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CASE REPORT

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A case of dissecting aortic aneurysm with cystic medial necrosis in systemic lupus erythematosus

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Abstract A 46-year-old woman with systemic lupus erythematosus (SLE) was admitted owing to a sudden severe back pain. She had been receiving corticosteroids throughout the course of long-standing nephritis, and had a family history of dissecting aortic aneurysm. Computed tomography (CT) and magnetic resonance imaging (MRI) examinations of the chest with intravenous contrast revealed a dissecting aortic aneurysm. Surgical replacement of the aortic arch was performed. The histological findings in the thoracic aorta were compatible with cystic medial necrosis. We report this rare case, and review the literature relating to this complication.

Key words Cystic medial necrosis · Familial dissecting aortic aneurysm · Systemic lupus erythematosus (SLE)

Introduction

Dissecting aortic aneurysm in systemic lupus erythematosus (SLE) has been recognized as a rare life-threatening complication, but its pathogenesis is still obscure. Possible mechanisms are thought to be atherosclerosis related to long-standing steroid therapy, or lupus vasculitis. Here we describe the case of a patient who had a family history in which her father had died of a dissecting aneurysm and her brother had died of acute heart failure. There have been only a few reports about familial dissecting aneurysm. Furthermore, familial dissecting aortic aneurysm in SLE has not previously been reported. The present case of dissecting aortic aneurysm with cystic necrosis in SLE is thought to be the first of its kind to be reported.

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Case report

A 46-year-old woman with SLE was hospitalized in April 1997 because of sudden back pain, hypertension, dyspnea, and systolic murmur. The patient had a family history in which her father had died of dissecting aneurysm and her brother had died of acute heart failure. Our patient and her parents showed no physical or laboratory evidence of Marfan's syndrome or Takayasu's disease. SLE had been diagnosed in July 1993 because of the presence of arthritis, fever, pleural effusion, a positive LE test result, lupus nephritis (WHO class III), leucopenia, high titers of antinuclear antibodies (ANA) (shaggy pattern), anti-DNA antibody, hypocomplementemia, and SLE retinopathy without hypertensive change. The patient had than been treated with glucocorticoids (prednisolone 60 mg/day orally). After the proteinuria and retinopathy had improved, her prednisolone dosage was reduced. Cyclophosphamide was started in March 1994 because of a recurrence, and thereafter urine protein was 1 g/day.

At the time of admission on April 27, 1997, a systolic murmur, gallop rhythm, and friction rubs were evident. The patient's body temperature was 36.7°C, her blood pressure was 260/130 mmHg, and her heart rate was 140 beats/min. Other test results were normal. Laboratory investigations showed a WBC count of $9900 \times 10^4/\mu l$ with a left shift, Hb 5.9 g/dl, platelets $19.8 \times 10^4/\mu l$, and C-reactive protein (CRP) 0.6 mg/dl (Table 1). All other blood parameters were within normal limits. Chest X-ray revealed pleural effusion on the left (Fig. 1). Echocardiography showed pericardial effusion.

Chest X-ray on the following day revealed a left hypolucent lung. On the second day, magnetic resonance imaging (MRI) revealed a dissecting aortic aneurysm and hemothorax (Fig. 2). On April 28, the patient underwent an

Table 1. Laboratory data

Peripheral blood	
WBC	9000/μl
RBC	192×10^4/μl
Hb	5.9 g/dl
Ht	19.4%
Platelet	14.1×10^4/μl
Blood chemistry	
Na	132 mEq/l
K	3.6 mEq/l
Cl	97.6 mEq/l
GOT	37 IU/l
GPT	21 IU/l
ChE	2.5 IU/l
γ-GTP	49 IU/l
TP	5.0 g/dl
Albumin	2.1 g/dl
BUN	48 mg/dl
Cr	2.5 mg/dl
CRP	0.6 mg/dl
Urinalysis	
pH	
Protein	5.0 g/dl
Sugar	(2+)
Keton	(−)
Sediment	(−)
RBC	Many
WBC	Many
Arterial blood Gas	
pH	7.509
PCO_2	28.0 mmHg
PO_2	82.8 mmHg
HCO_3	22.3 mEq/l
BE	−0.5 mEq/l

RBC, red blood cell; Hb, hemoglobin; Ht, hematocrit; ChE, cholinesterase; BUN, blood urea nitrogen; CRP, C-reactive protein; WBC, white blood cell; TP, total protein; γ-GTP, γ-glutamyltranspeptidase

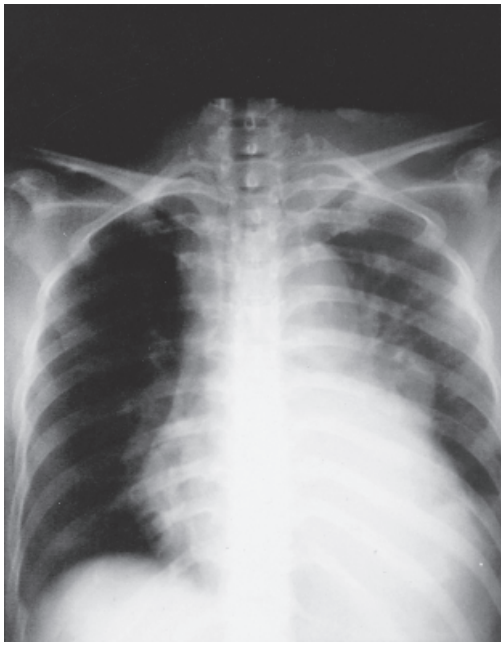

Fig. 1. Chest X-ray on the first day showed pleural effusion on the left and markedly widened mediastinum due to enlargement of the descending aorta

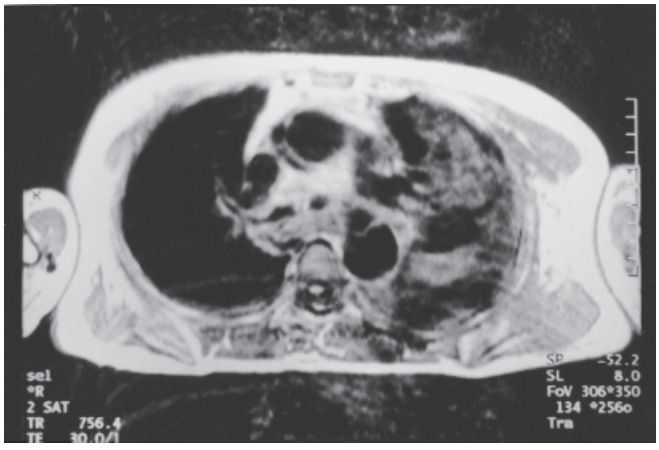

Fig. 2. MRI (T1). On the second day, magnetic resonance imaging (MRI) showed a dissecting aortic aneurysm and a left hemothorax

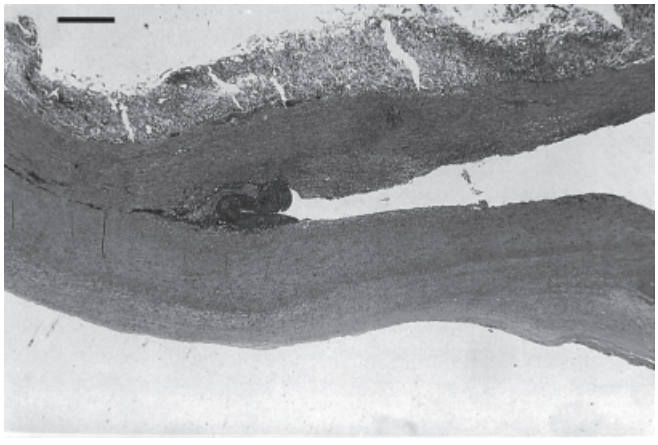

Fig. 3. The ascending aorta was dissected in the media with hemorrhage and thrombus formation (HE staining ×20, *Bar* 1000 μm)

emergency graft replacement of the ascending aorta and the total aortic arch, and had an uneventful postoperative course. Histologically, the aorta was dissected in the media with hemorrhage and thrombus formation (Fig. 3). There was no evidence of vasculitis, and the arteriosclerosis change was mild. Medial elastic fibers were distorted and fragmented, showing mucoid microcystic change of the media consistent with cystic medial necrosis (Fig. 4). There was also fibrinous hemorrhagic pericarditis of both acute and chronic origin. Despite intensive therapy, the patient died of renal failure on July 30.

Discussion

Acute dissecting aortic aneurysm has been recognized as a complication with poor prognosis, occurring in patients with aneurysms, syphilis, Marfan's syndrome, and Ehlers–Danlos syndrome, usually in the presence of hypertension as a background factor.[1]

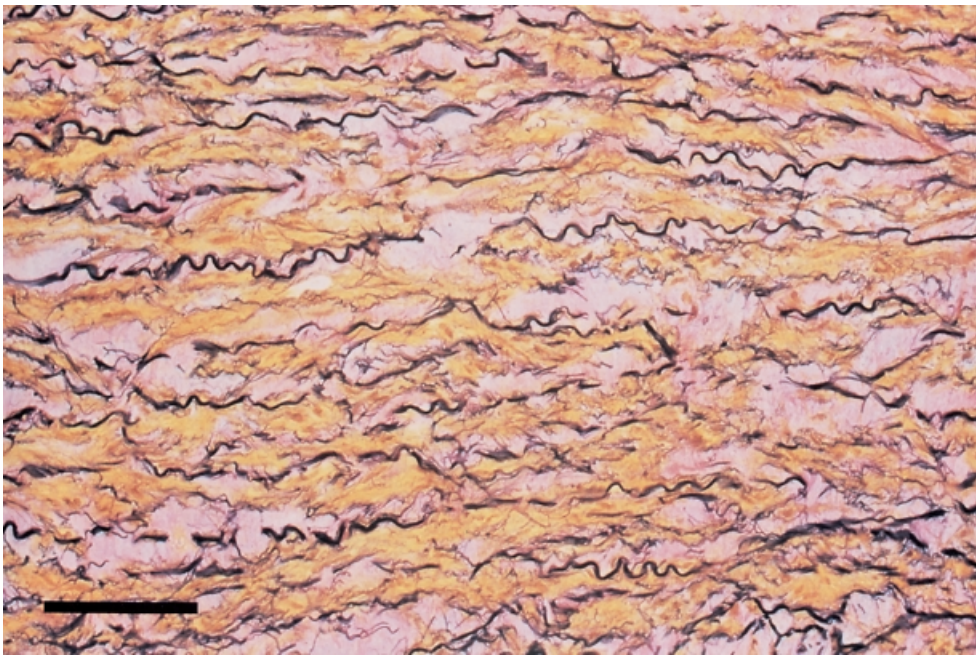

Fig. 4. Elastic fibers of the media were degenerated and destroyed (elastica van Gieson staining ×200, *Bar* 50 μm)

Recently, as the mortality of patients with SLE has improved because of the introduction of glucocorticoid therapy, cardiovascular disease has become a fatal complication of SLE in the long term.[1–4]

Previous reports[3–9] have indicated that the mean age at onset of this complication in SLE patients is 45.4 years old. On average, symptoms develop 13.3 years after primary disease onset. Nephrotic syndrome and acute renal failure are observed in almost all cases.[5–8] One of the reasons is that nephrotic syndrome can accelerate atherosclerosis. Two possible mechanisms are (1) that atherosclerosis could be caused by hypercholesterolemia of nephrotic syndrome, or (2) that hypertension based on renal pathological changes in nephrotic syndrome could promote atherosclerosis.[10,11] Further, hypertension, congestive heart failure, and coronary stenosis are important adverse effects of glucocorticoids in SLE patients.[1] The duration of glucocorticoid treatment plays an important role in the pathogenesis. Bulkey and Roberts[1] reported that high blood pressure, left ventricular hypertrophy, and arteriosclerosis are increased if glucocorticoid therapy is continued for 1 year or more. It is interesting to note that an inflammatory process in the vasa vasorum is often seen,[3,8] but pathological change in the great vessels is rarely a feature of SLE.

In our patient, significant aortic artery hardening could not have occurred over this short period of time on glucocorticoid therapy. In fact, excision specimens of the aortic artery revealed fibrinoid arteritis with slight inflammatory cell infiltration. Neither atheroma denaturation nor calcification consistent with atherosclerosis was seen.

Furthermore, elastica van Gieson staining revealed dissected collagen fibers, and alcian blue-periodic acid staining indicated mucus degeneration. All these findings were compatible with cystic necrosis. Although there are only a few reports giving histological details about SLE with dissecting aortic aneurysm, all of them described the cause of the dissecting aortic aneurysm as atherosclerosis. As the present case showed cystic necrosis with fibrinoid arteritis, it is more likely that the cause of dissection was associated with genetic factors relevant to the patient's family history or to other unknown factors based on lupus vasculitis rather than atherosclerosis.

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