

Aorta coarctation and systemic lupus erythematosus

A case report

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Abstract

Rationale: Vasculitis is one of the common pathological hallmarks of systemic lupus erythematosus (SLE). Vascular lesions in SLE commonly involve medium- and small-sized vessels. Rarely, vasculitis in SLE may involve large vessels such as the aorta leading to life-threatening complications. Reported cases of large vessel lesions in SLE included aortic aneurysm and aortic dissection.

Patient concerns: Here, we report a 52-year-old Chinese woman with SLE, who was stable on oral glucocorticoid, but showed sudden intractable hypertension and heavy proteinuria before we found aorta coarctation in her computed tomography (CT) scan of the aorta.

Diagnoses: This patient's large vascular lesions were likely secondary and not a primary manifestation of lupus activity.

Interventions and outcomes: After endovascular stent graft repair of the abdominal aorta, her hypertension and proteinuria were controlled.

Lessons: In the context of reported cases of large vessel lesions in SLE, our case further supports the significance of having a wide differential for vascular lesions in SLE, especially when an SLE patient presents sudden hypertension and heavy proteinuria. This case also demonstrates that vascular lesions in SLE may lead to serious, potentially fatal consequences.

Abbreviations: ACR = American College of Rheumatology, AoA = aortic atherosclerosis, BP = blood pressure, CT = computed tomography, DBP = diastolic blood pressure, ESR = erythrocyte sedimentation rate, MMF = mycophenolate mofetil, SBP = systolic blood pressure, SLE = systemic lupus erythematosus, SLEDAI = SLE disease activity index.

Keywords: aorta coarctation, hypertension, systemic lupus erythematosus, vascular lesions

1. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease with multi-organ involvement. Specifically, patients with

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SLE have an increased risk of aortic lesions including aneurysm and dissection. SLE patients were found to have increased risk of aortic lesions compared with age- and sex-matched controls, and the incidence of aortic lesions in SLE patients was estimated at 4.26 per 10,000 person-years in on population-level study.^[1,2] However, coarctation of the aorta has not been reported in patients with SLE. The present case describes a 52-year-old Chinese woman with aorta coarctation secondary to SLE. We use this case to emphasize the seriousness of visceral vascular lesions in SLE, especially when SLE patients present with sudden-onset hypertension and heavy proteinuria.

2. Case report

A 52-year-old woman, with a 7-year history of recurrent multi-joint pain, alar rash, and dental ulcers was diagnosed with SLE in 2007 according to the 1997 American College of Rheumatology (ACR) Classification Criteria for SLE, with evident low complement, positive poly-autoantibodies (anti-nuclear, anti-dsDNA, anti-smith, anti-SSA, and anti-SSB). She had no history of proteinuria, and her family history was negative for autoimmune diseases. Her SLE has been stable and controlled by oral glucocorticoid that was gradually tapered from 45 to 10 mg/d. She was not treated with additional immunosuppressive agents. In August 2015, she progressively developed generalized edema and proteinuria (1 g/24 h, 1200 mL). She received oral corticosteroid (30 mg/d) and mycophenolate mofetil (MMF, 1.5 g/d) for 3 months, but she had no improvement in her proteinuria or edema. On January 4, 2016, she was admitted

to our department with hypertension. On physical examination, hand deformities were noted including swan neck deformity of multiple digits, Z deformity of the thumb, and ulnar deviation of metacarpophalangeal joints. The blood pressure (BP) of the upper limbs (left: 176/92 mmHg, right: 172/88 mmHg) was higher than that of the lower limbs (left: 130/64 mmHg, right: 128/60 mmHg). Dorsalis pedis artery pulse was detected. She had no audible murmurs. A spectrum of SLE antibodies was tested and showed the presence of anti-nuclear antibodies, anti-dsDNA, anti-Ro52, anti-SSA, anti-nucleosome, and anti-histone antibodies. Laboratory testing showed high 24 hours urinary protein content (1.95 g/24 h, 1280 mL), low complement (C4 0.054 g/L; C3 0.34 g/L), and high cholesterol and triglyceride levels. Other pertinent laboratory results included creatinine, C-reactive protein, and erythrocyte sedimentation rate (ESR) that were all within normal range. Anti-cardiolipin antibodies were negative. Conventional radiographic imaging showed deformities in the fingers without bone erosions. Carotid ultrasound did not show intimal-medial thickening or plaques bilaterally. She was stable according to SLE disease activity index (SLEDAI) score with 4 points.

The patient continued to have uncontrolled hypertension despite treatment with a dihydropyridine calcium channel blocker (Levamlodipine 2.5 mg bid) and an adrenergic β blocker (Arotinolol 10 mg bid). During her admission, 24-hour dynamic blood pressure monitoring showed that the systolic blood pressure (SBP) ranged from 156 to 174 mmHg, and the diastolic blood pressure (DBP) ranged from 90 to 103 mmHg. Renal artery ultrasound revealed reduced diameter both renal arteries at their origin from the abdominal aorta, plaque formation within different sites of the abdominal aorta, local luminal stenosis, and distal dilation of the abdominal aorta. Follow-up computed tomography (CT)-scan of the aorta showed highly stenotic aortic segment, annular thickening of the aortic walls, abdominal aortic calcification, and penetrating atherosclerotic ulcer (Fig. 1A).

Following diagnosis with coarctation of aorta, she was offered surgical treatment. Endovascular stent graft repair of the abdominal aorta was done on January 26, 2016. Following her surgery, her blood pressure was within normal limits, and she did not require anti-hypertension agents. At 3-month follow-up visit, her proteinuria was reduced to 0.5 g/24 h, and her blood pressure was within normal limits. She was started on a glucocorticoid taper from 30 to 10 mg/d and discontinued MMF treatment. CT-scan of the aorta showed dilated aorta and no new aortic stenosis or mural thrombus (Fig. 1B).

3. Discussion

The presented middle-aged woman had a confirmed diagnosis of SLE as evident by the presence of Jaccoud Arthropathy, oral ulcer, cutaneous lupus, and immunologic abnormalities according to the 1997 ACR Classification Criteria for SLE. She has a long history of glucocorticoid use to control her symptoms, and her disease was stable without immunosuppressive agents. She presented with acute-onset intractable hypertension, proteinuria and vascular lesions mainly shown as aortic stenosis and arteriosclerosis. Based on her history and presentation, our differential diagnosis included 3 main conditions: primary vascular lesions, hypertension secondary to lupus activity, or secondary vascular lesions of SLE. Congenital aortic stenosis includes a series of stenotic lesions starting in the anatomic left ventricular outflow tract and stretches toward the ascending aorta. The vascular lesions in this patient were located in the abdominal aorta, an uncommon location for congenital lesions. The serum levels of inflammatory indicators were negative, and her SLE was inactive according to the SLEDAI score. Thus, primary vascular lesions and lupus activity were unlikely, and her vascular lesions are most likely secondary to or associated with her SLE.

SLE is a systemic autoimmune disease mainly caused by immune-complex induced inflammation which can potentially affect any organ system during the course of the disease.^[3] Vascular lesions are one typical characteristic of SLE. Vascular



Figure 1. (A) CT scan of the aorta before the surgery showing abdominal aorta coarctation. (B) CT scan of the aorta after the surgery showing dilated aorta. CT=computed tomography.

lesions in SLE are usually seen in medium- and small-sized vessels such as cutaneous vessels, renal glomeruli, coronary and cerebral vessels, and lung alveoli, while they are rarely reported in large vessels such as the aorta. Clinical features of vascular injury in SLE depend on the involved vessels. Life threatening injuries may result from lesions of visceral vessels such as the aorta.^[4] Vascular injuries in SLE are mainly immune-system related and involve the activation and dysfunction of endothelial cells. Appel et al^[5] provided a pathological classification for lupus vascular lesions, including uncomplicated vascular immune complex deposits, non-inflammatory necrotic vasculopathy, thrombotic microangiopathy, true vasculitis, arteriosclerosis, and arteriolosclerosis. These classes of lesions may occur alone or concurrently in the same patient.^[5] In a meta-analysis of 35 cases of vascular lesions in lupus, 2 principal patterns of aortic pathology were observed, thoracic aneurysms associated with cystic medial degeneration likely due to vasculitis, and abdominal aneurysms and plaques associated with the duration of steroid therapy and atherosclerosis.^[6] These pathological phenomena may contribute to the lesions observed in this patient, especially arteriosclerosis. SLE duration, activity and damage, corticosteroid treatment, metabolic syndrome, chronic kidney diseases and age at SLE diagnosis are independent predictors of aortic atherosclerosis (AoA) besides traditional atherogenic factors.^[7] Aggressive non-steroidal immunosuppressive therapy to control the inflammation in SLE patients may be needed, and aggressive interventions to control the atherogenic risk factors for AoA are needed for lupus vascular lesions.^[8] Surgical interventions in combination with immunosuppressive agents may be useful for lupus vascular lesions such as aortic stenosis, aneurysm, or dissection. When SLE patients present with sudden-onset hypertension, the possibility of visceral vascular lesions should be ruled out.

Author contributions

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