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Aortic aneurysm in systemic lupus erythematosus

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Systemic lupus erythematosus (SLE) is frequently associated with cardiovascular manifestations, but rarely complicated with aortic disease. We report a 28-year-old female patient with a 14-year history of SLE and a 3-year history of hypertension. She had suffered from palpitation and chest tightness for 1 month before admission. Heart echo showed thoracic to low abdominal level with low flow. A computed tomography (CT) scan confirmed aneurysms of the descending thoracic and upper abdominal aorta, down to the renal level. Diagnosis of aortic aneurysm should be considered in patients with SLE, especially those who have a history of hypertension, prolonged steroid use, palpitation and chest pain. Current imaging modalities, such as cardiac echo, CT and magnetic resonance angiography may provide earlier detection of subclinical disease, which may aid in preventing these fatal complications. It is important to control hypertension aggressively in patients with SLE. In addition to decreasing steroid doses, early use of immunosuppressive agents and accurate noninvasive image modalities may allow us to prevent severe damage to the aorta and avoid the fatal complications.

Key words: Aortic aneurysm, magnetic resonance angiography, systemic lupus erythematosus, X-ray computed tomography

Systemic lupus erythematosus (SLE) is a multisystem, autoimmune disease characterized by widespread inflammation of blood vessels and connective tissues. Steroid use, cyclophosphamide therapy and hemodialysis have decreased the mortality rates of SLE. Cardiovascular manifestations frequently complicate SLE [1]; cardiac complications have become significant clinical problems. However, aortic aneurysm is a very rare and life-threatening cardiovascular complication. Aortic aneurysm may be the result of weakness of the elastic lamina and atherosclerosis accelerated by prolonged steroid use, which can result in aneurysmal enlargement, aortic wall involvement and vasculitic damage [2]. We present a case of descending aortic aneurysm in a young woman with a 14-year history of SLE and corticosteroid treatment, using computed tomography (CT) for early evaluation and diagnosis.

Case Report

A 28-year-old female with SLE had complications of lupus nephritis, chronic renal failure under hemodialysis 3 times a week, and avascular necrosis of the bilateral hip joint s/p total knee replacement. She was diagnosed as having SLE in the presence of the following: serositis, renal involvement, anemia, thrombocytopenia associated with a high titer of antinuclear antibody, anti-DNA antibodies and hypocomplement. She had an initial presentation of a progressively puffy face and lower leg edema at 14 years old. She received methylprednisolone pulse therapy once and cyclophosphamide pulse therapy 7 times.

She was followed regularly at our immunology clinic. Nifedipine and propranolol were used for hypertension control. However, dizziness, occasional chest tightness and palpitation, and cold sweating were noted in the past month. Frequent hypotension was also noted by the patient. She returned to our hospital on April 26, 2002 for complaints of discomfort and, due to the symptoms described, she was admitted for further evaluation.

Laboratory investigations showed a white blood cell count at 6260/mm^3, hemoglobin 10.5 g/dL, haematocrit 32.3%, and platelet count 183,000/mm^3. Blood urea nitrogen was 113 mg/dL, creatinine 9.28 mg/dL, cholesterol 266 mg/dL, triglyceride 300 mg/dL, aspartate aminotransferase 13 U/L, creatine phosphokinase (CK) <20 U/L, CK-MB 2.6 U/L, troponin I <2 ng/mL, C3 129 mg/dL, and C4 49.5 mg/dL. A previous kidney biopsy revealed lupus nephritis, mixed membranous and membranous, and mesangial type. A chest radiograph revealed tortuous aorta and aneurysm with calcification,

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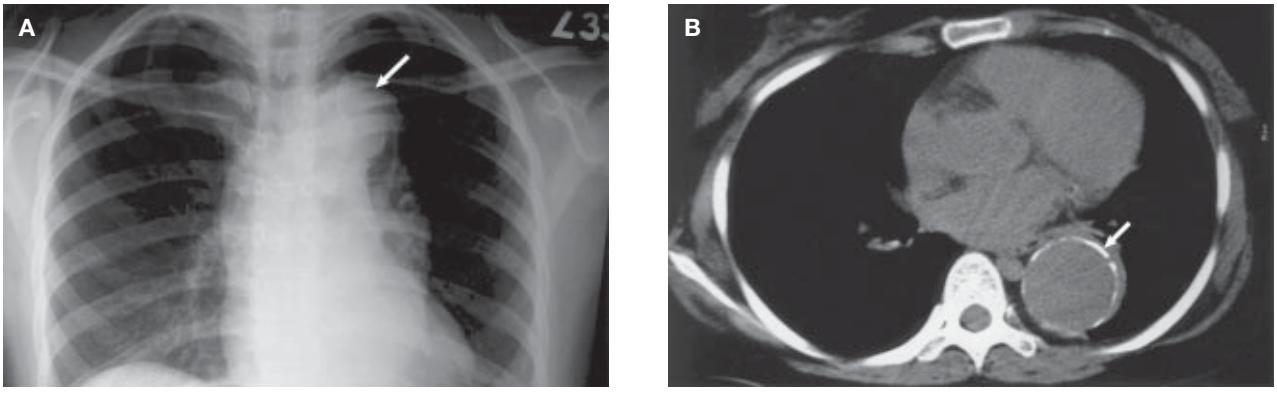

Fig. 1. A) Posteroanterior view of the chest X-ray film shows tortuous descending aorta (arrow). B) Computed tomography scan showing the aortic aneurysm in the descending aorta (arrow).

which had not been previously noted (Fig. 1A). A CT scan of the chest and abdomen showed fusiform dilation of the descending thoracic and upper abdominal aorta, down to the renal level (Fig. 1B). There was calcification along this aorta. Mural thrombosis was also found at the upper abdominal aorta. Aortic aneurysm was diagnosed. A cardiac surgeon was consulted, and because the size of the aneurysm was 4 cm, operation was not necessary at that time. She was then discharged and followed up in our clinic. In March 2003, surgical treatment was proposed, but the patient refused. She had no complaints and continued on antihypertensive, antianginal and antiplatelet treatment.

Discussion

The mortality rate of patients with SLE due to infection and renal failure has decreased after administration of antibiotics, immunosuppressive therapy and renal transplantation. Immunosuppressive agents, such as azathioprine, cyclophosphamide, methotrexate, and cyclosporine can be used to treat lupus nephritis and decrease morbidity of renal failure. However, there has been an increase in cardiovascular morbidity and mortality. Cardiovascular abnormalities in patients with SLE are frequent. Aortic aneurysm with or without dissection is one of the rare complications in patients with SLE. Common treatments for patients with aneurysm have included administration of a large dose, long-term corticosteroid therapy [3].

Our patient had received steroid therapy for at least 3 years, methylprednisolone pulse therapy once and cyclophosphamide pulse therapy 7 times. She had bilateral femoral avascular necrosis induced by steroids. Furthermore, she had suffered from hypertension and chronic renal failure under hemodialysis since 1999. Hyperlipidemia (cholesterol 266 mg/dL, triglyceride 300 mg/dL) was observed, and hypertension was poorly controlled during this period. The mechanisms of aortic aneurysm formation were not clear. However, prolonged steroid therapy may have played a major role in accelerating atherosclerosis, which can result in aortic aneurysmal enlargement, possibly together with primary aortic wall involvement and vasculitic damage in patients with SLE [2]. The patient refused surgical approaches, barring us from obtaining pathologic tissue proof.

It is widely understood that when aortic aneurysm is less than 5 cm in diameter, the risk of rupture is considerably lower compared to aneurysms of more than 5 cm [4]. In fact, the diameters of dissected aortic aneurysms reported in patients with SLE have been between 3 and 6 cm [5,6]. In our case, the cross-section of aortic aneurysm size was 4 cm. The patient hesitated to receive surgical correction. Up to now she has refused to take the risk of surgical operation. However, a surgical correction should be performed earlier in SLE patients than in patients without systemic disease. Some successful cases about surgery have been reported in such conditions [7].

Our patient suffered from cold sweating, palpitation, dizziness and chest tightness for 1 month. Aortic aneurysm was highly suspected after tracing her steroid use and hypertension history. However, the most common arteritic lesions complicating SLE are small vessel vasculitides and coronary arteritis. Coronary artery disease has also emerged as a prominent cause of death in young patients with SLE [8]. Her risk factors for coronary arteritis included hypertension, hyperlipidemia, a history of nephrotic syndrome and other organ system involvement, and a long history of prednisolone use [9]. Therefore, coronary arteritis could not be ruled out. Laboratory data showed CK <20 U/L,

CK-MB 2.6 U/L, troponin I <2 ng/mL, and aspartate aminotransferase 13 U/L. The above data was within normal ranges, and the myocardium did not seem to be involved. Cardiac echo revealed mild mitral regurgitation, aortic regurgitation, tricuspid regurgitation and thoracic to low abdominal level with low flow. However, we confirmed the diagnosis by CT. Sometimes CT with contrast studies may be contraindicated in SLE patients in the presence of renal failure. Transesophageal echocardiography and magnetic resonance angiography (MRA) are further options for early and noninvasive evaluation [10]. Often the initial presentation of aortic aneurysm is back pain or abdominal pain. The patient may present with varying quadrant pain depending on the location of aortic aneurysm. Back pain may be induced by aneurismal dilation of the ascending aorta, and abdominal pain may contribute to abdominal aortic aneurysm [11,12].

If a patient with SLE is suspected of having aortic aneurysm, aortic dissection, transesophageal echocardiography, CT and MRA are fast, safe, non-invasive and highly accurate for diagnosis [10]. They become useful in the evaluation and diagnosis of acute dissection of the aorta. Aortic dissection is a life-threatening disorder with high mortality rates; it requires prompt diagnosis and treatment to prevent death and to prolong survival.

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