Case Report

Hybrid Repair Combined with Open Surgery and Endografting for Lesions in Right Aortic Arch: Report of Three Cases

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Three cases with lesions in the right aortic arch (RAA) are presented. For two patients, whose RAA contained a retroesophageal segment, the primary treatment was total arch replacement (TAR) for acute type A dissection or severe arch angulation with thoracic endovascular aneurysm repair (TEVAR) as second-stage surgery. The third patient, who had RAA with mirror image branching, underwent supra-aortic bypass followed by TEVAR. There was no operative mortality, but the condition of two patients who underwent TAR was complicated by bilateral recurrent nerve palsy, which can be critical. The combination of TEVAR and supra-aortic bypass might thus be preferable for patients with RAA.

Keywords: right aortic arch, stentgraft, TEVAR, endovascular repair, hybrid repair

INTRODUCTION

Right aortic arch (RAA) is a relatively common anomaly which is classified by double or single arch and is categorized by a retroesophageal segment.1) The surgical treatment for aortic lesions involving RAA is complicated due to the anatomical relationship of the aorta with the trachea and esophagus, and exposure of all aortic segments may be possible only through a median sternotomy or an intercostal thoracotomy. However, thoracic endovascular aneurysm repair (TEVAR) is a viable therapeutic alternative for treatment of the descending aorta running on both sides of pleural cavities. The advantage of hybrid aortic repair utilizing TEVAR for patients with RAA is discussed.

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Patients

Clinical charts of three cases with RAA who underwent hybrid aortic repair were reviewed retrospectively.

Case 1 was a 67-year-old man who had been diagnosed with an infrarenal abdominal aortic aneurysm (AAA). He presented with sudden, severe back pain, and enhanced computed tomography showed an acute type A aortic dissection, associated with RAA. The right subclavian (RSCA), right common carotid (RCCA), left common carotid (LCCA), and left subclavian (LSCA) arteries branched in that order from the RAA with a retroesophageal segment. For emergency surgical treatment of the aortic dissection, the heart and the aorta were exposed through a median sternotomy. A cardiopulmonary bypass with bicaval venous drainage was then initiated via the right axillary and left common femoral arteries. When the RAA was opened under deep hypothermia and selective cerebral perfusion, it was found that Kommerell’s diverticulum behind the esophagus could not be approached because its location was too deep. Total arch replacement was, therefore, indicated, and distal anastomosis was initiated proximally to the diverticulum. To cover the entry, we inserted a 7-cm long elephant trunk graft into the true lumen of the descending aorta.
The patient’s postoperative course was uneventful except for hoarseness due to bilateral recurrent nerve palsy. AAA replacement was performed 3 months later. For enlargement (58 mm) of the middle descending aorta, TEVAR with GoreTAG (26 mm × 10 cm) was performed, extending from the elephant trunk to the distal descending aorta. One year postoperatively, the descending aorta showed no further enlargement.

Case 2 was a 74-year-old male who developed dilatation of Kommerell’s diverticulum and consequent descending aortic aneurysm (52 mm) with RAA containing a retroesophageal segment. The orifices of the LCCA, RCCA, and RSCA were close together at the top of the RAA and the LSCA arose from the diverticulum.

(B) Case 2 underwent total arch replacement and elephant trunk installation to create an adequate landing zone, and TEVAR with GoreTAGs (28 mm × 15 cm and 34 mm × 20 cm) was performed one week later, extending from the elephant trunk to the distal descending aorta. LCAA: left common carotid; RCCA: right common carotid; RSCA: right subclavian; RAA: right aortic arch; LSCA: left subclavian; TEVAR: thoracic endovascular aneurysm repair

Case 3 was an 81-year-old male with a history of malignant lymphoma. His RAA showed mirror image branching and the left brachiocephalic artery bifurcated to the LSCA, LCCA, RCCA, and RSCA in that order (Fig. 2). There were two saccular aneurysms, one just distal to the RSCA and the other at the proximal descending aorta. A supra-aortic bypass from the left axillary artery to the RCCA and right axillary artery was performed and TEVAR with GoreTAG (34 mm × 15 cm) two weeks later. To cope with the acute angle (311-degree) of the RAA, the proximal end of the wrapping of TAG was retracted with a snaring catheter.

Fig. 1
(A) Case 2 developed dilatation of Kommerell’s diverticulum and consequent descending aortic aneurysm (52 mm) with RAA containing a retroesophageal segment. The orifices of the LCCA, RCCA, and RSCA were close together at the top of the RAA and the LSCA arose from the diverticulum.
(B) Case 2 underwent total arch replacement and elephant trunk installation to create an adequate landing zone, and TEVAR with GoreTAGs (28 mm × 15 cm and 34 mm × 20 cm) was performed one week later, extending from the elephant trunk to the distal descending aorta. LCAA: left common carotid; RCCA: right common carotid; RSCA: right subclavian; RAA: right aortic arch; LSCA: left subclavian; TEVAR: thoracic endovascular aneurysm repair
which was inserted from the femoral artery. The patient has been unable to visit our facility because of a pelvic fracture, but his survival was confirmed by telephone interview, one year postoperatively.

**DISCUSSION**

The incidence of RAA among adults is reportedly 0.04% to 0.1%.3,4) The most frequent RAA has no retroesophageal segment or mirror image branching (77%). RAA with a retroesophageal segment is rare (4%).2) Our basic strategy for treatment of an aneurysm or dissection of the arch is total arch replacement.3) However, to deal with the risks or problems associated with patients with RAA, we chose hybrid repair including the use of TEVAR for the three cases.

In case 1, the entry tear through the median sternotomy was too deep, and additional treatment was necessary to close the entry. In case 2, the proximal anastomosis for replacement of the descending aorta was deemed too deep to be approached through the thoracotomy over Kommerell’s diverticulum. In addition, the orifices of the arch branches were too close together to create an adequate proximal landing zone for TEVAR. Case 3 was too old to undergo total arch replacement. However, RAA with mirror image branches features relatively long distances between the arch branches which allow for preservation of the blood flow to the left brachiocephalic artery and construction of an extrathoracic bypass to the right supraaortic branches.

Two of the three patients who underwent total arch replacement suffered from bilateral recurrent nerve palsy, and both of them had a retroesophageal aortic segment. In patients with RAA with a retroesophageal aortic segment, the left recurrent nerve approaches the back of the ductus arteriosus, encircles it from the left dorsal lateral
to the right ventromedial area, and goes upward along the left surface of the esophagus and trachea, whereas, the right recurrent laryngeal nerve loops the aortic arch from the right dorsal lateral area in the direction of the left ventromedial direction.4) During the surgery for both these cases, we took great care to preserve the functions of the recurrent nerves, we could not succeed. When total arch replacement is necessary for patients with RAA, such as acute type A aortic dissection, even greater care is needed. When an alternative operative procedure is feasible, the combination of a supra-aortic (extra-thoracic) bypass and TEVAR might be more effective for avoiding this serious complication of bilateral recurrent nerve palsy.

For the combination of total arch replacement and TEVAR, we utilized a synthetic graft to create an elephant trunk as the proximal landing zone for TEVAR. The placement of a stent graft inside a synthetic graft is not recommended in the instructions for use. However, the utility of this procedure, especially the utilization of an elephant trunk in the extended aneurysm, has been reported.5)

The RAA tends to have a shorter radius and more acute curvature of the arch than the usual gentle curve of a left aortic arch.6) In case 2, we anticipated a difficult passage of the GoreTAG through the acute angle, and we, therefore, decided on total arch replacement which turned out to be complicated with bilateral recurrent nerve palsy. In case 3, however, retraction of the GoreTAG with a snaring catheter was found to be useful. Moreover, the gap between the olive tip and the wrapping of stent graft has been made smoother (TAG over Molded Olive), so that the acute curvature of RAA may no longer be problematic when TEVAR with GoreTAG is used.

A major limitation of this study is the small number of cases with this rare condition. For validation of the long-term durability and efficacy of our strategy, careful follow-up will be essential.7)

**Conclusion**

Multiple-stage surgeries including endografting were found to be useful for overcoming anatomical difficulties in patients with RAA. Bilateral recurrent nerve palsy after total arch replacement can be critical, and the use of TEVAR after supra-aortic bypass may, therefore, be preferable for patients with RAA.

**Disclosure Statement**

The authors have no conflict of interest to declare.

**References**