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Case Report

Endovascular repair of thoracic aortic dissection associated with right-sided aortic arch: report of four cases

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Aortic dissection involving a right-sided aortic arch (RAA) is extremely rare with an incidence in adults of 0.04% to 0.1%[1]. Thoracic aortic dissection associated with RAA is even a more uncommon and life-threatening condition. For complicated aortic dissection, conventional open surgical repair is considered a standard therapy[2]. However, in such cases, endovascular repair has emerged as the procedure of choice as it is associated with favorable short- and mid-term results[3]. In this study, we present our experience with the use of thoracic endovascular aortic repair (TEVAR) technique for treatment of DeBakey III dissection in 4 patients with RAA. All procedures were successfully performed with a Siemens Artis Zeego DSA device.

Case report

Between September 2010 and January 2015, 3 Edwards type II[4] RAA (case 1 to 3, all with Kommerell's diverticulums) and 1 type I RAA patients (case 4, mirror image arch branches) with DeBakey III dissection were treated in our center. Radiography demonstrated entry tear in all 3 cases with Kommerell diverticulum. CT scan also defined the ordering of the aortic arch branches in these Edwards type II patients as the left common carotid artery (LCCA), right common carotid artery (RCCA), right subclavian artery (RSA) and aberrant left subclavian artery (LSA) with Kommerell diverticulum from proximal to distal, and right-sided descending aorta. With the excellent flexibility of Artis Zeego DSA, the C-arm can be easily rotated and adjusted to meet different right-sided arch angles. To obtain an expanded adequate view of the arch and its branches, the C-arm was positioned to left-anterior oblique 45° to 75°.

Case 1

A 68-year-old man was admitted to our institution with reoccurring chest and back pain for 10 years. The dissection extended from the orifice of the RCCA through the thoracoabdominal aorta (***Fig. 1A***). Measurements of the diameter of the RSA (Φ = 7.0 mm) as well as the landing zone in thoracic aorta (Φ = 37.4 mm in proximal and 26.6 mm in distal) were also obtained. A chronic aortic dissection with Kommerell's diverticulum where the aberrant LSA arose was then diagnosed (***Fig. 1D***). Willis circle, anterior artery and a dominant right vertebral artery were also clearly radiographed. The size of Kommerell's diverticulum was within the operational range. To create a prolonged proximal landing zone, a chimney stent (8 mm × 6 mm E-Luminexx; C.R. Bard, Inc., NJ, USA) was implanted retrograde into the RSA. Two stent-grafts (42 mm × 38 mm × 180 mm and 38 mm × 32 mm × 180 mm, Grikin, China) were sequentially positioned (proximal to the origin of the RCCA and distal to the descending aorta). We locked the vascular plug (14-16 lifetech, China) to occlude the LSA to prevent a type II endoleak. A control angiogram revealed complete flow exclusion of the false lumen and the correct position of the stent-graft (***Fig. 1B***). The subsequent postoperative course

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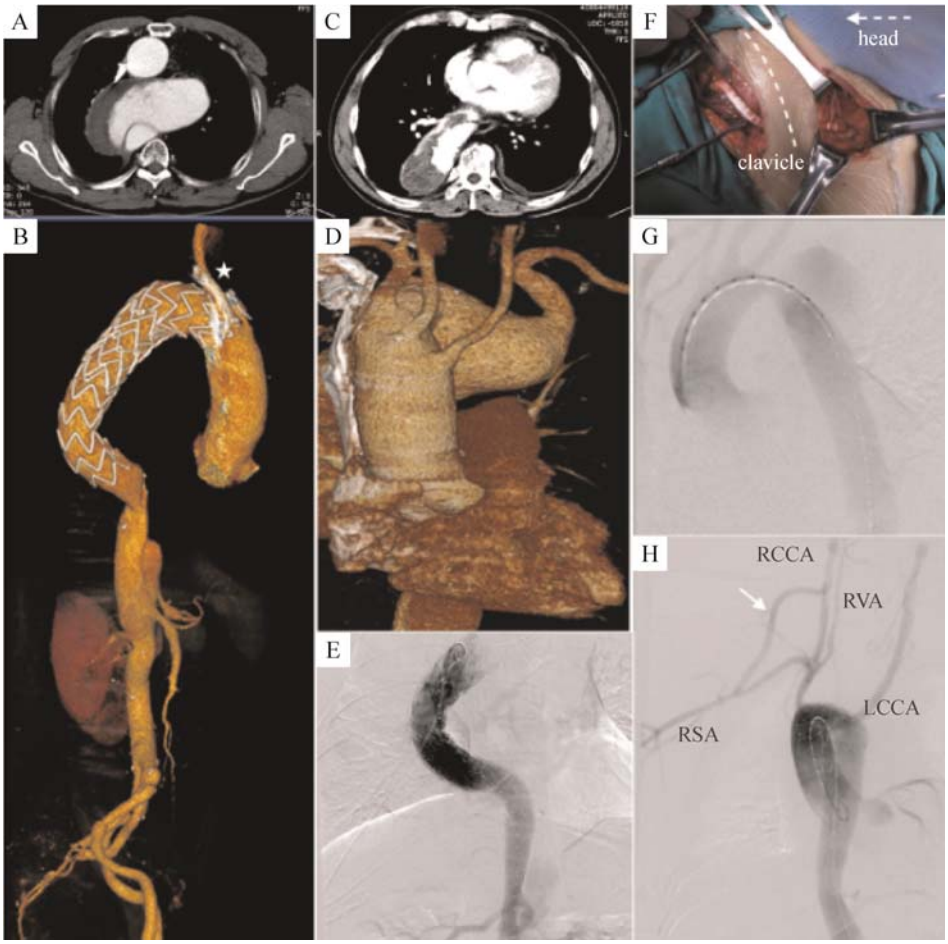

Fig. 1 **Images of Case 1 (A, B, C, D, E) and Case 2 (F, G, H).** A: At the level of the descending aorta, a dissection flap is seen in a right-sided arch. B: Stent-graft implantation with RSA chimney graft. C: A stent graft-induced distal redissection after thoracic endovascular aortic repair. D: The preoperative CTA 3D reconstruction view. E: After the secondary TEVAR, redissection was completely excluded. F: A right carotidsubclavian bypass. G: An impending DeBakey III dissected aneurysm in patient No. 3. H: Angiograph confirms the patency of the bypass, shows that the false lumen is completely excluded and the true lumen expands well. LCCA: left common carotid artery; RCCA: right common carotid artery; RSA: right subclavian artery; RVA: right vertebral artery; ☆: chimney graft.

was uneventful. A postoperative CT scan in 10th month showed thrombosis of the false lumen and a new-onset distal intimal tear (***Fig. 1C***), therefor an endovascular treatment was performed with extra stent-grafts (28 mm × 80 mm, Optimed and 36 mm × 36 mm × 80 mm, Shanghai MicroPort) (***Fig. 1E***). He remained asymptomatic 2 months after the procedure. No complaint has been reported so far.

Case 2

A 34-year-old man was intubated and transferred to our hospital because of an impending thoracic aneurysm and massive hemoptysis. In emergent operation, the angiograph revealed a type II right-sided arch and an impending DeBakey III dissected aneurysm tearing from the Kommerell's diverticula (***Fig. 1G***). A right carotid-subclavian bypass was first performed with a 6-mm straight woven Dacron graft with two separate incisions (***Fig. 1F***). Thereafter, an endoprostheses (32 mm × 26 mm × 180 mm, Grikin, China) were inserted through the right femoral artery and deployed from the orifice of his RCCA to the descending aorta. The LSA arising from the Kommerell's diverticulum was already occluded. The completion angiograph revealed a successful exclusion with no endoleak (***Fig. 1H***). His postoperative course was uneventful and discharged postoperative day 7.

Case 3

A 62-year-old male patient admitted from our emergency center in shock situation. CT scan revealed contained rupture of a dissected aneurysm arising from the Kommerell diverticulum and a great deal of ipsilateral pleural effusion (***Fig. 2A***). Emergency surgery was performed. We deployed a distal restrictive stent (24 mm × 80 mm Optimed, USA) through

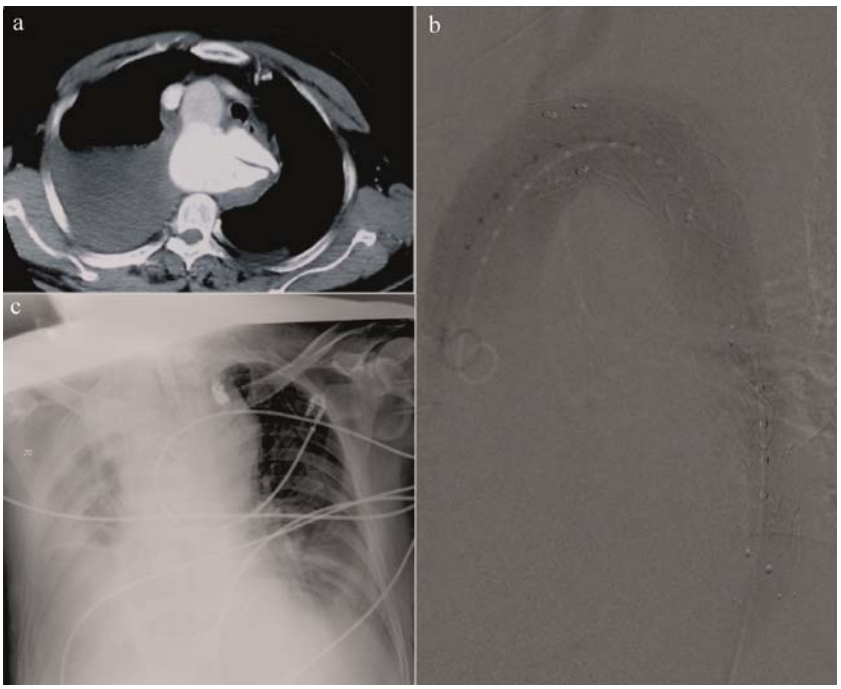

Fig. 2 **Images of Case 3.** A: A ruptured dissected aneurysm arising from the Kommerell diverticulum. B: Intraoperative angiograph showed complete exclusion of the aneurysmal sac. C: A plain chest film showed right pleural effusion 1 hour before death.

the vasculature, according to standard practice in the descending aorta at the T6 level. A Captivia stent-graft (30 mm × 30 mm × 200 mm Medtronic, USA) was then advanced into position and deployed just distal to the RSA precisely, overlapping 2 cm with the restrictive stent. After release, an additional angioplasty of left lower limb was performed with the use of two bare stents (12 mm × 80 mm, Zilver Cook) from retrograde puncturing of ipsilateral femoral artery. The completion angiogram revealed the successful exclusion (***Fig. 2B***) and restored left lower limb artery patency (not shown). Unfortunately, the patient died of a sudden massive hemodynamic shock 36 hours after the surgery (***Fig. 2C***). The family denied an autopsy.

Case 4

A 56-year-old man admitted for chest pain. CTA revealed an Edwards type I RAA with mirror imaged branches and an intimal dissection that originated distally to the RCCA opening. A GORE TAG Thoracic Endoprosthesis stent graft (38 mm × 32 mm × 180 mm) was then deployed. A completion angiogram showed occlusion of the origin of the left subclavian artery without evidence of any type of endoleak, and successful exclusion of the dissection. His follow-up computed tomographic scans have shown no evidence of endoleak, including the most recent scan performed 21 months after the operation (***Fig. 3***).

Fig. 3 **Complete occlusion of the false lumen in patient No. 4 (mirror arch).**

Discussion

Right-sided aortic arch that occurs in approximately 0.1% of the population is a rare variant of the thoracic

Table 1 **Major characteristics of studies involving aortic dissections with RAA**

Treatment	*n*	Gender		Outcome		Age (year)
				alive	death	
Open Surgery	13	Male	9	8	1	53.2 in avg.
		Female	4	1	3	62 in avg.
Endovascular	1	Male	1	1	n/a	62
		Female	n/a	n/a	n/a	n/a

vascular anatomy[1]. Approximately 40% of right-sided arches are associated with an aberrant left subclavian artery[4]. Kommerell's diverticulum occurs in this kind of abnormality with the RAA. The indications for surgical intervention as regards the size of the Kommerell's diverticulum are not clear because of the limitied reports of the anomaly[5]. As a result of the great stress during a cardiac cycle to the proximal descending thoracic aorta and of decreased vessel tissure density in dilated aneurysms, patients with Kommerell's diverticulum are predisposed to dissections. In former studies, most cases of aortic dissection with right-sided arch were treated open-surgically (***Table 1***)[1,3] We found a series of type III aortic dissections arising in the Kommerell's diverticulum. In all of our cases, the dissected tissue extended from the distal aortic arch through the thoracoabdominal aorta, which may increase operative difficulty and in-hospital mortality. Therefore, though such patients would be asymptomatic, we recommend an aggressive early surgical intervention because elective endovascular interventions are relatively safe. Since Yamazaki and Okada[6-7] successively established a feasible endovascular stent implantation strategy, endovascular prosthesis has developed rapidly and is now considered as an appealing alternative method for a descending thoracic aortic aneurysm with RAA. Recently, Croccia and colleagues have described the first case of a type B dissection associated with RAA treated with an endovascular approach[3]. Major advantages, compared with a standard surgical technique, include the lower level of invasiveness and the avoidance of more complex repairs and of the need for circulatory arrest. However, the management of acute type III aortic dissection in RAA patients is still one of the great challenges for vascular surgeons. The acute phase is defined as < 14 days from the onset of symptoms. An acute type III dissection is considered complicated when there is rupture or impending rupture, pain or hypertension refractory to medical therapy, progression of the dissection despite optimal medical therapy, and visceral or limb ischemia. In case 3, the patient presented evidence of impending rupture and malperfusion of his viscera and left lower extremities. The complicated acute aortic dissection requires an urgent intervention. Clearly, open surgery has significant risks and carries a high morbidity[8]. The advent of endovascular therapy is an appealing management of complicated acute aortic dissection, and offers an opportunity for improved outcomes and definitive treatment of this disease[9]. Patient 3 was believed to die of aorta rupture. However, because an autopsy was not performed, it is not justified to draw conclusions about the cause of aorta rupture, and we can only speculate that angulation and curvature at the aortic arch, pathological change of the aortic wall and involvement of the stent graft placed close to the top of lesser curvature seem to be risk factors. Successful TEVAR requires flat, straight, long, and regularly cylindrical landing zones, while a curve, severe taper and luminal surface irregularity continues to be a technical challenge. An RAA commonly has a greater curvature than that of a normal left aortic arch and exhibits a tortuous access route, which made an accurate deployment more difficult when performing TEVAR in this patient. Although an emergency surgery is now not recommended for stable patients with aortic dissection, it is sometimes the only option in cases with threatening major complications.

The necessity for revascularization of the LSA is controversial. Coverage of the LSA without revascularization may be justified only if thorough investigations on cerebral and vertebrobasilar circulation showed an acceptable low risk to the brain and spinal cord. In our series, we also accepted patients with landing zones < 2 cm. In such cases, overstenting of the left subclavian artery was performed. An emergency subclavian revascularization should be considered once the blood supply to the brain or spinal cord is interrupted. Strategies for handling the insufficiency of the proximal landing zone during endovascular repair are another major concern. In patient 1 and 3, the anatomic features made the landing zone between the RSA and the thoracic aortic intimal tear not sufficient to accommodate a stent-graft. Although good results have been obtained with fenestrated grafts, they have to be custom-made for most individual patients, which is a time-consuming and expensive procedure. In addition,

in situ fenestration refers to the slotting in the sealing region, which damages the integrity of the endograft and also increases the risk of endoleak[10]. For a promising long-term patency of carotid-subclavian artery bypass, a hybrid technique procedure (right carotid-subclavian bypass and graft stenting) was adopted in the 33-year-old patient. The chimney graft, which involves placement of a parallel stent in side branches of the aorta alongside the main endoprostheses, offers a readily available endovascular alternative in our elderly patients. In our observation, the E-Luminexx stent provided adequate radial strength to resist compression from the thoracic endograft and the tortuous aorta arch in the RAA and avoided overstenting the RCCA. The "chimney" is simple to use in those cases without sufficient landing zone, but it is still a disputed technique. We may try more bypass-hybrid procedure in future treatments. That is more convincing to achieve better prognosis for patients that do not have chance to obtain a fenestrated graft.

In conclusion, our recent experiences suggest that endovascular repair could be a more feasible option in treatment of RAA patients with a DeBakey type III aortic dissection, in comparison with conventional open surgical repair. However, the extra risks of emergent endovascular treatment should always be taken into account. Emergency surgery should be never considered in patients with stable hemodynamic situations. A hybrid procedure with bypass-reconstruction of the RSA is more convincing to achieve better prognosis for patients that do not have chance to obtain a fenestrated graft. In most cases, the aberrant LSA could be occluded safely. More evidence-based data are needed to certify long-term safety and efficacy of endovascular repair in treatment of thoracic aortic dissection associated with right-sided aortic arch.

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