Ascending aortic true aneurysm after acute aortic dissection in a patient with systemic lupus erythematosus

Ryosuke Kowatari[1], Hanae Sasaki[1], YUKI Imamura[1], Shintaro Goto[2], Akira Kurose[2], and Masahito Minakawa[1]

[1]Hirosaki Daigaku Igakubu Daigakuin Igaku Kenkyuka Seikei Gekagaku Koza
[2]Hirosaki Daigaku Igakubu Daigakuin Igaku Kenkyuka

August 22, 2022

Abstract

A 50-year-old woman presented to our hospital with shortness of breath on exertion and nocturnal dyspnea. She had undergone total aortic arch replacement for Stanford type A aortic dissection 17 years previously and was taking prednisolone for systemic lupus erythematosus. Computed tomography showed that the 63-mm ascending aorta near the proximal anastomosis site compressed the superior vena cava and right atrium. Cardiac catheterization showed occlusion of the left anterior descending branch. The patient underwent urgent surgery for ascending aortic aneurysm and coronary artery occlusion. Microscopic examination revealed that the aneurysm was true. This report highlights that in patients with systemic lupus erythematosus, aortic aneurysms can reoccur even after total arch replacement.

Ascending aortic true aneurysm after acute aortic dissection in a patient with systemic lupus erythematosus

Running title: Aortic aneurysm in SLE

Hanae Sasaki MD[1)], Ryosuke Kowatari PhD[1)], Yuki Imamura MD[1)], Shintaro Goto PhD[2)], Akira Kurose PhD[3)], Masahito Minakawa PhD[1)]

1. Department of Thoracic and Cardiovascular Surgery, Hirosaki University School of Medicine
2. Department of Pathology and Bioscience, Hirosaki University School of Medicine
3. Department of Anatomic Pathology, Hirosaki University School of Medicine

Address for correspondence:

Ryosuke Kowatari PhD

Department of Thoracic and Cardiovascular Surgery, Hirosaki University Graduate School of Medicine, 5 Zaifu-cho, Hirosaki, Aomori 036-8562, Japan.

Tel: +81-172-39-5074

Fax: +81-172-37-8340.

E-mail:*kowatari@hirosaki-u.ac.jp*

Funding : None.

Data availability statement : The datasets generated and/or analyzed during the current study are available from the corresponding author on reasonable request.

Disclosure : None.

Institutional Review Board waiver: The need for approval was waived by institutional review board.

Patient consent statement: The patient provided written informed consent for publication.

Acknowledgments : None.

ABSTRACT

A 50-year-old woman presented to our hospital with shortness of breath on exertion and nocturnal dyspnea. She had undergone total aortic arch replacement for Stanford type A aortic dissection 17 years previously and was taking prednisolone for systemic lupus erythematosus. Computed tomography showed that the 63-mm ascending aorta near the proximal anastomosis site compressed the superior vena cava and right atrium. Cardiac catheterization showed occlusion of the left anterior descending branch. The patient underwent urgent surgery for ascending aortic aneurysm and coronary artery occlusion. Microscopic examination revealed that the aneurysm was true. This report highlights that in patients with systemic lupus erythematosus, aortic aneurysms can reoccur even after total arch replacement.

Keywords: systemic lupus erythematosus, total aortic arch replacement, aortic aneurysm

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease with multi-organ involvement and is associated with the presence of autoantibodies. Patients with SLE often develop aortic events such as aortic aneurysms and dissections[1, 2]. Pseudoaneurysms are rare complications after aortic surgery and have been reported in patients with SLE[3]. However, little is known about true aneurysms of the residual ascending aorta after aortic surgery. We report the case of a patient with SLE who developed a true aneurysm of the ascending aorta 17 years after total arch replacement for an acute aortic dissection.

Case

A 50-year-old woman presented with shortness of breath on exertion. She had undergone total aortic arch replacement for Stanford type A aortic dissection 17 years previously. She had been taking prednisolone (7.5 mg) for SLE since she was 18 years old. Transthoracic echocardiography showed left ventricular diastolic and systolic diameters of 52 and 41 mm, respectively, left ventricular ejection fraction of 42.5%, and mild aortic regurgitation. Echocardiography revealed a large ascending aortic aneurysm compressing the right atrium. Computed tomography showed the 63-mm ascending aorta near the proximal anastomosis site compressing the superior vena cava and right atrium (Fig. 1) and a 53-mm thoracoabdominal aneurysm (TAA) and a 38-mm abdominal aortic aneurysm (AAA). Cardiac catheterization showed chronic total occlusion of the left anterior descending (LAD) branch. The patient underwent urgent surgery for ascending aortic aneurysm and LAD occlusion.

Cardiopulmonary bypass (CPB) was initiated via right axillary and right femoral artery perfusion, with inferior vena cava drainage through the right femoral vein. Superior vena cava drainage was performed via median sternotomy and CPB was established. The harvested great saphenous vein was anastomosed to the LAD during systemic cooling to a bladder temperature of 23°C. After circulatory arrest, the aneurysmal aortic wall was incised. The artificial blood distal to the aneurysm was clamped and circulation was resumed. Cardioplegic solution was infused from the coronary ostium and anastomosed graft to achieve cardiac arrest. Ascending aortic replacement was performed using a 24-mm J-graft (Japan Lifeline Co., Ltd., Tokyo, Japan). Proximal anastomosis was performed at the sinotubular junction level. The saphenous vein graft was anastomosed to the J-graft and the aorta was declamped. A 5-mm hole on the right atrium that had been adherent to the aneurysm was found and directly closed. The CPB weaning was uneventful and the duration of surgery, CPB, cardiac arrest, and circulatory arrest were 494, 229, 113, and 3 mins, respectively. There was no recurrence of ascending aortic aneurysm 2 years postoperatively.

Microscopic examination revealed that the aneurysm was true because the aneurysmal wall was composed of

all three layers including tunica intima, media, and externa and showed severe atherosclerotic changes. The tunica intima thickened with the formation of atheroma. The smooth muscles of tunica media decreased, and the elastic fibers were disordered. Some elastic fibers ruptured, and fragmented fibers were observed in the tunica media. (Fig. 2a, b).

Discussion

Aortic dissections and aneurysms are rare complications in patients with SLE; however, their incidence is higher in patients with SLE than in age- and sex-matched controls[1, 2]. Patients with SLE develop aortic aneurysms at a relatively early age[4]. Pathological changes in patients with SLE include cystic medial necrosis with mucopolysaccharide deposition and Marfan-like changes, such as destruction of elastic fibers[5]. Interestingly, histopathology results from the present case showed a tear in the tunica media elastic fibers, suggesting that this was the cause of the aortic dissection 17 years previously. Various factors, other than atherosclerosis, have been postulated to contribute to the formation of aortic aneurysms in patients with SLE. Mucoid degeneration, vascular injury, hypertension, arteriosclerosis, and steroid therapy are reportedly associated with aneurysm formation [6]. Long-term steroid therapy inhibits chondroitin sulfate and granulation tissue formation, affecting connective tissues and increasing the incidence of atherosclerosis[6, 7]. The thoracic aorta is considered more resistant to atherosclerosis than the abdominal aorta. The difference in susceptibility to lipid deposition and subsequent plaque formation suggests differences in the cellular and extracellular compositions of the thoracic and abdominal aorta. This supports a different pathophysiological mechanism for the development of TAA and AAA[8]. Vasculitis and cystic medial degeneration reportedly cause TAA, whereas atherosclerotic changes due to long-term steroid therapy cause AAA in patients with SLE[9]. The present case was of a TAA with severe atherosclerotic lesions. These lesions and her early age suggest that the TAA may have been influenced by SLE and steroid therapy.

The incidence of postoperative pseudoaneurysm after cardiac surgery is estimated to be $<0.5\%$[10]. Previous reports include pseudoaneurysm occurring 1 year after acute aortic dissection repair[3], and true aneurysm recurrence 5 months after AAA repair in SLE patients[11]. We should consider the possibility of true aneurysms developing in patients with SLE even after total arch replacement and consider using a technique that does not leave the aortic wall short of the sino-tubular junction.

Author contributions

Concept/design: HS, RK, MM; Drafting the article: HS; Critical revision of the article: RK; Approval of article: All authors

References

1. Wang SH, Chang YS, Liu CJ, et al. Incidence and risk analysis of aortic aneurysm and aortic dissection among patients with systemic lupus erythematosus: a nationwide population-based study in Taiwan. Lupus 2014;23:665-671.
2. Guy A, Tiosano S, Comaneshter D, et al. Aortic aneurysm association with SLE - a case-control study. Lupus 2016;25:959-963.
3. Shimada A, Yamamoto T, Endo D, et al. Pseudoaneurysm with a fistula to the right ventricle late after surgical repair of type A aortic dissection in a patient with systemic lupus erythematosus. J Cardiothorac Surg 2022;17:83.
4. Ohara N, Miyata T, Kurata A, et al. Ten years' experience of aortic aneurysm associated with systemic lupus erythematosus. Eur J Vasc Endovasc Surg 2000;19:288-293.
5. Choi KH, Rim SJ, Lee SK, et al. Dissecting aortic aneurysm with aortic-valve insufficiency in systemic lupus erythematosus. Nephrol Dial Transplant 1999;14:969-973.
6. Sato J, Kawakami T, Nakabayashi K, et al. Multiple aortic aneurysms complicated by a rupture in the systemic lupus erythematosus: a case report. Pathol Res Pract 2008;204:845-850.
7. Takagi H, Mori Y, Iwata H, et al. Nondissecting aneurysm of the thoracic aorta with arteritis in systemic lupus erythematosus. J Vasc Surg 2002;35:801-804.

8. Ruddy JM, Jones JA, Spinale FG, et al. Regional heterogeneity within the aorta: relevance to aneurysm disease. J Thorac Cardiovasc Surg 2008;136:1123-1130.
9. Kurata A, Kawakami T, Sato J, et al. Aortic aneurysms in systemic lupus erythematosus: a meta-analysis of 35 cases in the literature and two different pathogeneses. Cardiovasc Pathol 2011;20:e1-7.
10. Atik FA, Navia JL, Svensson LG, et al. Surgical treatment of pseudoaneurysm of the thoracic aorta. J Thorac Cardiovasc Surg 2006;132:379-385.
11. Washiyama N, Kazui T, Takinami M, et al. Surgical treatment of recurrent abdominal aortic aneurysm in a patient with systemic lupus erythematosus. J Vasc Surg 2000;32:209-212.

Figure legends

Figure 1: Computed tomography and its schema showing the 63-mm ascending aorta near the proximal anastomosis site. Arrows indicate an ascending aortic aneurysm compressing the right atrium.

Figure 2: Histological findings of the aneurysmal wall with severe atherosclerotic changes. (a) Hematoxylin and Eosin (H&E) staining highlights that the wall was composed of tunica intima (*), media (**), and externa (***). The tunica intima thickened with the formation of atheroma. (b) Masson-Goldner stain indicated the decrease of smooth muscle (red stained fibers) and the disordered arrangements of elastic fibers (brown to dark stained fibers). Some elastic fibers ruptured, and fragmented fibers were observed (white arrow).

Hosted file

220809 Figure Ascending aortic true aneurysm after acute aortic dissection in a patient with systemic lu
available at https://authorea.com/users/482614/articles/582717-ascending-aortic-true-aneurysm-after-acute-aortic-dissection-in-a-patient-with-systemic-lupus-erythematosus