**Abstract:**
Lupus aortitis is a rare and potentially life-threatening disorder. Previous studies have reported the utility of high-dose systemic glucocorticoids or surgery as the treatment, although there have been no related controlled trials. We herein report a 49-year-old woman with a 35-year history of systemic lupus erythematosus (SLE) who was diagnosed with aortitis. Her symptoms and laboratory and imaging abnormalities rapidly resolved upon the administration of moderate-dose glucocorticoids. We subsequently performed a literature review of similar cases to identify the appropriate treatment and discuss these cases. A study of further cases will be needed to identify the characteristics of patients who would benefit from moderate-dose glucocorticoid therapy.

**Key words:** systemic lupus erythematosus, lupus aortitis, moderate-dose glucocorticoid therapy

---

**Introduction**
Systemic lupus erythematosus (SLE) is an autoimmune disorder associated with multisystem organ damage mediated by autoantibodies and immune complexes. Aortitis is an uncommon complication of SLE (1-27). Aortic dissection and aortic aneurysmal rupture are potentially fatal complications of lupus-associated aortitis (lupus aortitis) (7-9, 12-17, 22, 24).

Many previous reports on lupus aortitis have stated the need for therapy with high-dose systemic glucocorticoids or surgery (5-10, 12-15, 17-27). However, glucocorticoid therapy is associated with both accelerated atherosclerosis, which causes cardiovascular events, and aortic aneurysmal enlargement (28-31). It is therefore important to limit the exposure to glucocorticoids, particularly in patients with aortic aneurysms. However, the appropriate amount and duration of glucocorticoid therapy for lupus aortitis remains unclear because of a lack of controlled trials.

We herein report a case of lupus aortitis that was successfully treated with moderate-dose glucocorticoids. The current report is significant because there are no previous reports in which remission was successfully induced by conservative therapy with moderate doses of glucocorticoids.

**Case Report**
A 49-year-old woman was admitted with a 1-week history of bilateral shoulder pain that migrated to the precordium and a 2-day history of a fever and dyspnea. Although the fever was resolved with oral loxoprofen, her chest pain remained.

She had been diagnosed with SLE at 14 years of age, after she presented with facial erythema, photosensitivity, and hair loss as well as laboratory results of positive antinuclear antibodies, positive anti-double stranded deoxyribonucleic acid (anti-dsDNA) antibodies, and hypocomplementemia. She subsequently developed both pericarditis and pleurisy several times. These conditions were resolved with prednisolone (PSL) at a dose of about 0.5 mg/kg/day. During all of her previous episodes, she had experienced chest pain that...
Figure 1. (a-d) Contrast-enhanced CT on admission showed abnormal thickening and enhancement of the aortic wall from the ascending aorta to the arch.
was exacerbated by movement and breathing, along with a fever, and elevated anti-dsDNA antibodies and elevated C-reactive protein (CRP) levels were seen. In addition, the episodes of pleurisy were accompanied by pleural effusion. PSL therapy was gradually reduced to 8 mg/day orally, and she visited the hospital regularly for observation while maintaining this dose for 29 months. She had also been diagnosed with Sjögren’s syndrome based on her dry mouth and positive findings for anti-Ro/SSA and anti-La/SSB antibodies.

She was completely alert on the day of admission, and her vital signs were as follows: blood pressure, 114/75 mmHg; pulse rate, 82 beats per minute; body temperature, 36.0°C; respiratory rate, 16 breaths per minute; and peripheral capillary oxygen saturation level on ambient air (SpO₂) of 98%. A physical examination showed a height of 156 cm and weight of 56.6 kg. Cardiovascular, respiratory, and abdominal examinations were normal, although she complained of tenderness over the sternum.

A laboratory examination revealed a white blood cell count of 5,860/μL, hemoglobin of 9.6 g/dL, platelet count of 369,000/mm³, CRP of 9.4 mg/dL, erythrocyte sedimentation rate of 89 mm/h, and anti-dsDNA antibodies of 13 U/mL (Table 1). She had presented with iron deficiency anemia for five years. Anti-dsDNA antibodies had been positive for 13 years and increased to more than 50 U/mL when she developed both pericarditis and pleurisy. Complement levels, creatinine, and a urinalysis showed no abnormalities.

Figure 2. (a-d) Contrast-enhanced CT performed two weeks after the PSL dose was increased revealed disappearance of the aortic wall thickening and periaortic soft tissue inflammation.

Plain chest X-ray, an electrocardiogram, and transthoracic echocardiography findings were all normal. However, contrast-enhanced CT revealed thickening of the aortic wall from the ascending aorta to the arch, along with periaortic soft tissue inflammation (Fig. 1).

Additional tests of blood culture, interferon-γ release assay, β-D glucan, HBs-antigen/HBs-antibody, HCV-antibody, IgG-4, myeloperoxidase-anti-neutrophil cytoplasmic antibody (ANCA), and Proteinase 3-ANCA after admission were negative. Her human leukocyte antigen was A2, B51, B62, although she had no history of oral or genital ulcers, ocular lesions, or cutaneous lesions, such as pathergy reactions, erythema nodosum and pseudofolliculitis suggestive of Behcet’s disease. She did not present with bloody diarrhea or abdominal pain. Anti-β2-glycoprotein I antibody, lupus anticoagulant, and anti-cardiolipin antibody were positive. She had no history of thrombosis and had had two pregnancies and deliveries. Brain magnetic resonance imaging revealed no ischemic changes. Antiretropenal antibody was negative. Based on these findings, we diagnosed her with lupus aortitis.

Since she strongly desired the same moderate-dose PSL therapy as before, and with the intention of minimizing the adverse effects of glucocorticoids, we started PSL at a dose of 30 mg/day (0.5 mg/kg/day). She did not approve of our suggestion that she should take hydroxychloroquine (HCQ) as standard therapy for SLE or another immunosuppressant as a glucocorticoid-sparing drug because she was afraid of...
| No | Authors | Age | Sex | Diagnosis | Symptoms | Treatment for admission | Site | Ankylosing Spondylitis | Diabetes | Hypertension | Arthritis | Outcome | Pathological findings |
|----|---------|-----|-----|-----------|----------|--------------------------|------|------------------------|---------|-------------|----------|----------|------------------------|
| #1 | Polhemus JA | 63 | F | Extremity claudication | Symptomatic, examination | PSL 5-30 mg | Celiac artery, abdominal aorta | Yes | Yes | No | Death | Polyarteritis nodosa |
| #2 | Shihada H. | 46 | M | Dispepsia | Angina | PSL 60 → 10 mg | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #3 | ElChaharty | 59 | F | Poor pulse in palpation | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #4 | Iwakiri T. et al. | 29 | M | Poor pulse in palpation | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #5 | Sato T. et al. | 29 | F | Poor pulse in palpation | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #6 | MacLeod O. | 47 | M | All heart failure | None | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #7 | Shihada W. | 56 | M | None | None | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #8 | Shihada T. et al. | 30 | F | None | None | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #9 | Grand RW. et al. | 60 | M | Chest pain radiating to back | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #10 | Hori-Tani et al. | 6 | F | Poor pulse in palpation | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #11 | Watanabe H. | 27 | F | Poor pulse in palpation | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #12 | Hasegawa K. | 40 | M | Chest pain, dyspepsia | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #13 | Sato T. et al. | 47 | F | Back pain | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #14 | Piscitelli A. | 37 | M | Back pain | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #15 | Wang L. et al. | 36 | M | Poor pulse | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #16 | Rojas-Leiva F. | 44 | F | Weight-loss, fatigue | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |
| #17 | Rojas-Leiva F. | 34 | F | Pericarditis | Angiography | None | Abdominal | No | Yes | Yes | Recovery | Polyarteritis nodosa |

**Table 2. Literature Review of Cases of Lupus Aortitis.**
| No | Authors                          | Age | Sex | Symptoms                       | Diagnosis                               | Site            | Aneurysm | Dissection | Prior treatment | Treatment for aortitis at admission | Outcome                          | Pathological findings                        |
|----|---------------------------------|-----|-----|---------------------------------|-----------------------------------------|-----------------|----------|------------|----------------|-------------------------------------|----------------------------------------|-----------------------------------------------|
| #18| Takagi H, et al.                 | 35  | F   | None                            | CT, resected specimen                   | Descending      | (+)      | (-)        | PSL            | PSL, surgery                         | Recovery                             | Obliterative endarteritis of the vasa vasorum in the adventitia, worm-eaten disruption of the elastic lamina in the media, perivascular lymphoplasmacytic infiltration in the adventitia and media, calcifications and atheroma within the thickened intima |
| #19| Caso V, et al.                   | 36  | F   | Left hemiplegia                 | MRA, angiography                        | Internal carotid artery, renal artery  | Arch            | (-)        | (-)        | None                       | PSL 75 mg, CY 2 mg/kg                | Recovery                             | Unknown                                      |
| #20| Silver A, et al.                | 30  | M   | Abdominal pain, vomiting        | CT, intraoperative findings             | Autopsy         | (-)      | (-)        | PSL 60 mg       | PSL 60 mg, MMF, Surgery               | Recovery                             | Small-vessel vasculitis accompanying intravascular thrombi in the pericardial vasculature |
| #21| Goel D, et al.                   | 32  | F   | None                            |                                         |                 | (-)      | (-)        | mPSL pulse, PSL, AZA, MMF, HCQ       | mPSL, HCQ                            | Death                                | Systemic small-vessel vasculitis including the vasa vasorum |
| #22| Breynaert C, et al              | 57  | M   | Fever, chest pain               | PET                                      | Thoracic        | (-)      | (-)        | mPSL 32 mg → Discontinued             | mPSL 32 mg                            | Recovery                             | Obliterative endarteritis in the adventitia, patchy necrosis in the media |
| #23| Brinster DR, et al              | 23  | F   | Fever, pleural pain             | CT, MRI, resected specimen              | Ascending       | (+)      | (-)        | None                       | Death                                | Death                                | Systemic small-vessel vasculitis including the vasa vasorum |
| #24| Soyooz A, et al                  | 28  | M   | abdominal pain, nausea          | Contrastenhanced CT                     | Thoracic-Abdominal | (-)      | (-)        | Warfarin                    | PSL pulse, PSL 60 mg, MMF             | Recovery                             | Unknown                                      |
| #25| Seo H, et al.                    | 30  | F   | chest pain                      | Contrastenhanced CT                     | Ascending-external Iliac                 | (-)      | (+)        | Betamethasone 1.5mg               | Betamethasone 3mg, Surgery, mPSL pulse, PSL 1 mg/kg/day | Recovery                             | Dissection of the elastic media, obliterative endarteritis of the vasa vasorum |
| #26| Sokahki D, et al                | 23  | F   | Fever, dyspnea, chest pain      | Contrastenhanced CT                     | Ascending       | (-)      | (-)        | None                       | mPSL pulse                           | Death (graft infection)                | Diffuse lymphocytic infiltration, disruption of the elastic lamina and necrosis of the media |
| #27| Medina G, et al                 | 17  | F   | Generalized edema               | Autopsy                                 |                 | (-)      | (-)        | None                       | mPSL pulse                           | Death                                | Systemic polyangitis, lymphocytic infiltration of all layers of the aorta |
| #28| Jung SY, et al                  | 21  | F   | Dyspnea                         | PET, resected specimen                  | Ascending-Arch | (-)      | (-)        | None                       | High-dose PSL                        | Recovery                             | Obliterative endarteritis and perivascular lymphocytic infiltration in the adventitia and media, necrosis with neo-vascularization of the media |
| #29| Our case                        | 49  | F   | Fever, dyspnea, migratory chest pain | Contrastenhanced CT                     | Ascending-Arch | (-)      | (-)        | PSL 8 mg                  | PSL 30 mg (0.5 mg/kg)                | Recovery                             | Unknown                                      |

AR: aortic regurgitation, A: Aortic valve; AZA: azathioprine, CT: computed tomography, CY: cyclophosphamide, HCQ: hydroxychloroquine, IVCY: Intravenous cyclophosphamide, MMF: mycophenolate mofetil, mPSL: methylprednisolone, MRA: magnetic resonance angiography, MRI: magnetic resonance imaging, MTX: methotrexate, PCI: percutaneous coronary intervention, PET: positron emission tomography, PSL: prednisolone, TEE: transesophageal echocardiography
developing an allergy. As a result, her chest pain resolved, and her inflammatory marker levels and anti-dsDNA antibodies became negative after two weeks. Contrast-enhanced CT performed two weeks after the increased PSL dose revealed disappearance of the aortic wall thickening and peri-aortic soft tissue inflammation (Fig. 2). She was discharged on day 21.

PSL was continued at the initial dose for two weeks. Subsequently, the dose was gradually reduced by 5 mg every 2 weeks to 15 mg, and then by 2.5 mg every 4 weeks to 10 mg. As of 22 months after the symptom onset, there has been no recurrence with a dose of 7.5 mg of PSL.

Discussion

We described the first case of lupus aortitis that responded to conservative treatment with moderate doses of PSL. There has been no textbook or systematic review describing the characteristics of lupus aortitis or its treatment strategy. There have been several cohort studies of vasculitis in SLE (32, 33), and aortitis was not mentioned in any of those reports.

In February 2020, we searched PubMed, Google Scholar, and Google for previous reports on lupus aortitis using the terms “SLE, aortitis” and “lupus, aortitis” in English and “SLE, aortitis” in Japanese. We identified 28 such cases of lupus aortitis in review papers and their cited references in English or Japanese (1-27) (Table 2). Since nine of the cases were fatal, lupus aortitis was thought to be a serious condition. The causes of death in many of these cases were attributed to complications from aortic dissection due to active aortitis or to postoperative complications (8, 9, 15, 25). Among the patients who recovered, surgery was performed for aortic dissection or aortic aneurysm, although most reports concerning surgical aortic repair did not mention the induction dose of corticosteroids (7, 8, 12-14, 17).

High-dose glucocorticoids have been recommended as the initial treatment for Takayasu’s arteritis or giant cell arteritis, which are major types of aortitis (34, 35). However, fatal cases of lupus aortitis without aortic dissection or aortic aneurysm despite high-dose glucocorticoid administration have been reported (25, 26). Since heterogeneous outcomes have been reported and there have been no controlled trials, the need for high-dose glucocorticoids as the initial treatment for lupus aortitis is unclear.

However, it has been reported that glucocorticoid administration itself can induce atherosclerotic changes and contribute to the fragility of the aortic tunica media, which might induce aortic aneurysmal enlargement (12, 13, 15, 28-31). Therefore, if possible, medical intervention with moderate-dose glucocorticoids seems beneficial for limiting the amount of glucocorticoids administered. It may thus be reasonable to consider initial treatment with moderate-dose glucocorticoids for lupus aortitis when there is no aortic dissection or aortic aneurysm formation at the initial evaluation and close follow-up is possible, or when there are additional factors that are relative contraindications to high-dose glucocorticoids.

Our patient showed no serious complications, such as aortic dissection or aortic aneurysm formation. After explaining the risks associated with insufficient treatment to this patient, moderate-dose glucocorticoid therapy was started, which successfully induced remission. The best immunosuppressant for lupus aortitis is unclear (18, 19, 22, 23). If the patient’s aortitis had not responded to the initial treatment, we would have increased the dose of PSL and persuaded her to take an immunosuppressant, such as cyclophosphamide or mycophenolate mofetil, while sharing information about the adverse events associated with the immunosuppressant. We intend to add HCQ if she agrees to take it, as HCQ is recommended for all SLE patients as the standard therapy (36).

There have been no previous reports of successful remission of lupus aortitis with moderate-dose glucocorticoids. It will therefore be necessary to examine more cases in the future to identify the characteristics of patients who are likely candidates for successful treatment with moderate doses of glucocorticoids.

The authors state that they have no Conflict of Interest (COI).

References

No funding was received for this manuscript.

1. Peloheimo JA. Obstructive arteritis of Takayasu’s type. Acta Medica Scandinavica. Supplementum 468: 7-15, 1967.
2. Shulman HJ, Christian CL. Aortic insufficiency in systemic lupus erythematosus. Arthritis Rheum 12: 138-146, 1969.
3. El-Ghobarey A, Grennan DM, Hadidi T, El-Bodawy S. Aortic incompetence in systemic lupus erythematosus. BMJ 2: 915-916, 1976.
4. Igarashi T, Nagaoka S, Matsunaga K, et al. Aortitis syndrome (Takayasu’s arteritis) associated with systemic lupus erythematosus. J Rheumatol 16: 1579-1583, 1989.
5. Saxe PA, Altmann RD. Aortitis syndrome (Takayasu’s arteritis) associated with systemic lupus erythematosus. J Rheumatol 17: 1251-1252, 1990.
6. MacLeod CB, Johnson D, Frable WJ. “Tree-barking” of the ascending aorta. Syphilis or systemic lupus erythematosus? Am J Clin Pathol 97: 58-62, 1992.
7. Strehbens WE, Delahunt B, Shirer WC, Naik DK. Aortic aneurysm in systemic lupus erythematosus. Histopathology 22: 275-277, 1993.
8. Shibata Y, Yamada T, Ishihara K, et al. A Case of Abdominal Aortic Aneurysm Associated with Systemic Lupus Erythematosus. Japanese Journal of Cardiovascular Surgery (Abstract in Japanese) 23: 217-220, 1994 (in Japanese).
9. Guard RW, Gotic-Graham I, Edmonds JP, Thomas AC. Aortitis with dissection complicating systemic lupus erythematosus. Pathology 27: 224-228, 1995.
10. Menon J, Karande SC, Kambekar KP, Lalwani SG, Nadkarni UB, Jain MK. Systemic lupus erythematosus with aortoarteritis. Indian Pediatr 33: 238-241, 1996.
11. Willett WF, Kahn MJ, Gerber MA. Lupus aortitis: a case report and review of the literature. J La State Med Soc 148: 55-59, 1996.
12. Hussain KM, Chanda H, Santhanan V, Sehgal S, Jain A, Denes P. Aortic dissection in a young corticosteroid-treated patient with
systemic lupus erythematosus—a case report. Angiology 49: 649-652, 1998.
13. Kameyama K, Kuramochi S, Ueda T, et al. Takayasu’s aortitis with dissection in systemic lupus erythematosus. Scand J Rheumatol 28: 187-188, 1999.
14. Peguero A, Rabb H, Morgan M, Rosen R, Bittle P, Ramirez G. Lupus aortitis and aneurysm case report and review of the literature. Journal of Clinical Rheumatology 5: 32-36, 1999.
15. Wang J, French SW, Chung CC, McPhaul L. Pathologic quiz case: an unusual complication of systemic lupus erythematosus. Arch Pathol Lab Med 124: 324-326, 2000.
16. Rojo-Leyva F, Ratliff NB, Cosgrove DM, Hoffman GS. Study of 52 patients with idiopathic aortitis from a cohort of 1,204 surgical cases. Arthritis Rheum 43: 901-907, 2000.
17. Takagi H, Mori Y, Iwata H, et al. Non-dissecting aneurysm of the thoracic aorta with arteritis in systemic lupus erythematosus. J Vasc Surg 35: 801-804, 2002.
18. Caso V, Paciaroni M, Parnetti L, et al. Stroke related to carotid artery dissection in a young patient with Takayasu arteritis, systemic lupus erythematosus and antiphospholipid antibody syndrome. Cerebrovascular Diseases 13: 67-69, 2002.
19. Silver A, Shao C, Ginzler E. Aortitis and Aortic Thrombus in Systemic Lupus Erythematosus. Archives of Rheumatology 26: 262-264, 2011.
20. Goel D, Reddy SR, Sundaram C, Prayaga AK, Rajasekhar L, Narasimulu G. Active necrotizing cerebral vasculitis in systemic lupus erythematosus. Neuropathology 27: 561-565, 2007.
21. Breynaert C, Cornelis T, Stroobants S, Bogaert J, Vanhoof J, Blockmans D. Systemic lupus erythematosus complicated with aortitis. Lupus 17: 72-74, 2008.
22. Brinster DR, Grizzard JD, Dash A. Lupus Aortitis Leading to Aneurysmal Dilatation in the Aortic Root and Ascending Aorta. The Heart Surgery Forum 12: E105-E108, 2009.
23. Soyuz A, Isik M, Dogan I, Kitic L, Kiraz S. Retroperitoneal Fibrosis and Aortitis as the Initial Findings of Systemic Lupus Erythematosus. Archives of Rheumatology 26: 262-264, 2011.
24. Seo H, Hirai H, Sasaki Y, Suehiro S. Aortic Dissection Due to Vasculitis in a Young Woman with Systemic Lupus Erythematosus. Nihon Kokkan Geka Gakkai Zasshi (The Japanese Journal of Vascular Surgery) (Abstract in English) 21: 33-36, 2012 (in Japanese).
25. Sokalski D, Spring TC, Roberts W. Large artery inflammation in systemic lupus erythematosus. Lupus 22: 953-956, 2013.
26. Medina G, González-Pérez D, Vázquez-Iúaíz C, Sánchez-Uribe M, Saavedra MA, Jara LJ. Fulminating systemic vasculitis in systemic lupus erythematosus. Case report and review of the literature. Lupus 23: 1426-1429, 2014.
27. Jung SY, Park HS, Jhee JH, et al. A Case of Aortitis with Systemic Lupus Erythematosus. Journal of Rheumatic Diseases (Abstract in English) 22: 205-208, 2015 (in Korean).
28. Varas-Lorenzo C, Rodriguez LA, Maguire A, Castellsague J, Perez-Guthmann S. Use of oral corticosteroids and the risk of acute myocardial infarction. Atherosclerosis 192: 376-383, 2007.
29. Ajeganova S, Svensson B, Hafström I, et al.; BARFOT Study Group. Low-dose prednisolone treatment of early rheumatoid arthritis and late cardiovascular outcome and survival: 10-year follow-up of a 2-year randomised trial. BMJ Open 4: e004259, 2014.
30. Tajima Y, Goto H, Ohara M, et al. Oral Steroid Use and Abdominal Aortic Aneurysm Expansion - Positive Association. Circulation Journal 81: 1774-1782, 2017.
31. Ohara N, Miyata T, Kurata A, Oshiro H, Sato O, Shigematsu H. Ten Years Experience of Aortic Aneurysm Associated with Systemic Lupus Erythematosus. Eur J Vasc Endovasc Surg 19: 288-293, 2000.
32. Drenkard C, Villa AR, Reyes E, Abello M, Alarcón-Segovia D. Vasculitis in systemic lupus erythematosus. Lupus 6: 235-242, 1997.
33. Ramos-Casals M, Nardi N, Lagrutta M, et al. Vasculitis in systemic lupus erythematosus: prevalence and clinical characteristics in 670 patients. Medicine (Baltimore) 85: 95-104, 2006.
34. Bossone E, Pluchinotta FR, Andreas M, et al. Aortitis. Vascular Pharmacology 80: 1-10, 2016.
35. Töpel I, Zorger N, Steinbauer M. Inflammatory diseases of the aorta. Gefasschirurgie 21 (S2): 80-86, 2016.
36. Fanouriakis A, Kostopoulou M, Alunno A, et al. 2019 update of the EULAR recommendations for the management of systemic lupus erythematosus. Ann Rheum Dis 78: 736-745, 2019.

The Internal Medicine is an Open Access journal distributed under the Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License. To view the details of this license, please visit (https://creativecommons.org/licenses/by-nc-nd/4.0/).