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CASE REPORT

Endovascular stent-graft placement for ruptured dissecting aortic aneurysm in an adolescent patient with systemic lupus erythematosus: case report

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Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune connective tissue disease with multi-organ involvement that is associated with the presence of autoantibodies, and it is influenced by genetic, endocrine, or environmental backgrounds such as exposure to ultraviolet rays and viral infections for the etiopathogenesis [1]. In the cardiovascular system, it frequently causes pericarditis, nonbacterial verrucous endocarditis, coronary artery disease, and cardiomyopathy [2]. Cardiovascular complications in SLE are generally presumed to be associated with inflammatory activity as well as premature development of atherosclerosis [1]. The overall morbidity and death from these complications have been increasing in recent years, and they are now the major cause of death in SLE [3].

Dissecting aortic aneurysm in SLE has been recognized as a rare life-threatening complication, but its pathogenesis is still obscure. The pathogenesis of aortic aneurysms and dissection in SLE has been attributed to aortic circulatory disturbances resulting from vasculitis and cystic medial degeneration as well as to atherosclerosis. The occurrence of atherosclerosis is considered to be due to long-term steroid treatment, hyperlipidemia, hypertension, and nephrotic syndrome [4].

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The importance of the high prevalence of premature atherosclerosis and cardiovascular disease in SLE should be addressed, even in young adolescents, and especially those with early childhood-onset disease. Nevertheless, aortic dissection is a life-threatening disorder that requires immediate diagnosis and treatment to prevent mortality. To our knowledge, this is the second adolescent patient with childhood-onset SLE complicated with aortic dissection, but the first case treated with endovascular stent-grafting.

Case report

A 17-year-old female patient was admitted to our hospital complaining of abdominal pain. She had been diagnosed with SLE 8 years earlier at the age of 9. SLE diagnosis was made based upon oral ulcers, photosensitivity, malar rash, arthritis, pancytopenia, mild pericardial effusion, positive antinuclear antibodies, and elevated anti-dsDNA antibodies. She was maintained on immunosuppressive therapy (prednisolone 30 mg/day and mycophonelate mofetil 1 g/day), antihypertensive therapy (amlodipine 10 mg/day, doxazosin 4 mg/day, minoxidil 5 mg/day, and propranolol 40 mg/day), antithrombotic therapy (acetylsalicylic acid 100 mg/day), and antilipidemic therapy (atorvastatin 10 mg/day). She suffered from chronic renal failure requiring dialysis, and she was also maintained on antacids 2 g/day, active vitamin D 0.5 μg/day, and erythropoietin 3,000 IU/week medication.

On admission, the patient had no fever or complaints of arthralgia besides abdominal pain. She was 151 cm and weighed 41 kg. Her blood pressure (BP) was 150/90 mmHg, and heart rate was 96/min with a regular rhythm. Admission laboratory testing revealed hemoglobin, 7.3 g/dl; white blood cell count, 14,700/mm^3; and total protein and albumin, 5.7 and 2.6 mg/dL, respectively. Electrolytes, blood glucose,

AST, ALT, GGT, amylase, cholesterol, and triglyceride levels were normal. Blood urea nitrogen (89 mg/dL) and serum creatinine (4.9 mg/dL) were elevated. Erythrocyte sedimentation rate (93 mm/h) and C-reactive peptide (37.5 mg/dL) values were higher than normal. C3, C4, anti-dsDNA, and antiphospholipid values were normal. The physical examination disclosed local tenderness restricted to the epigastric area with no peritoneal signs, and other physical findings including abdomen were normal. Direct abdominal radiography was normal. One hour later after admission, abdominal pain had started again but very severely, this time with accompanying back pain. On auscultation, there was a systolic murmur in umbilical region above the umbilicus as a new finding. Aortic dissection was considered, and cardiac echocardiography including aorta was performed. A dissected flap was seen in descending aorta with intact mitral valve. The medical condition was considered as a medical emergency; therefore, we decided to perform a contrast-enhanced computed tomography (CT) examination. Isotonic nonionic contrast agent, iodixanol (Visipaque™-270, GE Healthcare, Inc., Princeton, NJ, USA) was administered. Hemodialysis was planned immediately after the CT examination with the suggestion of consultant nephrologist. Contrast-enhanced 16-multidetector computed tomography (16-MDCT) revealed a dilated thoracoabdominal aorta (4 cm in diameter) and a dissection (Stanford type B) extending from the descending aorta to the origin of the celiac truncus (Fig. 1). There was also hematoma surrounding the descending thoracic aorta.

Emergency digital subtraction angiography was performed revealing a dissecting aneurysm of the thoracoabdominal aorta (Fig. 2a). A decision was made with the patient and her family to perform immediate stent-grafting of the thoracoabdominal aorta as an emergency. Preoperative prophylactic cerebrospinal fluid drain was placed to prevent spinal cord ischemia. Under general anesthesia, the right common femoral artery was exposed and cannulated. A 24×100-mm Medtronic Valiant thoracic stent-graft (Medtronic, Santa Rosa, CA, USA) was deployed from distal thoracic aorta to the level of the origin of celiac truncus. Control aortogram revealed a distal type I endoleak, but no additional endovascular treatment was considered (Fig. 2b, c). Coronal reconstructed multidetector CT image is provided in Fig. 2d.

After the endovascular procedure, the clinical course was uneventful, and the patient was discharged on the fifth postoperative day. On regular follow-ups, her blood pressure is under control with antihypertensive agents. She is taking the immunosuppressive and antihypertensive drugs and having hemodialysis three times a week. Follow-up CT scan after 6 months showed clot formation in the false lumina, and no progression of aortic dissection is observed.

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 [5]. Childhood onset SLE is observed in the 15–20% of the whole lupus patient population [6]. Cardiac manifestations in children with SLE

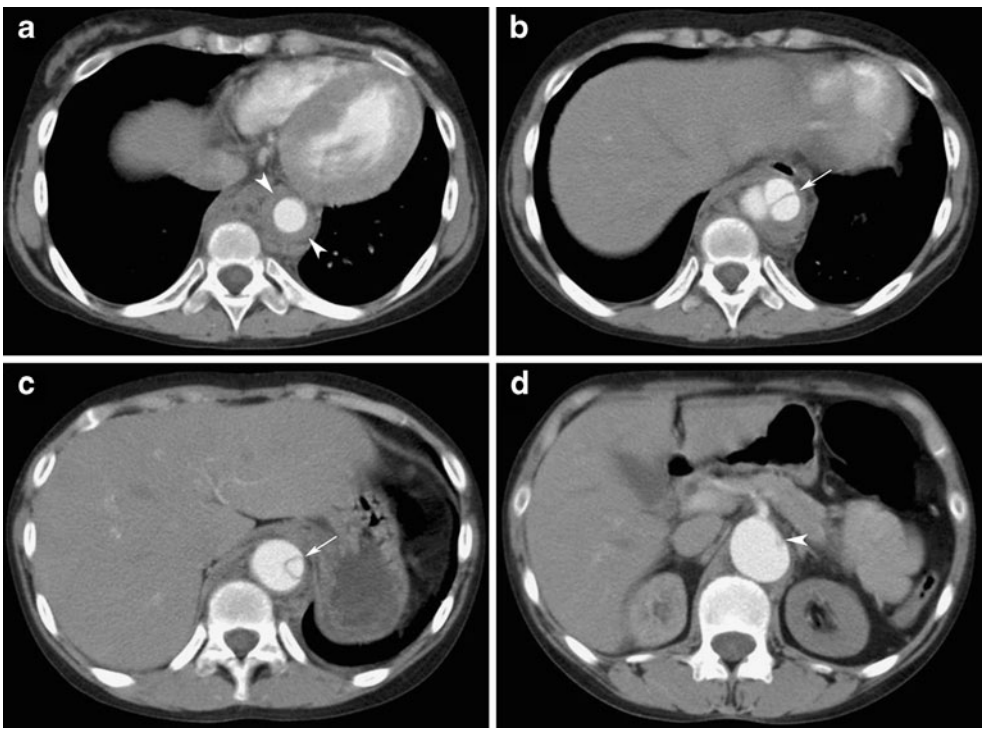

Fig. 1 Axial contrast-enhanced CT image through the thorax (**a**) shows hematoma (*arrowheads*) surrounding the descending thoracic aorta. Axial CT images through the lower thorax and upper abdomen (**b**, **c**) obtained at different levels show intimal flap (*arrows*) involvement of the thoracoabdominal aorta. Note the extension of the dissection flap (*arrowhead*) up to the origin of the celiac trunk from the abdominal aorta (**d**)

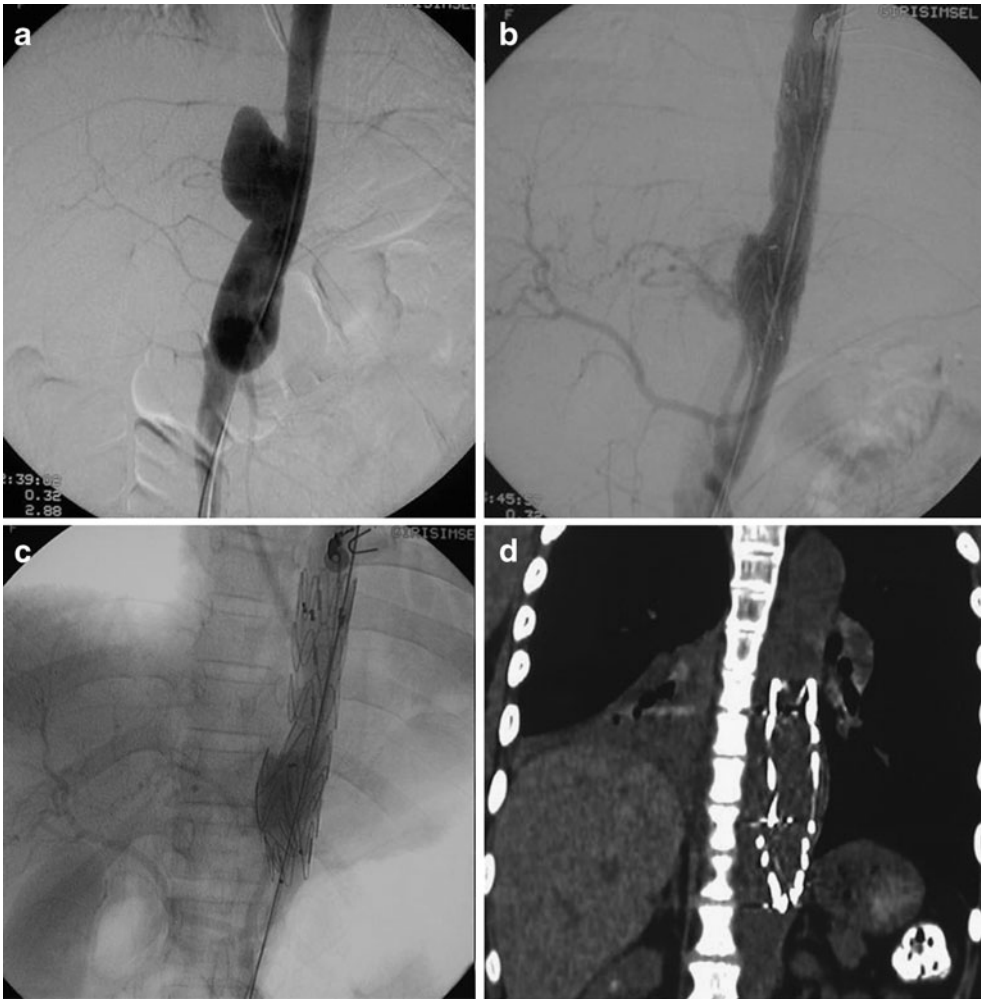

Fig. 2 Preprocedural digital subtraction angiography (**a**) shows dissecting aneurysm (Stanford type B) of the thoracoabdominal aorta. An aortogram after deployment of a stent-graft (**b**, **c**) revealed a distal type I endoleak, but no additional endovascular treatment was considered. Coronal reconstructed CT image 1 month after stent-graft deployment (**d**)

were reported in a wide range of 25–60% at any time of the disease [7], and the most common is pericarditis with pericardial effusion [8]. Myocardiopathy is also seen frequently. Valve pathology, arrhythmia, and conduction anomalies, respectively, are rare [9]. Despite recent improvements in patient survival in SLE, cardiovascular morbidity and mortality have been increased.

Aortic dissection is rare in children and young adults, occurring most commonly in the fifth to seventh decade of life. The known incidence is five to ten cases per million of the population [10]. Marfan syndrome is the most common inherited connective disorder in which an abnormality of fibrillin leads to progressive weakness of the aortic wall. Other implicated disorders of connective tissue include the Ehlers–Danlos syndrome, osteogenesis imperfecta, and Turner's and Noonan's syndromes. Patients with inflammatory processes such as Takayasu's arteritis and giant cell arteritis have also been reported with aortic dissection [11].

Aortic dissection in SLE is very rare and is often more common in patients with hypertension and during treatment with steroids [4]. The dissecting aneurysm in SLE may be due to many factors like aortitis, obliterative endarteritis of vasa vasorum of the aorta, mucoid degeneration, hypertension, atherosclerosis, and treatment with steroids, and all may work in concert to produce aortic aneurysmal dilatation with or without dissection in SLE. The vasculitis involving the vasa vasorum of the aorta leads to aneurysm formation. Antiphospholipid antibodies associated with SLE may predispose to premature atherosclerosis. The corticosteroid therapy may inhibit the formation of granulation tissue, chondroitin sulfate, interfere with the integrity of the connective tissue, and may contribute to aneurysmal enlargement by inhibiting repair of arterial injury. Primary proximal aortic wall involvement or vasculitic damage may be the initiating event for aortic complication in SLE [12]. Kurata et al. suggested that aortic aneurysms in patients with SLE may be classified into two principal patterns: one was the fatal nonatherosclerotic thoracic aneurysm which was associated with cystic medial degeneration and probably due to vasculitis [13]. The other was abdominal aneurysm, which was resulted from atherosclerosis due to prolonged steroid therapy, and this pattern was also associated with a relatively favorable prognosis. A similar proposal is presented in general patients besides SLE in

another literature, stating that thoracic aortic aneurysms are associated with medial degeneration, while abdominal aneurysms are associated with atherosclerosis [14].

In a recent case report and review of the literature, Wei et al. have reported that in English literature, there are only 21 cases of aortic dissection in SLE, and their patient was the first adolescent patient with this complication [15]. Our case is the second adolescent patient with childhood-onset SLE complicated with aortic dissection; however, this is the first published case treated with endograft stenting. Aortic dissection is a medical emergency and can quickly lead to death, even with optimal treatment because there are published reports in the literature about using stent-graft in neonatal, pediatric, and adolescent cases [16]. If the dissection tears the aorta completely, open, massive, and rapid blood loss occurs. Management of adolescent patients with aortic pathologies, as in adults, should focus on the size of aneurysm, caliber of the vessel, and location of the pathology when deciding to repair the vessel with endovascular techniques. This emphasizes the importance of coordinating general and vascular surgical services as well as the importance of stent choice and delivery mechanisms. These concerns are usually not as significant in the adult population, but present significant problems in the pediatric or adolescent population.

It is important to identify SLE patients at high risk of cardiovascular disease, even young adolescents. Atherosclerosis, cystic medial degeneration, vasculitis, and systemic hypertension play a key role in the pathophysiology of aortic dissection. Aortic dissection should be considered in patients with SLE who develop unexplained sharp abdominal, chest, or back pain, especially those who have a history of hypertension or prolonged steroid use. The main challenge in managing acute aortic dissection is to suspect and diagnose the disease as early as possible. Because of its rarity in the younger population, prompt diagnosis is often delayed. The outcome can then be catastrophic. As a conclusion, damage of the vessel wall related to SLE causing aortic dissection could be observed, even in young patients. Nonetheless, prompt diagnosis and endovascular stent-grafting could be life saving.

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