

A Right-sided Aortic Arch with Kommerell's Diverticulum of the Aberrant Left Subclavian Artery Presenting with Syncope

Ming-Hsun Yang¹, Zen-Chung Weng¹, Yu-Guo Weng², Hsiao-Huang Chang^{1*}

¹*Division of Cardiovascular Surgery, Department of Surgery, Taipei Veterans General Hospital, Taipei, Taiwan, R.O.C., and* ²*German Heart Institute Berlin, Berlin, Germany.*

A right-sided aortic arch with an aneurysm of the aberrant subclavian artery is a rare disease. We report a case of Kommerell's diverticulum of an aberrant left subclavian artery in a patient with a right-sided aortic arch. Fewer than 50 cases have been reported in the literature. A number of operative strategies are described. Right thoracotomy provides good exposure and avoids the morbidity associated with bilateral thoracotomy or sternotomy and thoracotomy. In our patient with symptoms of dysphagia, syncope, and left subclavian steal syndrome, a left thoracotomy was used. The repair was accomplished by division of a left ligamentum arteriosum, obliteration of the Kommerell's aneurysm, and an aorto-subclavian bypass. Postoperative complications included left vocal cord palsy and Horner's syndrome. Hoarseness and left ptosis recovered spontaneously 3 months after surgery, and the patient remained symptom-free at the 1-year follow-up. We believe a left thoracotomy for direct repair of Kommerell's diverticulum is a simple and safe method without the increased morbidity found in other procedures. [*J Chin Med Assoc* 2009;72(5):275-277]

Key Words: aberrant left subclavian artery, Kommerell's diverticulum, right-sided aortic arch, subclavian steal syndrome, syncope

Introduction

In 1936, Burckhard Friedrich Kommerell described the aortic diverticulum that bears his name.¹ According to the literature, the right subclavian artery arises as the last branch of the aortic arch and courses from the proximal descending aorta to the right arm, passing behind the esophagus. The aneurysmal diverticulum of the descending aorta at the origin of an aberrant right subclavian artery is called Kommerell's diverticulum, which consists of both an aneurysm of the thoracic aorta and an aneurysmal orifice of the aberrant subclavian artery. In this article, we report a patient with right-sided aortic arch in whom an aneurysm of the aberrant left subclavian artery (ALSA) presented with syncope.

Case Report

A 32-year-old man was referred for assessment of vascular anomaly. He had complained of occasional heartburn sensation and dysphagia since the age of 25 years. Esophagogastroduodenoscopy showed gastroesophageal reflux, and proton pump inhibitor was administered. He was a medical doctor, and had experienced 3 occasions of syncope during his clerkship when he was 29 years old. All the incidents had occurred when he was working as an assistant during surgery. Each time, he experienced visual field darkness initially, then cold sweating and tinnitus, before finally fainting. On physical examination, right arm blood pressure was higher than the left by 20 mmHg. Pulsations of the left brachial and radial artery were faint. Doppler



*Correspondence to: Dr Hsiao-Huang Chang, Division of Cardiovascular Surgery, Department of Surgery, Taipei Veterans General Hospital, 201, Section 2, Shih-Pai Road, Taipei 112, Taiwan, R.O.C. E-mail: shchang@vghtpe.gov.tw • Received: June 18, 2008 • Accepted: January 6, 2009

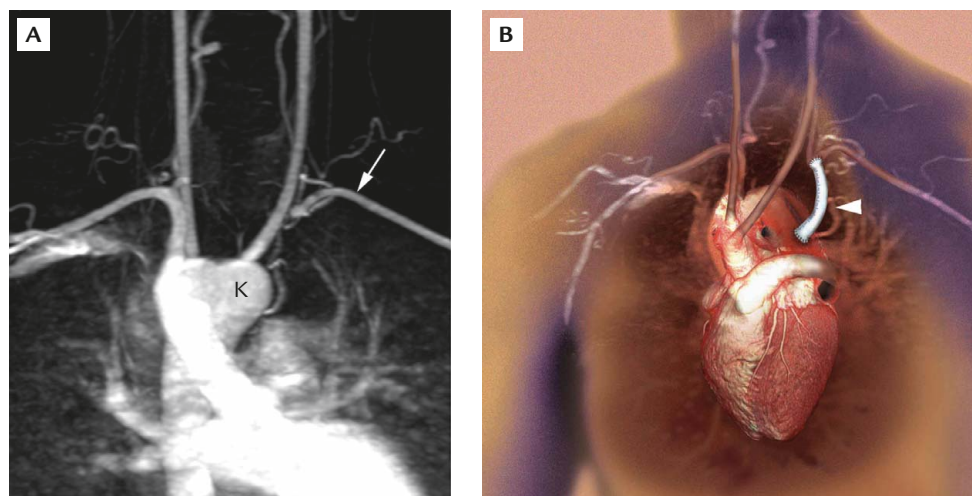


Figure 1. (A) Magnetic resonance angiography with 3-dimensional reconstruction demonstrates right-sided aortic arch with aberrant left subclavian artery (arrow) and Kommerell's diverticulum (K). (B) An illustration shows the obliteration of Kommerell's diverticulum and the aorto-subclavian bypass by artificial conduit (arrowhead).

ultrasound of the neck disclosed a reverse flow of the left vertebral artery. Chest computed tomography (CT) and magnetic resonance angiography displayed right-sided aortic arch with dilatation of the posterior part near the supposed ostium of an ALSA, assuming the appearance of a Kommerell's diverticulum (Figure 1A). The size of the Kommerell's diverticulum was about $2.9 \times 2.9 \times 2.8$ cm, with posterior decompression of the esophagus. Barium swallow esophagography also showed an extrinsic compression of the esophagus (Figure 2A).

Surgery was indicated for left subclavian steal syndrome and vertebrobasilar insufficiency. General anesthesia with a double lumen intubation was performed, followed by a left thoracotomy through the fifth intercostal space. The left subclavian artery arose as the last branch of the right-sided aortic arch and coursed from the proximal descending aorta to the left arm, passing behind the esophagus. The ALSA was occluded because of thrombosis and fibrosis. The aneurysmal diverticulum of the descending aorta at the origin of the ALSA, called Kommerell's diverticulum, was compressing the esophagus. The ligamentum arteriosum between the Kommerell's diverticulum and the origin of the left pulmonary artery was also compressing the esophagus. The vagus nerve and phrenic nerve were preserved. Division of an intact left ligamentum arteriosum sufficed to relieve external compression of the esophagus. A Satinsky clamp was placed on the saccular-type aneurysm without compromise of the descending aorta. Kommerell's diverticulum was obliterated by several pledgeted stitches of 3-O prolene continuously. Finally, aorto-subclavian bypass was achieved by a 6-mm GoreTex graft. The proximal end

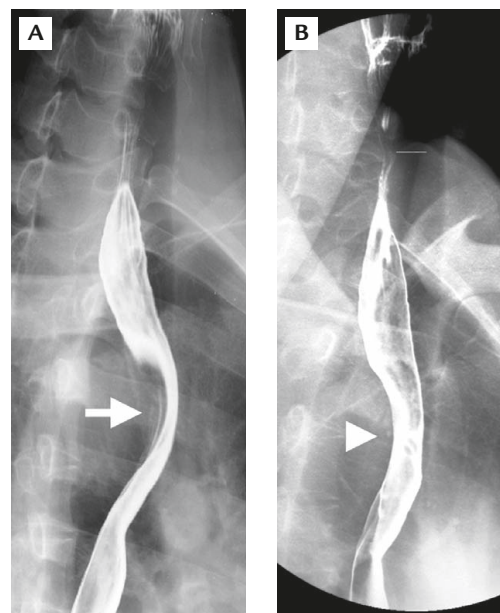


Figure 2. (A) Barium swallow esophagography before surgery shows significant compression of the esophagus by Kommerell's diverticulum (arrow). (B) Barium swallow esophagography after surgery shows a regressive change (arrowhead).

of the conduit was anastomosed to the descending aorta below Kommerell's diverticulum by 3-O prolene in running fashion, as was the distal end to the origin of the left subclavian artery (Figure 1B).

The patient was promptly extubated in the recovery room. He was able to swallow liquids on the second postoperative day, and then advanced to solid foods. Pulsations of the left brachial and radial artery were strong. Hoarseness because of left vocal cord palsy was confirmed by laryngoscopy. It improved significantly

1 week later. Horner's syndrome with left eyelid ptosis and left pupil miosis were noted but improved 3 months later. Barium swallow esophagography 2 weeks after surgery showed significant regression of external compression on the esophagus (Figure 2B). The results of chest CT 3 and 8 months after surgery were satisfying. The patient had no recurrence of syncope 1 year after surgery.

Discussion

Kommerell's diverticulum can occur in several anomalies of the aortic arch system, even in association with a left aortic arch with aberrant right subclavian artery (0.5–2.0% of the population),² or a right aortic arch with ALSA (0.05–0.1%).^{3,4}

According to a review by Cina and colleagues,⁵ there are many surgical strategies for repair of Kommerell's diverticulum. Reconstruction of the left subclavian artery is necessary to prevent arm claudication and subclavian steal syndrome. A left subclavian-to-carotid transposition is performed using the technique described previously.⁶ Surgery is performed in 2 stages during the same operative procedure. Thoracotomy, partial left-heart bypass, and endoaneurysmorrhaphy can be carried out before or after the left subclavian-to-carotid transposition. A right thoracotomy provides good exposure of the ascending transverse arch and descending thoracic aorta to allow reconstruction of these arteries and repair of Kommerell's diverticulum.

We abandoned the 2-stage surgery and the use of cardiopulmonary bypass. Endoaneurysmorrhaphy is complicated, and the use of partial cardiopulmonary or deep hypothermic circulatory arrest could not be avoided. An extended left thoracotomy provides good exposure for 1-stage surgery. Direct obliteration of Kommerell's diverticulum with pledgeted stitches of 3-0 prolene under assistance of a Satinsky clamp is a simple and safe method. The Satinsky clamp is placed on the saccular diverticulum and we kept the descending aorta patent. The use of cardiopulmonary bypass or deep hypothermic circulatory arrest is unnecessary. A left thoracotomy also provided good exposure for aorto-subclavian bypass. The GoreTex conduit was anastomosed between the descending aorta below Kommerell's diverticulum and the left subclavian without difficulty.

Syncope is a sudden and brief loss of consciousness associated with loss of postural tone.⁷ This patient experienced 3 episodes of syncope while acting as an assistant during operations. These operations were a laparoscopic appendectomy, a breast lumpectomy, and a circumcision. A sudden decrease in cerebral flow by left subclavian steal syndrome is reasonable. Interestingly, he did not use the left hand laboriously during these operations. Perhaps a vasovagal attack induced by the unpleasant smell during electrocauterization partly contributed to the syncope.

Dysphagia improved significantly after surgery, but the heartburn did not. Barium esophagography after surgery showed gastroesophageal reflux. Esophageal manometry is risky for this patient. The transient relaxation of both the lower esophageal sphincter and the crural diaphragm contributed to the gastroesophageal reflux.⁸

The neurologic complications included left recurrent nerve palsy and Horner's syndrome. A surgical contusion which resulted in temporary neurapraxy may be a better explanation. Fortunately, it was self-limited, and the patient recovered 3 months later. In conclusion, a left thoracotomy with direct repair of the diverticulum and intrathoracic reconstruction of the left subclavian artery is a simple and safe method for management of Kommerell's diverticulum and aberrant subclavian artery.

References

1. Kommerell B. Verlagerung des ösophagus durch eine abnorm verlaufende arteria subclavia dextra (arteria lusoria). *Fortschr Geb Roentgenstrahlen* 1936;54:590–5.
2. Freed K, Low VH. The aberrant subclavian artery. *AJR Am J Roentgenol* 1997;168:481–4.
3. Tsukube T, Ataka K, Sakata M, Wakita N, Okita Y. Surgical treatment of an aneurysm in the right aortic arch with aberrant left subclavian artery. *Ann Thorac Surg* 2001;71:1710–1.
4. Svensson LG, Crawford ES. Congenital anomalies of the aorta in adults. In: Svensson LG, Crawford ES, eds. *Cardiovascular and Vascular Disease of the Aorta*. Philadelphia: Saunders, 1997: 153–74.
5. Cina CS, Althani H, Pasenau J, Abouzahr L. Kommerell's diverticulum and right-sided aortic arch: a cohort study and review of the literature. *J Vasc Surg* 2004;39:131–9.
6. Cina CS, Safar HA, Lagana A, Arena G, Clase CM. Subclavian carotid transposition and bypass grafting: consecutive cohort study and systematic review. *J Vasc Surg* 2002;35:422–9.
7. Kapoor WN. Syncope. *N Engl J Med* 2000;343:1856–62.
8. Mittal RK, Balaban DH. The esophagogastric junction. *N Engl J Med* 1997;336:924–32.