Total arch replacement with stented elephant trunk technique for syphilitic thoracic aortic aneurysm

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After the advent of antibiotic treatment, tertiary syphilis is rarely observed over the last several decades, and its cardiovascular manifestations are particularly rare. Syphilitic aortitis typically involves the tubular portion of the ascending aorta, aortic arch and descending thoracic aorta, sparing the sinuses of Valsalva.[1] Although exceptional cases of treatment of syphilitic thoracic aortic aneurysm have been reported,[2-4] the optimal technique for treatment is not established. Total arch replacement combined with stented elephant trunk technique was proven to be feasible in patients with type A aortic dissection. Herein, we described our experience of total arch replacement with stented elephant trunk technique in three patients presenting with syphilitic thoracic aortic aneurysm involving the aortic arch and proximal descending aorta.

Three male patients (aged 53, 64, and 67 years, respectively) who initially presented with hoarseness or progressive dyspnea, were referred to our hospital. Their medical history was significant for hypertension. Chest computed tomography (CT) scans showed thoracic aortic aneurysm. Thereafter, computed tomographic angiography (CTA) demonstrated a large saccular thoracic aortic aneurysm involving the aortic arch and proximal descending aorta, as well as ascending aortic ulcer [Figure 1A and 1B]. The large aneurysm led to compression of the adjacent organs, such as recurrent laryngeal nerve leading to hoarseness, trachea, and left lung leading to progressive dyspnea. The cardiac examinations were unremarkable with no aortic root aneurysm and aortic valve insufficiency. CTA was used to exclude coronary artery disease. All data were collected from the database of the Division of Cardiothoracic Surgery with the permission of the Institutional Review Board of Changhai Hospital. This clinical series study was conducted after receiving patients’ consent.

At admission, the patients had no previous history or symptoms of syphilis. However, the routine pre-operative laboratory test for syphilis was highly positive, and the rapid plasma reagin (RPR) test was 1:64. Confirmatory fluorescent treponemal antibody was positive. Hence, a presumptive diagnosis of tertiary syphilis was made. Antibiotic therapy using penicillin was first administered, at least for two weeks before the operation. Furthermore, prednisone was orally administered to alleviate the Jarisch-Herxheimer reaction.

Ascending aorta and aortic arch replacement using a tetrafurcate graft, combined with stented elephant trunk implantation, was performed in all patients via a median sternotomy under cardiopulmonary bypass (CPB) with selective cerebral perfusion (SCP). Cannulation of the right axillary artery was routinely conducted for CPB and SCP. CPB and cooling were started after cannulation of the right axillary artery and the right atrium. Circulatory arrest was established when the nasopharyngeal temperature reached 27°C. The pathological ascending aorta was resected [Figure 1C and 1D]. The ascending aorta and aortic arch were replaced with a tetrafurcate graft. The distal arch was circumferentially transected proximal to the aortic aneurysm. If the aortic aneurysm involved the left subclavian artery and/or the left common carotid artery, they were reconstructed with a graft branch. Then, a 10 cm long stented graft (MicroPort Medical Co Ltd, Shanghai, China) in a bound was inserted into the aneurysm, and the terminal end of the stented graft was located at normal zone of descending aorta. Then the anastomosis between the tetrafurcate graft and the distal arch was conducted by the open aortic technique. The pathology examination of the aneurysmal wall revealed diffuse infiltration of lymphocytes and plasma cells, which is characteristic of syphilitic aortitis.
There was no hospital death, and the patients recovered uneventfully. Stroke, paraparesis, visceral malperfusion and lower extremity malfunction were not observed. Post-operatively, a follow-up CTA demonstrated successful repair with full exclusion of the aneurysm without endoleak [Figure 1E]. A serological test for syphilis was conducted, with additional administration of antibiotic therapy if necessary.

Syphilitic aortic aneurysm is a relatively rare type of cardiovascular syphilis, which is a tertiary manifestation, generally occurring 10 to 30 years after the original untreated infection. Vascular involvement in late syphilis is due to an obliterator small vessel endarteritis, usually of the vasa vasorum of the thoracic aorta, fostering loss of structural integrity of the tunica media, and subsequent fibrosis and calcification. Aortitis typically involves the tubular portion of the ascending aorta, aortic arch and descending thoracic aorta, sparing the sinuses of Valsalva.\cite{1} Definitive diagnosis of syphilitic aortitis can be challenging owing to prolonged latency from primary infection. Clinical diagnosis is most often made based upon serological confirmation of syphilis and a characteristic pattern of vascular involvement.

Patients with syphilitic aortic aneurysm can be asymptomatic for many years without significant clinical manifestations, but thoracic aortic aneurysm has high annual mortality rates. Once the aortic aneurysm is diagnosed, surgical treatment is necessary. The surgical procedure depends on the site involved in cardiovascular syphilis. Ascending aorta replacement is the most commonly performed surgical procedure for the treatment of syphilitic aortic aneurysm, because 50% of syphilitic aortic aneurysms involve the ascending aorta. For the syphilitic aortic aneurysm located at distal aortic arch, Yasuda et al\cite{2} adopted endovascular repair using a hand-made fenestrated stent graft. In order to secure an adequate landing zone, the left subclavian artery had to be occluded, which had the potential risk of ischemia of the brain and spinal cord. Additionally, the intima of proximal aortic arch and ascending aorta in patients with cardiovascular syphilis could be pathological, so serious complications may be observed after thoracic endovascular aortic repair, such as retrograde type A dissection, if the proximal landing zone of the stent is located in the proximal arch or the ascending aorta. One method to avoid these limitations is the hybrid (or staged) procedure.\cite{3} Although placing the conventional elephant trunk into the descending aneurysm is not very difficult, deaths caused by rupture of the remaining aortic aneurysm in the interval between the two procedures have been reported.\cite{5}

Given the above-mentioned problems, we adopted the total arch replacement with stented elephant trunk technique for three patients presenting with syphilitic thoracic aortic aneurysm involving the aortic arch and proximal descending aorta, which achieved satisfactory outcomes. This procedure combined the advantages of open surgical treatment and interventional methods while simultaneously avoiding the shortcomings of these approaches. First, the pathological aortic segment (from the proximal aortic lesion to the distal arch), which is susceptible to further dilation/rupture, was resected. Second, the procedure was simplified, because the aneurysm does not need to be incised, which is an effective way to prevent bleeding due to tissue weakness; the aortic arch was circumferentially transected proximal to the aortic aneurysm, and then the stented graft was inserted into the descending aorta during intra-operative view of selective antegrade brain perfusion. Furthermore, the aneurysm was completely eliminated, because the proximal end of the stent graft was firmly fixed to the distal end of the

![Figure 1: (A) Pre-operative computed tomographic angiography (CTA) demonstrated a large saccular thoracic aortic aneurysm involving the aortic arch and proximal descending aorta. (B) CTA showed the specific size of saccular thoracic aortic aneurysm. (C) Intra-operative exploration showed no aortic root aneurysm and aortic valve insufficiency. (D) Intra-operative exploration showed that the wall of ascending aorta and aortic arch aneurysm was moderately thickened with moderate inflammation presumably due to syphilitic aortitis. (E) Post-operative CTA showed successful repair with full exclusion of the aneurysm without endoleak.](image-url)
prosthetic graft using suture line and the distal aorta was expanded by the rigidly spread surgical stent. The proximal end of the stented graft had 1 cm of extravascular graft, facilitating proximal anastomosis to the prosthetic graft or lengthening proximal graft using another same-size graft in order to place the distal end of the stented graft at further landing zone. If the descending aorta is diffusely expanded, a late-stage procedure for the distal end is necessary.

Conflicts of interest
None.

References
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