Frozen Elephant Trunk Technique for the Treatment of an Extensive Syphilitic Thoracic Aneurysm

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Abstract

We present the successful application of the Frozen Elephant Trunk (FET) technique for repair of a syphilitic thoracic aortic aneurysm. A 66-year-old woman with untreated syphilis presented with double saccular aneurysms in the descending and dilated ascending aorta. Following total arch replacement using FET with distal landing at T8, postoperative shrinkage of the aneurysm occurred with no signs of paraparesis or endoleak. Adaptation of the distal landing of FET is essential to avoid spinal cord injury and pathological involvement of the ascending aorta should be considered during the management of extensive syphilitic thoracic aortic aneurysms. Long-term follow-up is crucial for the unresected part of the aneurysm.

Introduction

Syphilitic aortic aneurysm is a manifestation of the tertiary stage of syphilis and is rarely encountered now a days. Treatment of this aneurysm is complicated owing to its mycotic etiology and the extensiveness of the disease, therefore demanding intensive infection control during highly invasive surgery. We report the successful application of Frozen Elephant trunk (FET) for single-stage repair of an extensive syphilitic thoracic aortic aneurysm.

Case Presentation

An asymptomatic 66-year-old woman was referred to our hospital because of a protrusion of the aortic arch detected by chest radiography during a health screen. Her medical history included hypertension and untreated syphilis that was diagnosed 30 years prior. The treponemal test was positive with the rapid plasma reagin test of 1:16 and Treponema pallidum hemagglutination of 1:40960. Computed Tomography (CT) revealed double saccular aneurysms in the descending aorta with a maximal diameter of 60 mm and slightly dilated Ascending Aorta (AA) with a diameter of 40 mm, accompanied by atheromatous thickening in the intima (Figure 1a). The aneurysm extended to the level of T7 and the aortic valve was used as reference for T8 (Figure 1b). Considering her background, her thoracic aorta was believed to be affected by syphilitic aortitis, and surgical intervention was planned owing to the risk of the rupture of the descending aorta. Pre-operatively, she received 2 grams of intravenous ceftriaxone daily for two weeks followed by 2 grams of oral amoxicillin daily for six weeks.

After a median sternotomy, cardiopulmonary bypass was established by cannulation of the left subclavian artery interposed by a 9 mm Dacron graft (J-Graft, Japan Lifeline, Tokyo, Japan), which was subsequently anastomosed to an epiarterial branch of a 24 mm Dacron arch graft (J-Graft Shield Neo, Japan Lifeline, Tokyo, Japan). Macroscopically, the adventitia of the AA appeared red and swollen (Figure 2a). Edematous thickening was observed in the intima, and the aortic valve was intact (Figure 2b). During circulatory arrest with hypothermia of 26.0°C with selective antegrade cerebral perfusion, the FET with a diameter of 31 mm and a length of 90 mm (Frozenji J-Graft, Japan Lifeline, Tokyo, Japan) was deployed with a distal landing at the level of the aortic valve under the guidance of Transesophageal Echocardiography (TEE). The AA and aortic arch were replaced with the 4-branched Dacron graft described above. No endoleak was detected on TEE following resumption of the antegrade flow in the descending aorta. She was discharged on postoperative day 18 uneventfully without development of paraparesis. Histologically, lymphocyte infiltration was observed in the adventitia of the AA (Figure 3a). In the media, elastic fibers were degenerated, and smooth muscle cells were depleted, resulting in the loss of integrity of the media (Figure 3b). Follow-
up CT performed after 1 year showed shrinkage of the aneurysm (Figure 4a) and no endoleak (Figure 4b).

**Discussion**

In this antibiotic era, syphilitic aortic aneurysm is a rare entity. However, Robert et al. reported that cardiovascular syphilis, such as aortic aneurysm, aortic regurgitation, or coronary ostial stenosis occurred in up to 10% of untreated patients [1]. Aortic rupture accounted for 13% of deaths in necropsy cases of syphilitic aortitis [1]. Though Yasuda, et al. [2] reported that a syphilitic thoracic aortic aneurysm was treated successfully with a fenestrated stent-graft landing in the AA, we were concerned about the slightly dilated AA with the reverse taper forum, which impeded proper landing and could have caused an endoleak. Therefore, endovascular treatment was deemed unsuitable in this case. Tokuda, et al. [3] demonstrated the feasibility of the L-incision approach involving median hemisternotomy and left anterior thoracotomy for single-stage resection of the extensive aneurysm of the aortic arch. However, this approach was associated with a higher rate of postoperative respiratory complications than the median sternotomy approach [3] Kobayashi et al. [4] reported a successful staged repair of an extensive syphilitic aortic arch aneurysm by aortic arch replacement followed by open repair of the descending thoracic aorta. However, Papakonstantinou et al. [5] pointed out that the interval mortality, which occurred during the waiting time for the second stage procedure, was as high as 25%, and mostly resulted from aortic rupture; furthermore, single-stage repair of extensive aortic arch aneurysm using FET accounted for 5% of the in-hospital deaths. The risk of Spinal Cord Injury (SCI) following FET use in our patient seemed to be high as a result of deep implantation of the FET for the extensive aneurysm. Katayama et al. [6] showed that the occurrence of SCI following the FET technique ranged from 0.4% to 24%, and placement of the distal position of the stent-graft below T9 was identified as an independent risk factor. We presumed that placing the distal end of the FET at T8 could exclude the aneurysm with a distal end at T7. In addition, she had no history of aortic repair, which is another risk factor of SCI [6]. Consequently, we preferred to avoid preoperative spinal drainage, as the reported rate of drainage-related adverse events is 6.5% and accounts for 2.5% of severe complications, such as epidural hemorrhage, meningitis, and catheter-/drainage-related neurological deficits [7]. Intra-operative endoleak did not occur; otherwise, a second-stage endovascular surgery would have been necessary. Robert et al. [1] reported that the syphilitic process reportedly begins at the sinotubular junction and involves the aortic wall, leading to a disruption of the integrity in the media, and resulting in wall weakness and vulnerability to dilatation or focal saccular aneurysm formation. Actually, those findings were observed in our specimens from the AA and, thus, our procedure may have helped to prevent the long-term ascending aortic events. Although we demonstrated shrinkage of the aneurysm without an endoleak, Yasuda et al. [2] concluded that the long-term outcome of a stent-graft in an unresected syphilitic aneurysm was unclear, despite the disappearance of the aneurysm 2 years after the surgery. Hence,
we concluded that close follow-up is important.

**Conclusion**

Careful adaptation of distal landing is important to prevent SCI and pathological involvement of the AA should be considered during the management of extensive syphilitic thoracic aortic aneurysms. Long-term follow-up is required after implantation of FET for remaining unresected syphilitic aneurysms.

**References**