were used as indicators for evaluating the inflammatory response in deciding the amount of prednisolone to administer. The patient recovered uneventfully. A histological study of the aortic wall demonstrated no findings of vasculitis compatible with SLE and no myxomatous degeneration in the media.

DISCUSSION

SLE is a chronic systemic inflammatory disease and affects multiple organs. Since its cardiovascular involvement was first reported in 1924, several cardiovascular abnormalities have been recognized in SLE.⁷⁾ Despite recent improvements in patient survival in SLE, cardiovascular morbidity and mortality have increased.

The most characteristic cardiac lesion is endocardial involvement in the form of non-infective vegetations, as described by Libman and Sacks.⁷⁾ In our patient, typical thickening, nodularity and regurgitation of the aortic valve were found. In a study by Cervera and colleagues,⁸⁾ valvular abnormalities were found in 44% of patients with SLE, and the incidence of valvular lesions is on the increase because of recent advances in echocardiography for valvular disease.

The occurrence of aortic dissection in patients with SLE, however, is extremely rare. To our knowledge, only 14 cases of aortic dissection in SLE have been previously reported, as shown in the Table.^{2,6)} Thus, this is the 15th case described in the literature. Common features in such patients were their young age, presence of systemic disease, and administration of corticosteroid for a relatively long period of time. Systemic hypertension was also found in 13 of the 15 patients.

One of the interesting aspects in our patient is the coexistence of a bicuspid

Table. Reported Cases of Aortic Dissection in Patients with SLE

No.	Author (Year)	Age/Sex of Patients	SLE (years)	Steroid Therapy (years)	Hyper- tension	Surgery	Histological Findings Vasculitis/Degeneration		Pathogenesis of aortic dissection	Outcome
1	Walts (1977)	34/M	22	22	+	?	-	+	atherosclerosis	died
2	Okiye (1983)	33/M	6	6	+	+		No	atherosclerosis?	died
3	Yamakage (1984)	34/F	7	4	+	-		No	atherosclerosis	died
4	Pazirandeh (1988)	38/M	2.5	2.5	+	-	-	+	degeneration	died
5	Yoshimoto (1989)	28/F	11	2	+	-	-	-	atherosclerosis	died
6	Kimura (1989)	38/F	7	7	+	-		No	atherosclerosis?	died
7	Souza (1992)	33/F	3	3	+	+				alive
8	Dugo (1993)	30/F	0.9	0.9	+	-		No	vasculitis?	alive
9	Sclair (1995)	30/F	10	2	+	-		No	-	died
10	Guard (1995)	31/F	9	9	-	-	+	-	vasculitis	died
11	Ohge (1995)	55/F	11	11	?	+	-	-	-	alive
12	Sueda (1996)	52/F	26	21	+	+	-	-	atherosclerosis	alive
13	Lam (1997)	66/F	1	1	+	-	-	-	atherosclerosis	died
14	Motomura (2000)	45/F	10	9	+	+	-	-	-	alive
15	Present case (2002)	27/F	15	15	+	+	-	-	-	alive

?=Pathogenesis of aortic dissection surmised from clinical and laboratory findings.

aortic valve and aortic dissection. Bicuspid aortic valve is considered to be a cause of aortic dissection because of frequent association with medial degeneration of the aortic wall. Pathological investigation of the previous reports, however, demonstrated that medial degeneration was detected in only two patients^{9,10)} and vasculitis related to SLE in only one patient,¹¹⁾ as shown in the Table. Based on these results, many of the previous studies have indicated that atherosclerosis resulting from the side effects of long-term steroid therapy rather than vasculitis related SLE or medial degeneration is possibly the pathogenesis of aortic dissection in patients with SLE. Generally, steroid therapy is certainly effective for controlling inflammatory processes in SLE, however, its adverse effects on systemic diseases are also well known. Long-term steroid therapy increases the incidence and accelerates the severity of hypertension, together with promotion of atherosclerosis.¹²⁾ Furthermore, effects on connective tissue, as well as inhibition of the formation of granulation tissue and chondroitin sulfate have also been recognized.¹³⁾ In our patient, neither medial degeneration nor vasculitis was found in the aortic wall. Considering that systemic inflammation resulting from SLE was relatively well controlled in our patient, the etiology of aortic dissection in SLE is multifactorial. In addition to inflammatory involvement of the aortic wall by SLE, systemic diseases such as systemic hypertension, atherosclerosis, and inhibitive effects on the connective tissue related to relatively long-term steroid therapy probably contribute significantly to the development of aortic dissection in

SLE.

Another interesting finding in this case is that her twin sister had also been diagnosed with SLE. Since aortic dissection developing in twins or in the same family has occasionally been experienced,¹⁴⁾ strict management of the systemic hypertension and careful medical observation are also necessary for her twin sister.

In conclusion, we have described a case of acute type A aortic dissection that developed in a patient with SLE. The occurrence of aortic dissection in patients with SLE is extremely rare, and this is to the best of our knowledge, only the 15th case described in the literature. In addition to inflammatory involvement of the aortic wall by SLE, systemic diseases related to relatively long-term steroid therapy may have promoted the development of aortic dissection in this patient.

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