

## A Case of Abdominal Aortic Aneurysm Associated with Systemic Lupus Erythematosus

Seiji MARUBAYASHI<sup>1)</sup>, Keiji SUGI<sup>1)</sup>, Kouhei ISHIYAMA<sup>1)</sup>, Kazuyuki FUKUMA<sup>1)</sup>, Kazuro OKADA<sup>1)</sup>, Takao HINOI<sup>1)</sup>, Masahiro IKEDA<sup>1)</sup>, Haruki HARUTA<sup>1)</sup>, Keizou SUGINO<sup>1)</sup>, Toshimasa ASAHARA<sup>1)</sup>, Hironori KAWANO<sup>2)</sup> and Kiyohiko DOHI<sup>1)</sup>

1) *The Second Department of Surgery, Hiroshima University School of Medicine, 1-2-3 Kasumi, Minami-ku, Hiroshima 734-8551, Japan*

2) *Department of Hematology and Oncology, Division of Clinical Research, Research Institute for Radiation Biology and Medicine, Hiroshima University, 1-2-3 Kasumi, Minami-ku, Hiroshima 734-8553, Japan*

### ABSTRACT

A case of abdominal aortic aneurysm associated with systemic lupus erythematosus (SLE) is reported. A 45-year-old woman with a 18-year history of SLE was admitted with severe lumbago radiating to the bilateral inguinal region. CT and DSA showed a dumbbell shaped true aneurysm of the abdominal aorta. An aorto-biiliac Y shaped graft replacement was performed. SLE is rarely associated with aneurysm of the great arteries. We could find only 4 reports of abdominal aneurysm associated with SLE. Common features were the young age of the patient, the long term of the systemic disease, and administration of corticosteroid therapy for a relatively long period of time. We speculate that atherosclerosis, hypertension, and corticosteroid may all work in concert, possibly together with aortic wall involvement or vasculitic damage, to produce the rare abdominal aneurysm in SLE.

**Key words:** *Systemic lupus erythematosus, Abdominal aneurysm*

Cardiovascular complications in systemic lupus erythematosus (SLE) have been recognized for many years<sup>4)</sup>. Recently, patient survival in SLE is steadily improving due to antibiotic, immunosuppressive and renal replacement therapy which has markedly decreased mortality from infections and renal failure<sup>2,11)</sup>. However, there has been a parallel increase in cardiovascular morbidity and mortality, which is currently the third cause of death from SLE<sup>15)</sup>. This report, outlining a case of an atypical abdominal aortic aneurysm with SLE, appears to be the fifth reported association of SLE with an atypical non-dissecting aortic aneurysm.

### CASE REPORT

A hypertensive female aged 45 years, with a 18-year history of SLE treated by prednisolone was referred to our university hospital due to pain in the right foot in September 1990. She had a history of hypertension treated by a calcium-blocker. DSA examination showed a sudden obstruction of the posterior tibial artery and a poor blood flow from the foot joint to the peripheral region. These results show that the pain was probably due to poor blood perfusion due to angitis caused by SLE. In November 1995, she visited our hospital com-

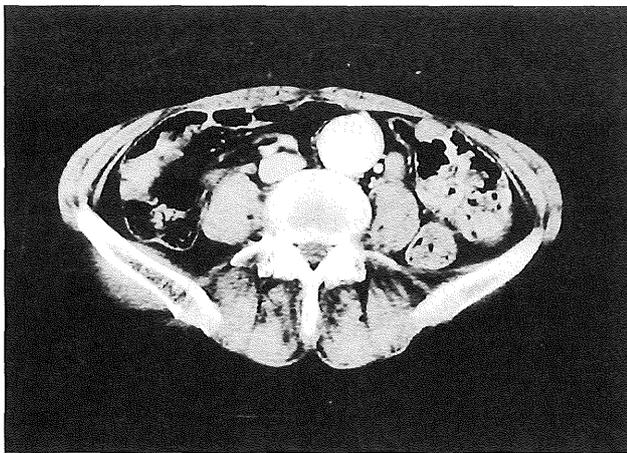
plaining of a periumbilical mass. Abdominal echo and CT examination showed an abdominal aneurysm (size; 3×3 cm). She had no clinical symptoms, so her case was followed by the outpatient clinic. On September 12 1996, she complained of severe lumbago radiating to the bilateral inguinal region. She was admitted to our hospital by ambulance.

On physical examination, she was in slight distress. A Cushingoid habitus was evident. The blood pressure was 180/130 mmHg, heart rate 80/min and regular, and temperature 36.8°C. Head, eye, ear and throat examination showed no abnormalities, and no distension of the neck vein was present. Her breath sound was equal and normal. On auscultation of the heart, a slight systolic murmur was heard at the apex, but no diastolic murmur was heard. The peripheral pulse was symmetric and the remainder of the physical examination was within normal limits.

In the laboratory data, the hemoglobin was 10.8 g/dl with normocytic normochromic anemia, and the hematocrit was 0.31. The white blood cell and platelet count were normal. Elevation of creatinine was observed (Cr; 1.0 mg/dl and CCr; 60 ml/min). She had positive anti-nuclear antibodies

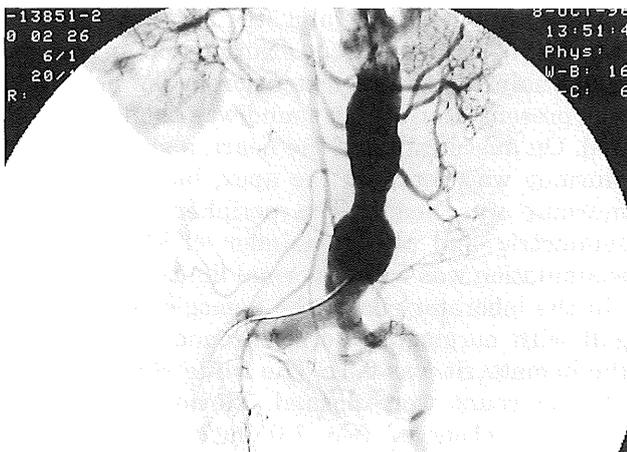
(1:320), but CRP was normal. The values of serum cholesterol, triglycerides, HDL, LDL, VLDL were normal. Other biochemical profiles were within normal limits.

CT and DSA showed a dumbbell-shaped true aneurysm (3.2×2.8 cm and 3.3×2.8 cm) of the infra-renal aorta involving the right common iliac artery (Figs. 1 and 2). Rupture and dissection of the aneurysm was not observed. Electrocardiograph and chest radiography showed no abnormality. Echocardiography showed a trivial mitral valve regurgitation and good left ventricular function. An aorto-biiliac Y-shaped polyester graft replacement was performed. The vascular anastomosis was covered with dacron-felt. A slight atherosclerotic change was observed in the aortic wall (Fig. 3). The postoperative course was uneventful and the patient discharged 4 weeks after the operation. She showed no clinical or radiological evidence of the disease for one year after the operation.



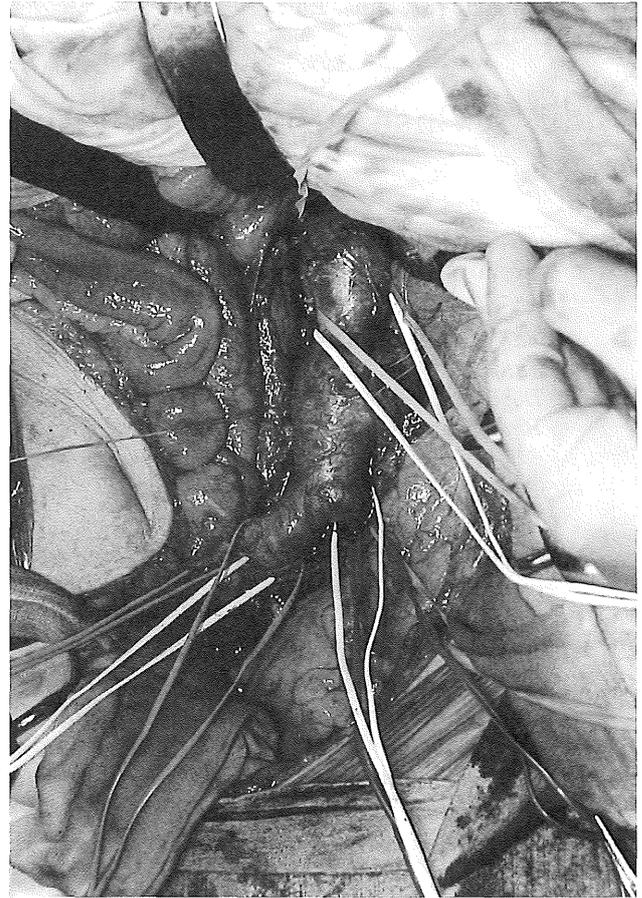
**Fig. 1.** Abdominal CT

Abdominal CT showed true aneurysm of the infra-renal aorta.



**Fig. 2.** DSA examination of abdominal aorta

DSA examination showed a dumb-bell shaped true aneurysm of the infra-renal aorta involving the right common iliac artery.



**Fig. 3.** Operative finding of abdominal aneurysm

The Infra-renal aorta and bilateral common iliac artery was taped. A dumb-bell shaped aneurysm was seen and a slight atherosclerotic change was observed in the aortic wall.

## DISCUSSION

Systemic lupus erythematosus commonly affects the cardiovascular system with hypertension being found in 44% of patients<sup>14)</sup>. The disease is frequently associated with vasculitis, which usually resembles polyarteritis or, less often, Takayasu's arteritis<sup>7)</sup>, Kawasaki disease<sup>8)</sup> or mycotic arteritis<sup>5)</sup>. The aortic and peripheral vascular changes in this case suggest that SLE contributed to the aneurysm formation, although an histologic examination was not performed.

Several cardiovascular abnormalities are recognized in SLE. The most common type of cardiac involvement is effusive pericardial disease, although constrictive pericarditis or cardiac tamponade are distinctly rare<sup>1)</sup>. We could find only 4 reports of abdominal aneurysm<sup>10,12-14)</sup> (Table 1). The average age was 51 years, which is about a decade younger than that of other abdominal aortic aneurysm patients. The average period of steroid therapy was 18.2 years. Among the 7 recorded aortic aneurysms, 2 were pseudoaneurysms; another 5 were true aneurysms. Two patients have died.

**Table 1.** Abdominal aortic aneurysms associated with systemic lupus erythematosus

Author	Age/Sex	Duration of steroid therapy (years)	Status	Etiology	Outcome
Seyama et al <sup>12)</sup>	54/male	1.5	intact	inflammation	discharged
Shibata et al <sup>13)</sup>	30/female	17	intact	inflammation	died due to postoperative bleeding
Sato et al <sup>10)</sup>	45/female	27	pseudoaneurysm	atherosclerosis	discharged
	75/female	32	pseudoaneurysm	atherosclerosis	discharged
	52/female	16	intact	atherosclerosis	died due to ascending aortic dissection
Stehbens et al <sup>14)</sup>	56/male	16	intact	inflammation	discharged
Marubayashi et al	45/female	18	intact	atherosclerosis and inflammation	discharged

One was the result of postoperative bleeding immediately after the operation. The other patient's death was due to an ascending aortic dissection 2 years after the operation. Common features were the young age of the patients with one exception (75 years old), the long term of the systemic disease and administration of corticosteroid therapy for a relatively long period of time.

We believe that the etiology of abdominal aneurysm in SLE is based on various factors. In this collected series, 3 cases of abdominal aneurysm may have been caused by inflammation and another three cases by atherosclerosis. Bulkley and Roberts<sup>1)</sup> found that the long-term steroid therapy in SLE decreased the frequency of certain cardiac lesions as found by autopsy, but accelerated the severity of hypertension, together with promotion of atherosclerosis. The effect of steroids on connective tissue is also well-known, with inhibition of the formation of granuloid tissue and of chondroitin sulfate<sup>4)</sup>. Atherosclerosis, hypertension, and corticosteroids may all work in concert, possibly together with aortic wall involvement or vasculitic damage, to produce the rare abdominal aneurysm in SLE.

(Received January 30, 1998)

(Accepted May 6, 1998)

#### REFERENCES

1. Bulkley, B.H. and Roberts, W.C. 1975. The heart in systemic lupus erythematosus and changes induced in it by corticosteroid therapy: a study of 36 necropsy patients. *Am. J. Med.* **58**: 243-263.
2. Cervera, R., Front, J., Pare, C., Azqueta, M., Perez, V.F., Lopez, S.A. and Ingelmo, M. 1992. Cardiac disease in systemic lupus erythematosus: prospective study of 70 patients. *Ann. Rheum. Dis.* **51**: 156-159.
3. Doherty, N. E. and Siegel, R. J. 1985. Cardiovascular manifestations of systemic lupus erythematosus. *Am. Heart J.* **110**: 1257-1265.
4. Dugo, M., Liessi, G., DeLuca, M., Arduini, R., Susanna, F. and Cascone, C. 1993. Dissection of the thoracic-abdominal aorta in a young adult with systemic lupus erythematosus. *Clin. Nephrol.* **39**: 349-351.
5. Goldman, J.A., Fleischer, A.S., Leifer, W., Parent, A., Schwarzman, S.W. and Raggio, J. 1979. Candida albicans mycotic aneurysm associated with systemic lupus erythematosus. *Neurosurgery* **4**: 325-328.
6. Gross, L. 1940. The cardiac lesion in Libman-Sacks disease with a consideration of its relationship to acute diffuse lupus erythematosus. *Am. J. Pathol.* **16**: 375-407.
7. Igarashi, T., Nagaoka, S., Matsunaga, K., Kaloh, K., Ishigatsumo, Y., Tani, K., Okubo, T. and Lie, J.T. 1989. Aortic syndrome (Takayasu's arteritis) associated with systemic lupus erythematosus. *J. Rheumatol.* **16**: 1579-1583.
8. Laxer, R.M., Cameron, B.J. and Silverman, E.D. 1988. Occurrence of Kawasaki disease and systemic lupus erythematosus in a single patient. *J. Rheumatol.* **15**: 515-516.
9. Parum, D.V. 1990. The spectrum of chronic periaortitis. *Histopathology* **16**: 423-431.
10. Sato, O., Takagi, A., Miyata, T. and Takayama, Y. 1995. Aortic aneurysm in patients with autoimmune disorders treated with corticosteroids. *Eur. J. Endovasc. Surg.* **10**: 366-369.
11. Scair, M., Nassar, H., Bar-Ziv, Y. and Putterman, C. 1995. Dissecting aortic aneurysm in systemic lupus erythematosus. *Lupus* **4**: 71-74.
12. Seyama, K., Mutou, S., Furuya, H., Ohara, T., Orisaka, M., Yamaki, M., Terao, T., Asano, T., Fujisawa, M., Hosoda, S., Katou, M., Miyano, H., Yamaguchi, T., Fukushima, A. and Saitou, K. 1989. A case of upper abdominal aortic aneurysm associated with systemic lupus erythematosus. *Jin to Toseki (Jpn)* **27**: 1139-1114.
13. Shibata, T., Yamada, T., Ishihara, K., Suzuki, N., Eirai, M., Fujii, K., Suehiro, S., Sasaki, Y. and Ueda, M. 1994. A case of abdominal aortic aneurysm associated with systemic lupus erythematosus. *Jpn. J. Cardiovas. Surg.* **23**: 217-220.
14. Stehbens, W.E., Delahunt, B., Shirer, W.C. and Naik, D.K. 1993. Aortic aneurysm in systemic lupus erythematosus. *Histopathology* **22**: 275-277.
15. Wallace, D.J., Podell, T., Weiner, J., Kinenberg, J.R., Forouzes, S. and Dubois, E.L. 1981. Systemic lupus erythematosus: survival patterns. Experience with 600 patients. *JAMA* **245**: 934-938.