Clinical Case Report

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Application of thoracic endovascular aortic repair (TEVAR) in treating dwarfism with Stanford B aortic dissection

A case report

Jian Qiu, MD, Wenwu Cai, MD, Chang Shu, MD*, Ming Li, MD, Qinggen Xiong, MD,
Quanming Li, MD, Xin Li, MD

Abstract

Rationale: To apply thoracic endovascular aortic repair (TEVAR) to treat dwarfism complicated with Stanford B aortic dissection.

Patient concerns: In this report, we presented a 63-year-old male patient of dwarfism complicated with Stanford B aortic dissection successfully treated with TEVAR.

Diagnoses: He was diagnosed with dwarfism complicated with Stanford B aortic dissection.

Interventions: After conservative treatment, the male patient underwent TEVAR at 1 week after hospitalization. After operation, he presented with numbness and weakness of his bilateral lower extremities, and these symptoms were significantly mitigated after effective treatment. At 1- and 3-week after TEVAR, the aorta status was maintained stable and restored.

Outcomes: The patient obtained favorable clinical prognosis and was smoothly discharged. During subsequent follow-up, he remained physically stable.

Lessons: TEVAR is probably an option for treating dwarfism complicated with Stanford B aortic dissection, which remains to be validated by subsequent studies with larger sample size.

Abbreviations: CSF = cerebrospinal fluid, CT = computed tomography, CTA = computed tomography angiography, MRI = magnetic resonance imaging, SCI = spinal cord ischemia, TEVAR = thoracic endovascular aortic repair.

Keywords: dwarfism, Stanford B aortic dissection, thoracic endovascular aortic repair

1. Introduction

Stanford B aortic dissection occurs when the blood flows between the layers of the wall of aorta. It can rapidly lead to death due to insufficient blood flow into the heart or the aorta rupture, which is a severe and fatal threat to the affected patients and constantly requires emergent management.[1] Considering the varying symptoms of Stanford B aortic dissection relying upon the degree of the dissection, the final diagnosis is difficult to deliver. Hence, the imaging techniques, such as chest X-ray, computed tomography (CT), magnetic resonance imaging (MRI), and transesophageal ultrasound, are performed. Endovascular stent graft was first introduced in the treatment of aortic dissection by Dake et al[2] in 1999, which was gradually applied by many surgeons.[3] In this article, we reported a 63-year-old male dwarf complicated with Stanford B aortic dissection. We applied thoracic endovascular aortic repair (TEVAR) in treating such condition for the first time. Clinical efficacy and safety were evaluated by subsequent follow-up.

2. Case report

A 63-year-old male dwarfism with a height of 1.30 m was admitted to our hospital due to the complaints of acute-onset chest and back pain for 10 hours, as illustrated in Fig. 1. The patient was diagnosed with hypertension 8 years and the high blood pressure was uncontrolled. Upon admission, the blood pressure was measured as 175/68 mm Hg with a pulse rate of 83 beats/minute. The white blood cell count was 14.18×10^9/L including 94.81% neutrophilia. No other abnormalities or complications were noted. Computed tomography angiography (CTA) revealed multiple aortic ulcers distal to the origin of the left common carotid artery with local aortic dissection in the descending aorta. Other vital visceral arteries were not affected, as illustrated in Fig. 2A. Pleural effusion was observed on the left side and blood flow was noted in the left subclavian artery (Fig. 2B). Written informed consent was obtained from the patient for publication of this case.

The patient was hospitalized and underwent TEVAR. CTA detected multiple aortic ulcers distant from the left common carotid artery with local aortic dissection, as the arrows indicated

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Both JQ and WC contributed equally to this article.

The authors report no conflicts of interest.

Department of Vascular Surgery, Second Xiangya Hospital, Central South University, Changsha, Hunan, P.R. China.

* Correspondence: Chang Shu, Department of Vascular Surgery, Second Xiangya Hospital, Central South University, No. 139, Renmin Road, Changsha 410011, Hunan, P.R. China (e-mail: shuchang@csu.edu.cn).

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Figure 1. A picture illustrating the body shape of the patient.

in Fig. 3. The vascular stents were selected based upon the measurement of the vascular diameter. Two modes of covered vascular stents of 32 to 26 mm × 180 mm and 30 to 24 mm × 180 mm in size were employed (Lifetech Scientific Co., Ltd., Shenzhen, China). Repeated CTA demonstrated that the first stent covered the origin of the left subclavian artery. Two stent grafts were overlapped for 8 cm. No stent endoleak or dislocation occurred. The ulcers and false lumen disappeared (Fig. 3). The patient remained stable at 1 day after TEVAR, and he suddenly presented with numbness and weakness of bilateral lower extremities. He could merely move in the horizontal direction on bed. The possibility of paraplegia induced by spinal cord ischemia (SCI) was considered. Multiple interventions, including maintaining systolic blood pressure at 120 to 140 mm Hg, administration of methylprednisolone at a dose of 60 mg/day, subcutaneous injection of low-molecular-weight heparin at a dose of 12500 U/12 hours, intravenous drip of mannitol at a dose of 250 mL/12 hours, and neurotrophin were delivered immediately. These symptoms were significantly mitigated within 1 week after TEVAR, though the patient still felt mild numbness of bilateral lower extremities. He was able to rise and walk with external support. Repeated CTA detected that the innominate artery, the left common carotid artery, and other visceral arteries were patent, and the aortic ulcers and false lumen disappeared. No stent endo-leak or dislocation was observed. The patient was discharged at 2 weeks following hospitalization. During subsequent 3-month follow-up, he could independently rise and walk. Repeated CTA revealed that the branches of aorta were restored, and the left pleural effusion was mitigated. He reported no other abnormalities or postoperative complications.

3. Discussion

Since the introduction of aortic stent grafting for the treatment of thoracic aortic aneurysms, TEVAR has gained widespread clinical application as a less invasive technique for Stanford B aortic dissection compared with the open surgery. However, the use of TEVAR is associated with the risk of postoperative complications, which probably result in poor long-term survival and even high rate of morbidity and mortality. Indications for TEVAR in treating thoracic aortic aneurysms include paraplegia, visceral ischemia, acute rupture, chronic aneurysm, etc. Paraplegia induced by SCI is the most severe acute complication after

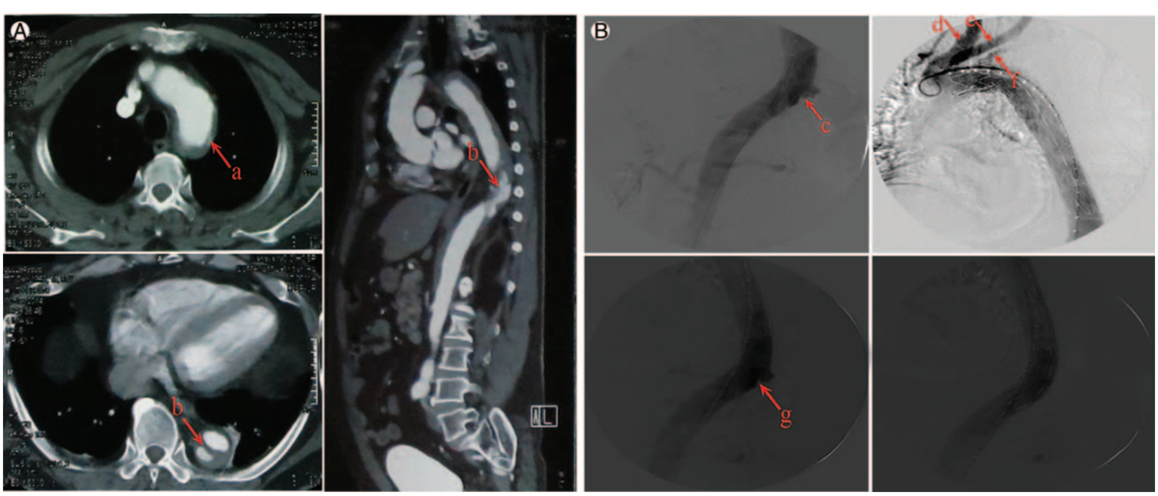

Figure 2. (A) The findings of preoperative CTA examination. The arrow (A) indicated the aortic ulcers, and the arrow (B) indicated the local aortic dissection. (B) The findings of CTA examination at 1 week after TEVAR illustrating the conditions of innominate artery (C), left common carotid artery (D), and other visceral arteries were restored normally; the aortic ulcers and false lumen disappeared, but there were still residual pleural effusion (E). Blood flow was noted in the left subclavian artery (F). The aortic branches were properly restored, the aortic ulcers and false lumen disappeared, and the sign of the left pleural effusion disappeared (G).

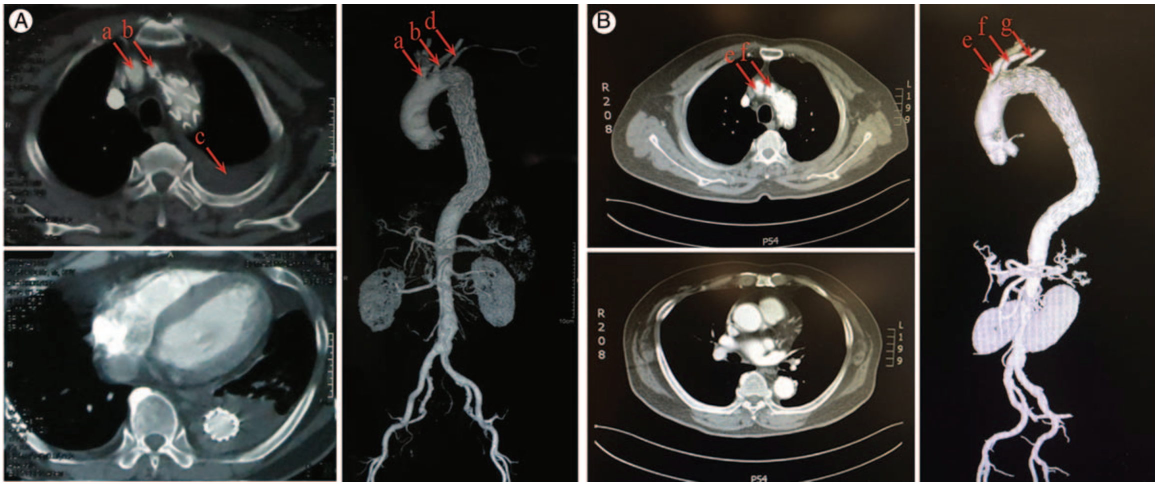

Figure 3. (A, B) The findings of the initial aortography illustrating multiple aortic ulcers (A) and local aortic dissection in the descending aorta (B). The findings of the aortography after vascular stents were released. When the first covered stent was released, the conditions of innominate artery (C) and the left common carotid artery (D) were restored well, but the local aortic dissection (B) was not repaired, which disappeared after the release of the second covered stent.

TEVAR.[4] Previous studies have demonstrated that the incidence of both immediate and delayed paraplegia in patients undergoing TEVAR can be as high as 12%, compared with 2% to 21% in their counterparts after open surgery.[5,6]

The mechanism underlying the incidence of paraplegia post-TEVAR remains to be completely understood. Numerous studies have been conducted to identify the risk factors of paraplegia following TEVAR, potentially including the left subclavian artery coverage without prior revascularization, the use of multiple stent grafts, episode of hypotension, concomitant open abdominal aorta surgery, renal failure, severe atherosclerosis of the thoracic aorta, aneurysm as an underlying pathology, and use of iliac conduit as vascular access, etc.[4,6–8]

In our patient, the intentionally covered left subclavian artery was delivered without prior revascularization to achieve an ideal repair. Compared with healthy individuals, the relatively shorter aorta probably compromises the spinal cord derived blood supply and contributes to the progression of paraplegia.

In this case, the local aortic dissection could not be covered after the placement of the first stent of 32 to 26 mm × 180 mm in size. Subsequently, we had to adopt the second stent of 30 to 24 mm × 180 mm in size, which potentially aggravated the severity of paraplegia. Consequently, it is highly recommended to retain the left subclavian artery and shorten the length of the aorta covered by the stent graft by experienced surgeons. Repeated CTA at 1 week and 3 months after TEVAR revealed that there was still blood flow in the left subclavian artery, probably owing to the retrograde flow from the left vertebral artery, which may accelerate the recovery of the patient.

Early diagnosis and treatment of postoperative paraplegia is of great significance to prevent further aggravation. Multiple measures, including augmentation of arterial pressure by volume expansion and vasopressor therapy, use of steroids, anti-coagulation, and administration of cerebrospinal fluid (CSF) drainage, have been employed to manage the paraplegia after TEVAR.[7,8] A randomized trial showed that lumbar CSF drainage could decrease the risk of SCI and reduce neurologic deficits after open thoracoabdominal aortic surgery.[9] However, the CSF hemodynamics differs between the endovascular and open repair of thoracic aortic lesions, which remain to be studied.[10] Fatic et al[11] analyzed the morphological predictors of in-hospital mortality in acute type III aortic dissection and demonstrated that inner curvature intimal tear localization, large intimal tear, and elliptic shape of true lumen are recommended. A previous study [12] compared the advantages of different endovascular techniques in the treatment of complicated aortic dissection. CSF drainage is an invasive operation, probably leading to severe postoperative complications, including spinal headache, epidural or subdural hematoma, infection or even abscess, etc. Therefore, much attention should be paid before CSF drainage to prevent the risk of severe postoperative complications. In the present case, although the patient did not receive CSF drainage, he still fully recovered. Al-Jughiman et al [13] previously reported 1 male case of achondroplastic dwarfism who presented with acute type-A aortic dissection with aortic insufficiency. He underwent Bentall and hemiarch repairing surgery. However, this surgical technique might be extremely complicated if the patients' thorax size is irregular and abnormal. In addition, it is challenging to precisely identify the position of hypoplastic femoral arteries. Consequently, the application scope of this surgical technique is probably limited and constrained.

4. Conclusion

After full preoperative preparation, precise intraoperative localization, and intimate postoperative monitoring, TEVAR is probably an option for the treatment of the dwarf complicated with Stanford B aortic dissection. Nevertheless, the clinical efficacy and safety remain to be validated by further investigations with larger sample size and longer postoperative follow-up.

Author contributions

Conceptualization: Jian Qiu, Wenwu Cai, Chang Shu, Qinggen Xiong.
Data curation: Chang Shu, Ming Li, Quanming Li.
Formal analysis: Xin Li.
Investigation: Quanming Li.
Methodology: Qinggen Xiong, Quanming Li, Xin Li.
Project administration: Jian Qiu, Wenwu Cai, Chang Shu.

Resources: Jian Qiu, Wenwu Cai.
Writing – original draft: Ming Li, Qinggen Xiong, Xin Li.
Writing – review & editing: Ming Li, Xin Li.

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